

Surgical & Cosmetic Dermatology

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Surgical & Cosmetic Dermatology

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Subcutaneous pedicle flap shaped like a shark on perinasal region

Retalho de pedículo subcutâneo em formato de tubarão na região perialar

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ABSTRACT

The nasal region is often affected by cutaneous neoplasm, especially in individuals of low phototypes, where the incidence of basal cell carcinoma is high. Surgical defects in the nasal wing and perinasal region constitute a challenge to its reconstruction since it involves several cosmetic units and preservation of the nasal groove. The preservation of the limits between these units is thus fundamental for good functional and aesthetic results. This article aims to show the application of the Shark Island Flap for the correction of defects in the nasal and perinasal regions.

Keywords: Nasal Surgical Procedures; Neoplasms, Basal Cell; Nose Neoplasms

RESUMO

A região nasal é frequentemente acometida por neoplasias cutâneas, especialmente em indivíduos de fototipos baixos, em quem a incidência de carcinoma basocelular é elevada. Defeitos cirúrgicos na asa nasal e região perinasal constituem desafio à sua reconstrução, uma vez que envolve várias unidades cosméticas e preservação do sulco nasal. A preservação dos limites entre essas unidades mostra-se fundamental, portanto, para o bom resultado funcional e estético. Este artigo tem como objetivo mostrar a aplicação do retalho de pedículo subcutâneo em formato de tubarão para correção de defeitos em asa nasal e região perinasal.

Palavras-chave: Carcinoma Basocelular; Neoplasias Nasais; Procedimentos Cirúrgicos Nasais

INTRODUCTION

Skin neoplasms frequently affected the nasal region, especially in individuals with low skin phototypes, in whom the incidence of basal cell carcinoma is high.

According to a study conducted at the Brazilian National Cancer Institute (INCA), basal cell carcinoma was responsible for nasal defects in which partial or subtotal nose reconstructions were performed. Moreover, squamous cell carcinoma was the main responsible in cases of total nasal reconstruction.

Non-melanoma skin cancer is the most frequent among Brazilian men and the second among women. Approximately 93% of basal cell carcinomas occur in the head and neck and, of these, 25% appear in the nasal pyramid. Thus, it is undoubtedly the most common cancer of the head and neck.¹

The nose region has unique characteristics that surgeons should consider when planning the surgical reconstruction of

Case report

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the defect. The complex topography, with concave and convex surfaces, needs to be preserved. The nasal wing's free margin can be easily raised and distorted if the reconstruction is not carefully planned. The lower third of the nose's skin has reduced mobility and is not easily recruited for primary wound closure without causing anatomical distortions. It is also a region with a unique texture and coloring characteristics, not resembling the surrounding skin. Finally, the nose's functional aspect must always be considered, preserving the bone, cartilage, and mucosa structure, never compromising the airway.²

The lateral nasal wing, lateral side of the nose, malar region, and supralabial skin delimit the nasal and perinasal region.³

Surgical defects involving this region are challenging at the time of its reconstruction. This is because the combination of these defects involves several cosmetic units of the face, and transgressing from one unit to another, at the time of reconstruction, can lead to the erasure of the nasal sulcus and the loss of the limits of these units, causing an unsatisfactory aesthetic result.⁴

Options for managing deep defects in the nasal wing and perinasal region include primary closure, healing by secondary intention, local flaps, interpolation flaps, and skin grafts.

Performing a flap using skin adjacent to the surgical defect is ideal for obtaining a good functional and cosmetic result. In this case, the subcutaneous shark island pedicle flap proved to be effective, as it is a procedure performed in a single surgical procedure, under local anesthesia, with skin that preserves the same characteristics of texture, color, and concentration of sebaceous glands in the dry area, besides preserving the nasal sulcus and maintaining the limits between the cosmetic units of the face.

CASE REPORT

A 58-year-old man presented nodular, erythematous-pigmented lesion, with telangiectasias, of about 5 mm x 6 mm, with pearly edges, in the nasal sulcus and right perinasal region, suggestive of basal cell carcinoma (Figure 1).

Due to the surgical lesion location (Figure 2), the subcutaneous shark island pedicle flap proved to be one of the options to be considered.

The pedicle flap's demarcation was performed so that the upper arm rotated 90 degrees concerning the surgical defect as if the "shark's fin" fits the alar edge of the nose (Figure 3).

Subsequently, the tissue was rotated, causing the rest of the flap to form an inverted cone of skin redundancy (Figure 4), resulting in the natural creation of the nasal wing and sulcus, without the need for anchoring the flap.

Finally, the flap's lower arm was sutured, positioning the scar along the nasolabial fold, which provided a satisfactory immediate postoperative result without altering the anatomy of the wing and nasal introitus (Figures 5 and 6).



FIGURE 1: Lesion demarcation and shark island pedicle flap



FIGURE 2: Final surgical defect



FIGURE 3: Preparation of the lower flap arm with detachment of the subcutaneous pedicle

DISCUSSION

The nasal surgical reconstruction options must contemplate the region's functional and aesthetic preservation, always trying to use techniques that camouflage the scars in the face's folds and natural lines.⁵



FIGURE 5: Sixty days after surgery. Preservation of the nasal sulcus



FIGURE 4: Redundancy cone after 90° rotation of the upper arm



FIGURE 6: Sixty days after surgery. No change in wing anatomy and nasal introitus

A surgical option in this region is healing by second intention and primary closure, quick and straightforward solutions for small defects. However, in more extensive cases, such as the one described, they can retract the nasal wing and functional impairment when modifying the nasal introitus's anatomy.

On the other hand, the skin graft is a good option for large defects, especially in areas where there is no tissue distensibility and in cases where it is not possible to delimit the intraoperative tumor area. However, professionals should avoid its use whenever possible, as it does not give a satisfactory aesthetic aspect. It may occur because it is a technique that does not always use skin with similar characteristics to the area to be reconstructed regarding color, texture, thickness, and concentration of sebaceous glands, which can lead to local hypopigmentation.

Bilobed transposition flaps would not be a good option because they are more indicated in defects more centralized in the nasal dorsum or the nose's lateral wall. Besides, they also have a chance of skin redundancy with a trapdoor effect.

An option that could be used in this surgical wound is the pedicle advancement skin flap or island pedicle flap. It preserves the anatomy of this concave region of the face and presents a pattern of myocutaneous vascularization, which gives the flap great viability.

In our case, we chose the shark island pedicle flap, which receives this name because it resembles the shape of a shark, with the "fin" and ventral mouth formed from the surgical defect.

The advancement of the flap with the rotation movement of the "shark's fin" region, spontaneously, creates an inverted cone of skin redundancy, which redefines the groove of the nasal wing, and the perinasal tissues restore the contour of the nasal wing region, maintaining a well-defined boundary between the cosmetic units of the malar and nasal regions.⁶

Therefore, the shark island pedicle flap is a surgical reconstruction technique for defects in the nasal and perinasal region, reproducible, performed in a single operative time, and offers a very satisfactory functional and cosmetic result without distorting the anatomy and of the area. ●

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Pilomatricoma (calcifying epithelioma of Malherbe) of the temporal region - a case report

Pilomatricoma (Epitelioma Calcificante de Malherbe) da região temporal - Case report

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ABSTRACT

Pilomatricoma is an uncommon benign neoplasm of hair follicles, most commonly seen in children. The tumor appears as a solitary and firm nodule, covered with normal epidermis. We report the case of a 26-year-old woman with a lesion in the temporal region. In histological sections, there was an expansive proliferation of islets of epithelial cells configured irregularly, showing the shadow of lost nuclei (ghost cells) in the central area. Pilomatricoma should be considered in the differential diagnosis of skin nodules, especially those of the head, neck, and upper limbs.

Keywords: Hair Follicle; Neoplasms; Pilomatricoma

RESUMO

O pilomatricoma constitui uma neoplasia benigna incomum de folículos pilosos, observado mais frequentemente em crianças. O tumor apresenta-se como um nódulo solitário e firme, recoberto com epiderme normal. É relatado o caso de uma paciente feminina de 26 anos com lesão na região temporal. Nos cortes histológicos, observou-se uma proliferação expansiva de ilhotas de células epiteliais conFiguradas de forma irregular, mostrando na área central sombra de núcleos perdidos (células-fantasma). O pilomatricoma deve ser considerado no diagnóstico diferencial de nódulos cutâneos, especialmente aqueles de cabeça, pescoço e membros superiores.

Palavras-chave: Folículo Piloso; Neoplasias; Pilomatricoma

INTRODUCTION

Pilomatricoma is an unusual benign neoplasm of hair follicles, first described by Malherbe and Chenantois in 1880. In 1961, Forbes and Helwig suggested the name pilomatricoma.^{1,2} Most of them appear in the first two decades of life, with a second peak occurring in older patients. A solitary and firm nodule, covered with normal epidermis, usually represents the tumor. The tumor can be solid or partially solid and cystic, with calcium deposits also observed, thus justifying the synonym “calcifying epithelioma of Malherbe”.¹ The superficial coverage of the skin can turn violet and, in some cases, bleed. It is usually solitary, but literature also describes multiple tumors.³ Most are benign and, rarely, malignant changes are mentioned in the literature.⁴

Case report

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CASE REPORT

A 26-year-old woman presented a lesion in the left temporal region, with progressive growth and intermittent pain. The physical examination revealed a solid tumor lesion of approximately 0.5 x 1.2 cm in the left temporal region, adhered to the deep plane (Figure 1). The patient reported no previous illness or similar cases in the family. The clinical evaluation and pre-operative laboratory tests did not change. Surgical resection of the lesion was performed with safety margins of 1 cm, under local anesthesia. The histological sections showed an expansive proliferation of islets of irregularly configured epithelial cells, which presented distinct boundaries, and a central stained area that showed a shadow of lost nuclei (ghost cells) (Figure 2). The examination demonstrated various stages of maturation of basaloid cells in shadow cells. Therefore, based on histopathological characteristics, the diagnosis of pilomatricoma was confirmed.

DISCUSSION

Pilomatricoma, or calcifying epithelioma of Malherbe, is a benign epithelial neoplasia. Malherbe and Chenantois described it in 1880 as a calcifying epithelioma derived from sebaceous glands.^{1,5} Forbis and Helwig later defined the origin of pilomatricoma as the hair follicle's outer root sheath.^{2,6} These



FIGURE 1: 26-year-old woman with a 0.5x1.2 cm lesion in the left temporal region adhered to deep planes

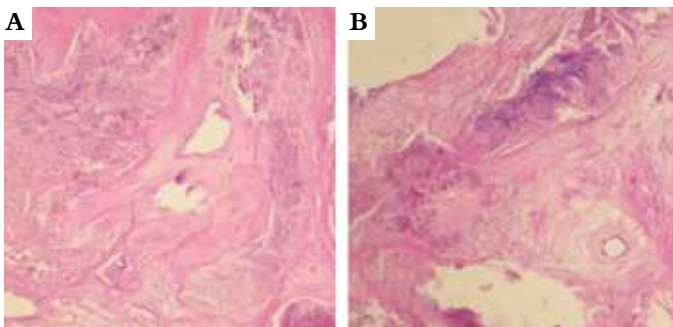


FIGURE 2: Histological examination observed an expansive proliferation of epithelial cell islands with irregular configurations but with distinct limits, showing loss of nuclei (ghost cells) (A) - Various maturation stages of basaloid cells. (B) - Hematoxylin and Eosin - original magnification X 400)

tumors are commonly present in the head and neck, but other parts of the body have also been reported. They are most common in the age group from 0 to 20 years.^{7,8} Pilomatricoma usually presents as a single asymptomatic nodule. The skin on the tumor is generally normal, but occasionally it may be reddish or bluish.⁹ It is generally well-circumscribed, with an oval or spherical shape, and can sometimes be encapsulated.^{9,10} Multiple tumors are associated with Gardner's syndrome, Turner syndrome, myotonic dystrophy, sarcoidosis, and Steinert's disease.¹¹ Malignant transformation of pilomatricoma, although reported, is rare.^{8,12} The histopathological features of pilomatricoma are a well-circumscribed tumor, lined by a capsule of connective tissue. Pilomatricoma is usually located in the subcutaneous cell tissue, composed of islands of epithelial cells made up of varying amounts of basaloid matrix cells and some cystic changes.^{6,9} As the tumor matures, there is a central degeneration of the basaloid cells that constitute the ghost cells (anucleated shadows cells) of the uncolored central area, a histopathological characteristic of pilomatricoma.^{6,9} These basaloid cells tend to decrease in number, and the ghost cells start to predominate.¹¹ The presence of an inflammatory reaction, central calcifications, giant foreign body cells, and keratin debris is also characteristic. With the use of von Kossa stain, 75% of calcium deposits are found in tumors.¹¹ Trichilemmal cysts with loss of nuclei and calcification must be histopathologically differentiated from pilomatricoma. There is a palisade pattern of peripheral basophilic cells in trichilemmal cysts that is not found in pilomatricomas.^{6,9,12} The differential clinical diagnosis of these tumors should include sebaceous, dermoid, and epidermoid cysts, as well as metaplastic bone formation, foreign body reaction, trichoepithelioma, and basal cell carcinoma.^{8,12} Surgical excision is the treatment of choice. After adequate excision, tumor recurrence is rare, and long-term follow-up is mandatory.⁶

CONCLUSION

Pilomatricomas are benign tumors of skin appendages derived from primitive basal cells of the epidermis, which differ in cells of the hair matrix. They are a relatively rare condition and are more frequent in children.

The treatment of choice is complete surgical excision of the lesion, including the skin covering it in the sample. The recurrence is not frequent, nor the occurrence of the malignant variant of the lesion.

The relative rarity of the tumor justifies this report, and health professionals differentiate it from other tumors in soft tissues. ●

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Cutaneous pilar cysts: description of an innovative technique

Cistos cutâneos no couro cabeludo: descrição de técnica inovadora

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ABSTRACT

Cysts are benign epithelial tumors widespread. The trichilemmal cysts, originating from the anagen's isthmus, represent about 20% of the cysts and are located more frequently in the scalp (90%). Several methods can be used for the removal of the cysts. The choice of technique depends on its characteristics, such as size, mobility, consistency, quiescence, inflammation, and quantity. However, they may grow again if the capsule is incomplete removed. The authors present a simple variant of the marsupialization technique that allows its complete removal.

Keywords: Ambulatory Surgical Procedures; Cysts; Scalp Dermatoses

RESUMO

Cistos são tumores epiteliais benignos extremamente comuns. O cisto triquilemal ou pilar tem origem no istmo do pelo anágeno, representa cerca de 20% dos cistos e localiza-se mais frequentemente no couro cabeludo (90%). Vários métodos podem ser usados para a retirada dos mesmos. A escolha da técnica depende de suas características, como: tamanho, mobilidade, consistência, quiescência, inflamação e quantidade. Porém, eles podem reaparecer se houver remoção incompleta da cápsula. Os autores apresentam uma variante simples da técnica de marsupialização que permite a sua retirada total.

Palavras-chave: Cistos; Couro cabeludo; Procedimentos cirúrgicos ambulatoriais

INTRODUCTION

Cysts are widespread benign epithelial tumors. They consist of a space lined by an epithelium filled with liquid or pasty content, mobile concerning the deep planes, located in the dermis or subcutaneous.^{1,2} They are usually asymptomatic and are located mainly in the cephalic segment, neck, and upper trunk.¹

Epidermoid or follicular cysts are more common in adults and can originate from the hair infundibulum or follicular ostium (comedone). The pillar or trichilemmal cyst derives from the isthmus of the anagen hair and the steatocystoma, from the sebaceous glands, therefore being true sebaceous.³

Trichilemmal cysts are less common than epidermoid cysts, representing about 20% of cysts. They have a preference for women, middle-aged adults, or older than 60 years. They are more frequent on the scalp (90%) and rare on the face, limbs, and trunk.^{4,5} Unlike the epidermoid cyst, they have no

orifice. They present a firmer consistency and are more mobile than the epidermoid cyst.³

Several methods can successfully remove the cysts. The choice depends on the cyst's size, the quality of the overlying skin, the existence or not of the exit hole of the cyst, among other variables. However, they can relapse if there is incomplete removal of the capsule.⁶

We present a surgical marsupialization technique that allows safety in the total removal of the capsule, a technical variation not described in the literature.

METHODS (SURGICAL DESCRIPTION)

A cyst (about 1.5 cm in its largest diameter) in the scalp's midline, of elastic consistency, painless on palpation, not adhered to deep planes (Figure 1).

1. Marking of the incision on the line with the largest diameter of the cyst and its limits (Figure 1)
2. Local asepsis and antisepsis
3. Local anesthesia (lidocaine with vasoconstrictor), 2ml, in the incision, in the plane between the dermis and the anterior wall of the cyst (Figure 2)
4. Skin incision with a surgical scalpel blade n. 15 (Figure 3)
5. Incision in the cyst capsule with a surgical scalpel blade n. 15
6. Drainage of cyst content (Figure 4)
7. Deep anesthetic infiltration (1 ml) in the plane between the posterior wall of the cyst and the periosteum (Figure 5)
8. Expression of the cyst towards the skull (Figure 6)
9. Removal of the cyst using a Halsted clamp (Figure 7)
10. Suture



FIGURE 2: Anesthesia with local vasoconstrictor in the central incision

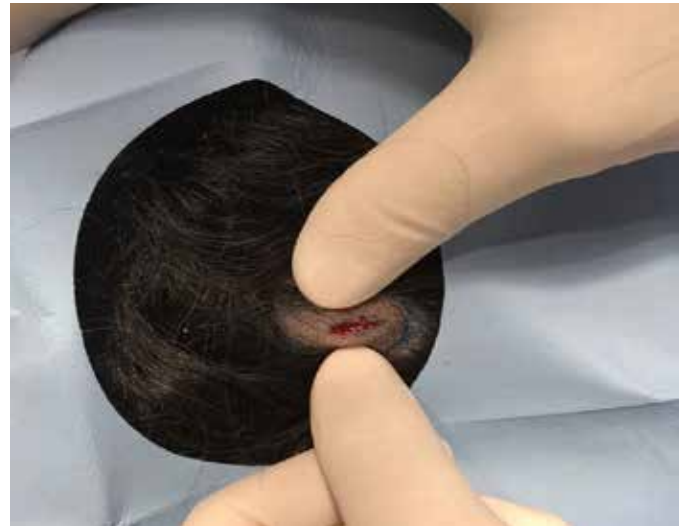


FIGURE 3: Central incision in the cyst



FIGURE 1: Marking of lesion boundaries and central incision site



FIGURE 4: Drainage of the cyst content

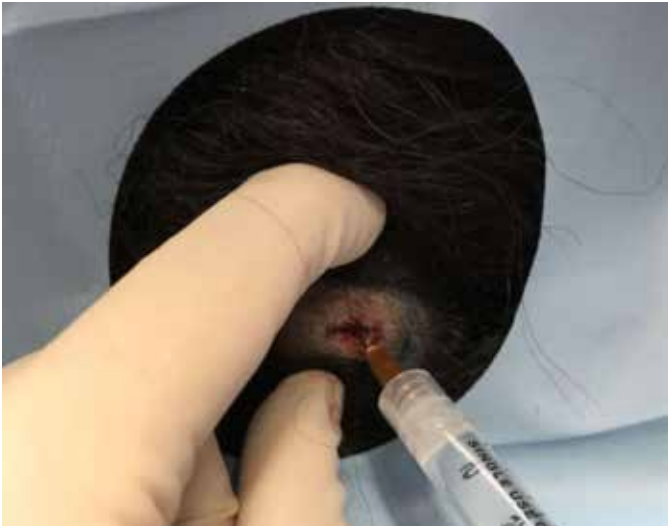


FIGURE 5: Lidocaine infiltration by the periosteum



FIGURE 7: Cyst removal, showing the total removal of the capsule



FIGURE 6: Compression of the pericyst region

DISCUSSION

Epidermoid or follicular cysts can have a central orifice and are often found in the dermis, hypodermis, and subcutaneous tissue. Although they are a very common and easily resolved clinical entity, they still depend on surgical management.³

The need for resections of the skin segment above the cyst depends a lot on the skin's quality since the cyst may have suffered inflammation and/or infections. Another fact requiring skin removal is the presence of the orifice that identifies the follicular ostium from which the cyst derives, which also must be removed.

Conventional removal is performed with infiltration anesthesia around and superficially to the anterior cyst wall, and it is one of the most used techniques in treating follicular cysts.⁴ It is performed through an incision on the skin surface, exposing the underlying cyst. The Halsted clamp is used to facilitate the detachment of the cyst's lateral and deep walls with delicately curved iris scissors until the total removal of the structure.^{1,6,7}

Although this technique is highly efficient, the possibility of rupture of the cyst in the tissues' dissection increases the probability of the permanence of capsule residues, potentiating a recurrence.

The technique presented performs the infiltration in the cyst's posterior region, which, with a two-finger compression maneuver, everts the cyst towards the incision when it is pulled. The inflammatory and infectious cyst recurrence processes can lead to the formation of a perilesional granuloma and reactive fibrosis, making it difficult to remove the capsule. In such cases, the presented technique's applicability may not be indicated since the existing fibrosis in the region would hinder the cyst's complete removal. Thus, removing the block structure, that is, skin and cyst, is recommended, which, although increasing the procedure's volume, guarantees the complete removal of the capsule.

The advantage of the presented technique lies in the fact that it is not necessary to separate the cyst's tissue and neighboring tissues, which, in turn, decreases bleeding and, consequently, the surgical time. This technique is simple, quick and applicable to places like the rigid scalp surface.³ ●

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Surgical correction of facial burn scar in ambulatory

Correção ambulatorial de cicatriz de queimadura

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ABSTRACT

Muscle, mucosae, and skin anatomically compose the lips. Neoplasms, congenital malformations, or local trauma can cause lesions on the lips. The surgical reconstruction of lip defects may need flaps, grafts, or microsurgery. The following case report describes the ambulatory surgical correction of a lower lip complex scar, using local skin flaps, with satisfactory aesthetic and functional outcomes.

Keywords: Ambulatory Surgical Procedures; Surgery, Plastic; Face; Surgical Flaps

RESUMO

Os lábios são compostos por estruturas musculares, mucosas e pele. As lesões em lábios podem ser de etiologia neoplásica, malformações congênitas ou traumáticas. A reconstrução cirúrgica dos defeitos nos lábios pode necessitar de técnicas de retalhos, enxertos e microcirurgia. Descreve-se no presente relato a correção cirúrgica ambulatorial do lábio inferior utilizando-se retalho cutâneo local, com evolução estético-funcional satisfatória.

Palavras-chave: Cicatriz; Cirurgia Plástica; Retalhos Cirúrgicos; Procedimentos Cirúrgicos Ambulatoriais

INTRODUCTION

The lips are aesthetic units of the face composed by muscle, skin and mucosae, which exert an important function related to the oral system. This important function is performed by controlling the orbicularis muscle of the mouth, where some fibers of this muscle are arranged horizontally, starting at one commissure and going towards the other, crossing the lip, making muscle-cutaneous insertions, columns and joints and these muscle fibers compress the lips. Besides the orbicularis muscle of the mouth, it is necessary to mention the elevator muscles of the upper lip, the zygomatic major muscle, and the elevators of the angle of the mouth. In addition to these, the depressor muscles of the lower lip, the mentalis, and the elevators of the central portion of the lower lip, compose the functions and aesthetic features of the lips¹. Lesions on the upper or lower lips can be of various etiolo-

gies, including trauma and neoplasms caused by sun exposure, especially in the lower lip. Depending on the type of lesion, the structures involved, its location and extent, there will be a need for a certain type of reconstruction, with the most varied options, from local, distant and microsurgical flaps.^{1,2} Considering the variables of the lesion and the flaps involved, we will have a proportional aesthetic-functional result⁴. In this report, we describe a case of lower lip reconstruction with local skin flap, performed in an outpatient setting, under local anesthesia, evolving with a good aesthetic-functional result.

CASE REPORT

A 56-year-old afro-descendant female patient, hypertensive and epileptic, arrived at the Dermatology Service of the Medical School of UNISA, complaining of the constant loss of saliva due to scarring of the lower lip and neck. The scar was caused during an epileptic seizure and consequent third-degree burn, 4 years earlier.

Upon physical examination, the patient had a scar on the right hemiface, on the lower third, which extended from the vermilion of the lower lip to the ipsilateral anterior cervical region.

In the transition regions between vermilion and lower lip and lower third of the face and neck, the scar caused the formation of flanges, in addition to the vitiligo-like areas secondary to the primary lesion (photo 1). At the static inspection of the lower lip, there was a small failure to close the upper and lower vermilion. Preoperative evaluation and release for surgery under local anesthesia was scheduled.



FIGURE 1: Pre-operative and surgery planning

Surgical procedure

The patient was placed in horizontal dorsal decubitus position; asepsis and antisepsis of the face with alcoholic chlorhexidine was performed, followed by bilateral mental nerve block and local anesthesia, with saline solution, Xylocaine and adrenaline (1/250000) in the amount of 10 cc, in the subdermal and subcutaneous planes of the lower lip. The first incision was made in the mucosal cutaneous line of the lower lip, bypassing the V-shaped defect, close to the right commissure and extending in a medial direction, extending 3 cm, and followed by the rotation of the flap of the mucosa (vermilion) on its own axis, to the right commissure and suture with 5-0 mononylon. Finally, it was made an advanced skin flap rotation, in a medial-medial direction and 5-0 mononylon suture.

Post-surgery period

The post-operative orientation was carried out, concerning the dressing change, the follow-up and the return for the removal of the stitches (photos 2 and 3).

Progress

Around the 14th post-surgery day, the cutaneous flap evolved with a small dehiscence in its most distal portion (photo 4), healing by second intention (photo 5). The last photographic documentation was done on the 28th post-surgery day. The patient was advised to maintain follow-up through the next 12 months, but that did not occur.

DISCUSSION

Performing the functional and aesthetic reconstruction of the lips is a challenging task for plastic surgeons and dermatologists^{2,3}. Surgical correction of these defects may lead to oral incompetence, sialorrhea and difficulty in speech³, depending on the flap used. Successful reconstruction seeks to preserve oral competence, maximum oral openness, speech and sensibility, as well as to improve aesthetics^{3,4}, and sometimes the achievement of these variables is partial.

The literature describes more than one hundred different reconstruction modalities³, but up to now, no technique is considered ideal for lip reconstruction⁵, since they involve many factors, such as type and size of the lesion. In defects comprising less than one third of the lip, the primary closure produces good functional and aesthetic result^{3,6}. Local flaps are a good option for defects that affect one to two thirds of the lip, as in the case reported^{3,6}. When the defect exceeds two-thirds of the lip, regions with previous reconstruction, or irradiated tissue, it is recommended microsurgical flap reconstruct³, among them, the antebrachial flap.

In reconstructions in general, especially in lip reconstructions, the donor area of the flap should be considered, as sometimes the use of the contralateral lip of the lesion is indicated. In addition, these muscular flaps include important structures for oral competence^{3,4}.

We emphasize that none of lesions required tissue removal compromising the perimeter of the mouth, but a repo-



FIGURE 2: Immediate post-op



FIGURE 4: Two weeks post-op. Small dehiscence area in healing stage



FIGURE 3: Seventh post-op



FIGURE 5: Four weeks post-op. Complete cicatrization and salivary continence obtained

sitioning of the ectopic tissues, showing an improvement of the oral competence, the movement, and the speech. Prolonged follow-up, up to 12 months, is recommended, since the development of retractions and thickening or keloids might be serotinous.

The local flaps used, both mucous and cutaneous, aimed at repositioning and to minimize the size of the procedure itself,

without compromising other structures and donor areas, since the clinical conditions of the patient allowed only local anesthesia. However, the occurrence of dehiscence may have been due to the quality of the skin of the flap, generating necrosis, since this skin was also involved in the primary trauma. Even with this complication, the evolution was very favorable in aesthetic and

functional terms, with the total resolution of the bridle between the vermilion and the lip and the repositioning of the inferior vermilion, leading to total oral competence.

CONCLUSION

In the case reported, mucosal and cutaneous flaps were performed under submental block and local anesthesia, which resulted in satisfactory oral competence and good aesthetic result. ●

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Atypical presentation of dermatofibrosarcoma protuberans: Case report

Dermatofibrossarcoma protuberans de apresentação clínica atípica: relato de um caso

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ABSTRACT

Introduction: Dermatofibrosarcoma protuberans (DFSP) is a local aggressive sarcoma that presents 60% of recurrences. Rarely it presents lung metastasis too.

Methods: 41 years old man presented a soft tumor in his right hallux for two years. Histopathology has shown spindle-shaped cells arranged in a storiform manner, and immunohistochemistry was CD34 positive.

Discussion: The literature describes some clinical types of DFSP. The most common are: confluent nodules forming a plaque, sometimes similar to keloids, tumoral lesions, and atrophic plaques.

Conclusion: DFSP cases simulating a subcutaneous cyst have been described in the literature; nevertheless, it is an unusual presentation of this tumor.

Keywords: Dermatofibrosarcoma; Sarcoma; Surgical Procedures, Minor

RESUMO

Introdução: O dermatofibrossarcoma protuberans (DFSP) é um sarcoma localmente agressivo que apresenta recidiva local em até 60% dos casos e raras metástases pulmonares.

Métodos: Paciente masculino, de 41 anos, apresenta tumoração amolecida à palpação, localizada no hálux direito há dois anos. Histopatologia com células fusiformes em arranjo estoriforme. A imuno-histoquímica foi focalmente positiva para o CD34.

Discussão: Algumas variantes clínicas são descritas. As principais são: lesões nodulares confluentes formando uma placa, muitas vezes com aspecto clínico semelhante ao de quelóide; lesão tumoral; placa atrófica.

Conclusão: Casos de DFSP simulando cisto subcutâneo foram encontrados na literatura, porém trata-se de uma apresentação clínica inusitada de DFSP.

Palavras-chave: Dermatofibrossarcoma; Procedimentos Cirúrgicos Menores; Sarcoma

INTRODUCTION

Described previously by Taylor in 1890, dermatofibrosarcoma protuberans (DFSP) is a locally aggressive sarcoma that presents local recurrence in 60% of the cases besides rare metastases, which are also available to lymph nodes.^{1,2,3}

DFSP is a rare and infiltrating connective tissue tumor that represents 1–2% of all sarcomas. DFSP has a prevalence of 0.8 to 5.0 cases per million population per year¹ and occurs primarily among youth in the third and fifth decades of life age, although it may occur to people at any age.^{3,4} Due to the slow and asymptomatic growth of DFSP, the cases diagnosed in adulthood begin in childhood and approximately 20% occurred in children. A study observed that the occurrence of DFSP in black-skinned patients happens two times more in relation to white-skinned ones, while a gender distribution among them is the same.^{4,5}

The trunk is the most affected site (50–60% of the cases), mainly the thorax and shoulders, followed by the proximal region of the limbs (20–30%), being more common in the upper limbs.^{4,6} Approximately 10 to 15% of cases occur in the head and neck, mainly in the scalp and malar. In children, a tendency to acral presentation of the tumor has been described.⁷ Hands and feet are affected in about 1% of the cases.^{8,9}

Approximately 85–90% are low-grade lesions whose behavior is an indolent tumor with an average evolution of about 6.4 years at the time of diagnosis.^{1,3} Occasionally, some areas of differentiation for high-grade sarcoma within the DFSP are found. These cases present a more aggressive behavior and have high rates of recurrence and metastases.^{3,10}

Metastasis are rare and occur in 2–5% of cases. Therefore, CT scans and laboratory tests are not routinely performed. Metastasis can spread hematogenously, usually in more advanced lesions or recurrent lesions, and chest X-rays should be requested for all patients and chest tomography only for those with pulmonary metastasis.^{4,8,9}

The mean size of the DFSP varies from 2 to 5 cm. In cases with late treatment, some lesions may reach 20cm in diameter and present multiple satellite nodules. It is usually restricted to the skin. However, recurrent or old lesions may invade other structures, such as fascia, striated muscle, periosteum, and bones.⁴

Some reports associate the emergence of DFSP with the HIV virus infection, renal transplantation and antibody deficiency.¹¹ A previous history of trauma is described in about 10–20% of cases, being considered a possible etiologic agent.⁹

In 1993, the immunoreactivity of this tumor was described for CD34 that today continues as its main immunohistochemical marker, especially if associated with negativity for XIIIa factor.¹²

Even after extensive resections, there may be local recurrences due to the presence of unexpectedly positive margins, secondary to the microscopic infiltrative pattern of tumor growth. In these cases, a new excision accompanied by a careful histopathological analysis is indicated.^{3,8,9} The mean time to recurrence is about 2–3 years after excision.^{3,13,14} We report hitherto the case of a patient with DFSP of unusual location and atypical clinical presentation.

METHODS

A 41-year-old male presented a softened tumor in the right hallux with growth for two years (Figure 1 and 2). Pain, dyspnea or other symptoms were reported. Chest radiography without alterations.

A right-sided CT scan revealed a nodular formation with a density of soft tissues and contrast medium of 1.8 x 1.5cm in the subcutaneous tissue near the interphalangeal joint of the first pododactyl. It also presented discrete erosion of the cortical bone in the epiphysis of the proximal phalanx in contiguity to injury. The patient was referred to the dermatologic surgery for lesion excision. It was performed the hallux garrotrum and fusiform incision above the lesion (Figure 3), with tumor removal and approach of the borders by simple suture with 4-0 wire (Figure 4).

During the surgical procedure, a softened lesion was confirmed, simulating cystic lesions. The histopathological appearance revealed spindle cells in a stent-like arrangement which, upon infiltration of the subcutaneous tissue, shows a honeycomb appearance and a focally positive immunohistochemistry for CD34 (Figure 5). In light of the data presented, the diagnosis of DFSP with positive surgical margins was confirmed. Margin enlargement with freezing was performed and the patient was sent for a radiotherapy procedure.

DISCUSSION

The progression of DFSP is slowly over a long period of time until it enters a rapidly growing phase.¹⁵ Initially, DFSP appears as a hardened, purplish or brownish plaque, attached to the skin, but not to the subcutaneous. After a period that may vary from years to decades, it occurs the proliferation of multiple



FIGURE 1: Clinical presentation



FIGURE 2: Clinical presentation-medial view



FIGURE 3: Tumor excision



FIGURE 4: Post operative

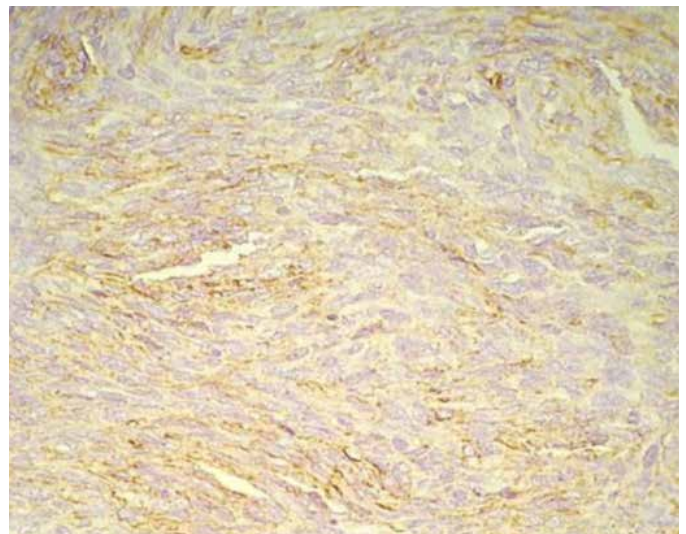
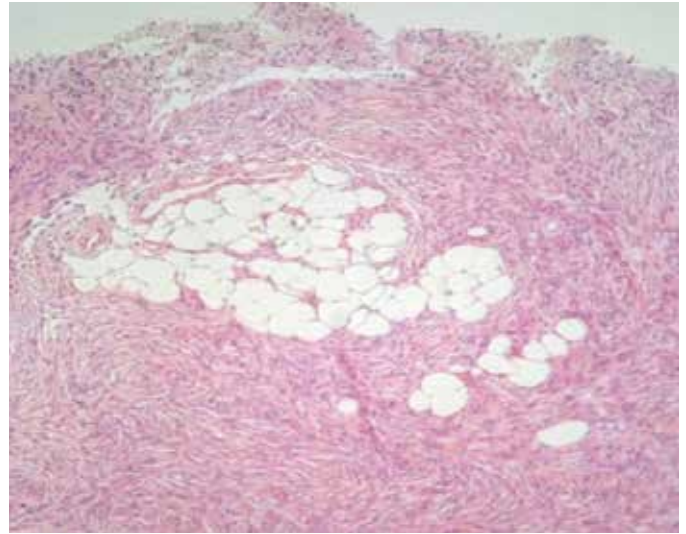


FIGURE 5: On the left side, the histopathological appearance revealed spindle cells in a stent-like arrangement which, upon infiltration of the subcutaneous tissue, shows a honeycomb appearance (hematoxylin and eosin, 100x). On the right side, focally positive immunohistochemistry for CD34

nodules on the plaque, justifying such a name as protuberans. The DFSP can also appear as a single cutaneous nodule, but it is an unusual clinical presentation.^{4,8} In the course for the tumor phase or when it already appears in this phase, some differential diagnoses must be remembered, such as epidermal cysts, lipomas or dermatofibromas.⁴

Some clinical variants are described in the literature, as long as the main ones are: a) confluent nodular lesions forming a plaque, often similar to the keloid one b) tumor lesion c) atrophic plaque. In addition, simulating subcutaneous cyst cases were found in the literature, with similar clinical characteristics to those presented by the current patient.^{7,16,17}

Cystic presentation is unusual.^{18,19} Gielli et al. presented a series of 27 cases of cystic DFSP observed in a hospital in Italy during a period of 14 years.¹⁸ Shivartsbeyn et al. showed by molecular methods that the cell line that formed the pseudocystic

part of the DFSP was the same as the main tumor line, allowing the confirmation of the diagnosis and emphasizing that the cyst region was of neoplastic origin and did not represent a secondary reactive proliferation.¹⁹

The treatment pillar is performed with complete surgical removal with free margins under light microscope. Two centimeters margins have been suggested in the literature and margins of 5 cm may reach less than 5% recurrence. However, morbidity is proportional to tumor size, which can lead to an increase in complications resulting from the procedure, such as bleeding and surgical wound infections.^{4,8,20} Furthermore, the surgical procedure may require complex reconstructive techniques and cause functional or cosmetic loss.

After surgery, patients should be followed up every six months for the first three years and annually after that period. Inspection and palpation of the surgical scar are essential.²¹

Adjuvant radiotherapy is indicated in cases with positive margin when further surgery is not possible. Imatinib (PDGF receptor inhibitor) has been used as the first safe and effective drug in DFSP systemic therapy. Lesions initially assessed as unresectable tumors with metastasis in initial presentation or indication of mutilating surgeries are examples in which the use of such a drug assists in the reduction and subsequent excision of the tumor.³

CONCLUSION

The cystic aspect is more common in larger tumors. The cause of this cystic transformation is unknown and represent a degenerative phenomenon rather than a histopathological variant.¹⁹ However, in the aforementioned patient, the tumor presented reduced size.

Cystic recurrence is rarely observed in sarcomas in general and should be considered as a pseudocystic modification, taking into account that sarcomas, because they are originated from the mesenchymal lineage, would not have true epithelial tissue to form the wall of a cyst. Other sarcomas that may present with formation are endometrial stromal sarcoma, pulmonary synovial sarcoma, and fibroblast sarcoma of the small intestine.¹⁹

In high-grade tumors, the cystic portion can be related to degeneration, due to areas of ischemia or necrosis. However, in the aforementioned presented case, the tumor did not have these characteristics as justification of its morphology. In light of that, it is vitally important to emphasize the recognition of this form of DFSP to avoid its misinterpretation as a benign cystic lesion. ●

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Giant basal cell carcinoma of the scalp: report of late reconstruction

Carcinoma basocelular gigante em couro cabeludo: relato de reconstrução tardia

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ABSTRACT

The giant BCC is a rare variant, and the repair of large defects in the scalp, generated after oncologic resection, is a surgical challenge. We report the case of a patient submitted to late scalp reconstruction after oncologic resection of giant BCC in the scalp with the association of techniques. Reconstruction of scalp lesions depends on a comprehensive understanding of the anatomy of this site, knowledge of the arsenal of surgical techniques, and a detailed assessment of patient factors and expectations, given the impact on the quality of life and morbidity related to each therapeutic option.

Keywords: Carcinoma, Basal Cell; Reconstructive Surgical Procedures; Scalp Dermatoses

RESUMO

O carcinoma basocelular (CBC) gigante é uma variante rara, sendo o reparo de grandes defeitos no couro cabeludo, gerados após ressecção oncológica, um desafio cirúrgico. Relata-se o caso de um paciente submetido à reconstrução tardia do couro cabeludo após ressecção oncológica de CBC gigante no couro cabeludo, com associação de técnicas. A reconstrução das lesões do couro cabeludo depende de uma compreensão abrangente da anatomia desse local, um conhecimento do arsenal de técnicas cirúrgicas e uma avaliação detalhada dos fatores e expectativas do paciente, tendo em vista o impacto na qualidade de vida e a morbidade relacionada a cada opção terapêutica.

Palavras-chave: Carcinoma Basocelular; Dermatoses do Couro Cabeludo; Procedimentos Cirúrgicos Reconstructivos

INTRODUCTION

Basal cell carcinoma (BCC) is the most common cancer in light-skinned populations, accounting for more than 75% of non-melanoma skin cancers. It occurs in the head and neck region in approximately 80% of cases. Giant BCC (GBCC), defined as a lesion larger than 5 cm in its largest diameter, is a rare variant of BCC, with an incidence of 0.5%.¹⁻³ In contrast to the small lesion, the giant variant develops on non-exposed skin, including the back, shoulders, groins, and thighs. Scalp involvement is extremely rare. The lesion can invade the underlying deep tissue, compromising treatment options and challenging reconstruction methods.

Many therapeutic modalities have already been reported as surgical options, with varying results. The main ones include surgical excision and reconstruction, radiotherapy, and Mohs micrographic surgery, the latter being the gold standard treatment for giant BCC.^{1,2}

Case report

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Scalp reconstruction after ablative surgery can be challenging. Physicians must evaluate each case individually. The best proposal for reconstruction must be based on the patient's health, psychosocial aspect, and the therapeutic planning of the underlying disease, without compromising the oncological surgery principles.^{4,5}

This study aims to report the case of a patient who underwent late scalp reconstruction after oncological resection of giant basal cell carcinoma on the scalp, with the association of techniques.

CASE REPORT

A 74-year-old woman, retired, living in a rural area, presented a papular-nodular lesion on the scalp eight years ago. The lesion had slow and progressive growth, with central healing, bleeding, and local itching.

Upon dermatological examination, we observed a scalp with an extensive plaque of scarring alopecia, presenting an atrophic center, with exulcerations and crusts, periphery with papulous, pigmented, lacy-looking edges, measuring 17 x 17 cm, located in the right frontotemporo-parietal region. (Figure 1). Dermoscopy showed the presence of large, unfocused blue-gray cells, with edges tending to form rounded structures and tortuous vessels with arboriform ramifications (Figure 2). Anatomopathology was compatible with pigmented basal cell carcinoma.

We performed a scalp lesion excision, respecting oncological surgical margins, aided by dermoscopy. Healing by secondary intention was chosen with late reapproach (Figures 3 and 4).

After four months, partial skin grafting of the lateral aspect of the left thigh was performed on the recipient area's granulation tissue, using a dermatome.

The patient evolved with good response to the graft, presenting necrosis of approximately 10% of its extension and subsequent total healing (Figure 5).

DISCUSSION

The repair of large scalp defects generated after cancer resection is a surgical challenge. Modern surgery techniques have provided a greater capacity for resolving lesions and as a reduction in perioperative and postoperative complications, and a better aesthetic and functional result.⁶

Size, location, local tissue's quality, and scalp distortion help choose the best reconstructive method. Practitioners must also consider the patient's intrinsic and psychosocial factors, given the impact on their quality of life.^{4,6}

Healing by secondary intention can be used when the pericranial layer with extensive exposure is present, and when a shorter surgical time is required. The disadvantages of this method include delayed healing, alopecia, telangiectasias, and thin coverage.⁴

The partial skin graft has been used in large defects due to the technique's speed and ease, the possibility of monitoring tumor recurrence, and fast healing.⁷ It should be used when aesthetic issues are not a concern, as they can lead to alopecia,



FIGURE 1: Patient at the time of diagnosis with extensive plaque of scarring alopecia, periphery with papulous, pigmented edges



FIGURE 2: Dermoscopy with the presence of large defocused blue-gray cells, tortuous vessels with arboriform branching



FIGURE 3: Patient in the preoperative period with lesion measuring 17 x 17cm



FIGURE 5: Patient after three months of partial skin graft in the second surgical procedure



FIGURE 4: Patient in the immediate postoperative period after resection of the lesion with healing by secondary intention

hypopigmentation, contour deformity, and morbidity of the donor site.⁴

A retrospective study assessed 65 patients who underwent skin grafts on the scalp, with grafts from various sites, including six thigh flaps. This study reported a low rate of surgery complications and satisfactory and long-lasting cosmetic results. It recommended using anterolateral thigh (ALT) flaps for surgeries in the scalp's temporal-lateral region.⁸

As in the case reported, the association of healing by secondary intention techniques and partial skin graft can be used. The formation of granulation tissue is related to greater adherence of the skin graft due to greater vascularization, enabling better results.⁴

CONCLUSION

The reconstruction of scalp lesions depends on a comprehensive understanding of the scalp's anatomy, knowledge of the surgical techniques' arsenal, and a detailed assessment of the patient's factors and expectations. The reconstructive goal must be adapted for each patient individually, aiming at the success of the reconstruction and a satisfactory aesthetic-functional result. ●

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Dermatofibrosarcoma protuberans simulating keloid

Dermatofibrossarcoma protuberante mimetizando queleide

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ABSTRACT

Dermatofibrosarcoma protuberans is a locally aggressive sarcoma of intermediate malignancy that predominates in the range of young adults to middle age. Congenital or childhood lesions are rare but have been reported. It can be misdiagnosed and incompletely excised in the early stages, which increases the risk of recurrence. The present article reports the case of a 24-year-old male patient presenting a hyperchromic macula in the abdominal region from birth that evolved into a tumor after several surgical approaches. Biopsy and immunohistochemistry confirmed the diagnosis of dermatofibrosarcoma protuberans, and the lesion was excised with a 3 cm margin.

Keywords: Dermatofibrosarcoma; Neoplasms, Fibrous Tissue; Sarcoma

RESUMO

O dermatofibrossarcoma protuberante é um sarcoma localmente agressivo, de malignidade intermediária, que predomina na faixa de adultos jovens à meia-idade. Lesões congênitas ou na infância são raras, mas foram relatadas. Nos estágios precoces, pode ser mal diagnosticado e ser incompletamente excisado, o que aumenta o risco de recorrências. Relata-se o caso de paciente do sexo masculino, de 24 anos, com história de mácula hipercrômica em região abdominal desde o nascimento que evoluiu para tumoração após duas abordagens cirúrgicas. Biópsia e imuno-histoquímica confirmaram o diagnóstico de dermatofibrossarcoma protuberante, e a lesão foi excisada com margem de 3cm.

Palavras-chave: Dermatofibrossarcoma; Neoplasias de Tecido Fibroso; Sarcoma

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a fibrohistiocytic tumor of intermediate malignancy, which, although rare, represents the majority of cutaneous sarcomas. It is locally aggressive, with high rates of recurrence but low metastatic potential. DFSP occurs more in the age group of 20 to 50 years and has a slight predominance in males.^{1,2,6}

The most frequent location is the trunk (40–60%), followed by the limbs (20–30%) and the head and neck (10–15%).³ It is exceptionally described on the abdominal wall, as it represents the soft tissue tumor, less frequent in this region.⁴

In its initial phase, DFSP presents as a hardened plaque, asymptomatic, skin color, violet, reddish-brown, or hyperchro-

Case report

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mic, similar to keloid. Eventually, DFSP develops reddish-brown nodules up to several centimeters in diameter. In palpation, the lesion is solid and adhered to the subcutaneous tissue. There are reports of lesions that develop after trauma or in previous surgical scar.^{1,3,7}

In histopathological examination, DFSP presents a uniform, dense collection of spindle cells arranged as short fascicles in a “storiform” or mat-like arrangement with low nuclear pleomorphism and low mitotic activity. The cells infiltrate the subcutaneous tissue forming a “honeycomb” pattern. Deep cellular projections toward the fascia and muscle hinder delimitation and surgical removal of the DFSP.^{3,5,6}

The immunohistochemistry test of DFSP shows fusiform cells positive for CD34 and factor XIIIa negative, which helps in the differential diagnosis with dermatofibroma, in which cells are CD34 negative and factor XIIIa positive.^{2,7,8}

Complete surgical excision, including Mohs micrographic surgery, is the standard treatment for DFSP. Imatinib mesylate has been tried in patients with unresectable, recurrent, and/or metastatic DFSP.²

The aim of this study is to highlight the importance of early diagnosis of DFSP, which, although rare, can clinically simulate various cutaneous lesions, such as keloids. The first surgical approach with broad margins has a greater curative chance, reducing risk for local recurrences and high morbidity.

CASE REPORT

A 24-year-old male patient from Belo Horizonte, Brazil, with lesion in the abdominal epigastric region from birth, which began as a hyperchromic macula accompanied by pain at the site.

A biopsy was performed at 12 years of age, suggestive of keloid according to the patient, who did not present the anatomopathological result at the medical appointment. The patient underwent the surgical approach at 12 and 16 years of age, with posterior recurrence and lesion growth, which progressed to nodules and skin color plaques. He also did not have the histological results of these surgeries.

At age 24, he was submitted to a new biopsy requested by a dermatologist, which was suggestive of fibromixoma without signs of malignancy. The patient was referred to the Dermatology service of the Santa Casa de Belo Horizonte for propaedeutic extension. Physical examination showed a lobate erythematous-violet tumor with fibrous consistency of approximately 8 x 6 cm in the abdominal epigastric region (Images 1 and 2).

According to the clinical presentation, the main suspicion was DFSP and, therefore, a revision of the anatomopathological slide was performed. The review revealed spindle cells in a storiform arrangement and the material was submitted to immunohistochemical evaluation (Image 3). Immunohistochemistry revealed positivity for the CD34 antibody, being conclusive of DFSP, corroborating with the main clinical diagnostic hypothesis.

In addition, the patient was submitted to a nuclear magnetic resonance, to evaluate the extent of the lesion and local invasion. The examination showed multiloculated expansive le-



FIGURE 1: Erythematous-violaceous lobulated tumor of fibrous consistency of approximately 8 x 6 cm in the abdominal epigastric region



FIGURE 2: Erythematous-violaceous lobulated tumor in the abdominal epigastric region

sions in the skin and subcutaneous and a hepatic nodule and may correspond to secondary neoplastic involvement. However, the nodule propaedeutics did not show, until now, any relation with the cutaneous neoplasia.

The lesion was excised by the plastic surgery team, with a margin of three centimeters, with removal of the fascia of the rectus abdominis muscle and wall closure by reverse abdominoplasty, with good aesthetic results (Images 4 and 5). As discussed with a pathologist, the anatomopathological of the surgical

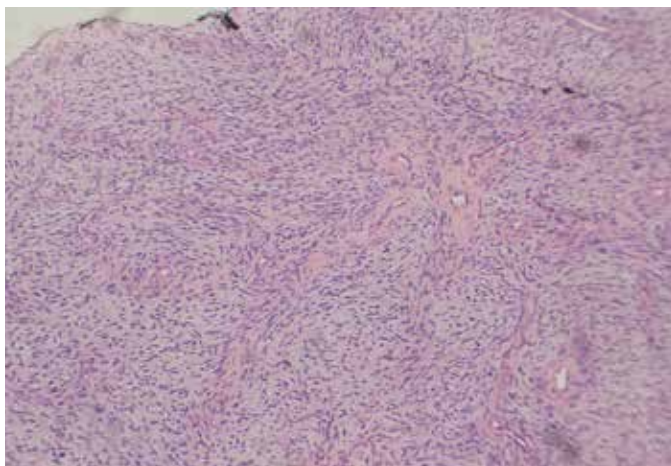


FIGURE 3: Anatomopathological showing spindle cells in a stent-like arrangement



FIGURE 4: Preoperative marking

specimen was compatible with DFSP with fragments of muscle fascia free from neoplasia and the possibility of more aggressive transformation in the part analyzed was excluded.

No adjuvant treatment was given. At the moment, one year after the surgical procedure, the patient has no evidence of local or distant recurrences.

DISCUSSION

The DFSP is a neoplasm of spindle cells with intermediate malignancy and low risk of metastasis. However, the indolent behavior of the tumor and its inaccurate characteristics often lead to delays in its perception by patients and physicians, leading to delays in diagnosis. Moreover, due to the higher prevalence of cases of basal cell carcinomas, squamous cell carcinomas and cutaneous melanomas, dermatologists are more familiar with the diagnosis, prognosis and treatment of these neoplasms.^{1,3}

The early stage of DFSP can be misdiagnosed as a benign tumor and incompletely excised because it is a nonspecific and asymptomatic lesion. The clinical differential diagnosis includes keloid, dermatofibroma, dermatomyofibroma and morphea.^{2,7}

Proper collection of material for anatomopathological (AP) is extremely important because superficial samples of material for AP may not produce diagnostic material.²

The first intervention is of extreme importance, since tumor spread after the first inadequate resection may lead to uncontrolled local growth or metastasis.¹

The high rates of local recurrence accompanied by the high morbidity of DFSP justify the effort in the early diagnosis and the need for initial treatment, which allows a higher rate of cure.^{3,7,9}



FIGURE 5: Removal of the surgical specimen and immediate postoperative period

Lesions with multiple recurrences after initial treatment appear to be more likely to differentiate into high-grade malignancies with increased risk of metastases.³

The main prognostic factors are inadequate surgical removal with compromised margins, large tumors located in the head and presence of fibrosarcoma areas in histology.³

The treatment with greater curative chances is surgical resection with wide local excision. The microscopic spread of

the tumor by projections of tumor cells, similar to tentacles, under clinically normal skin, makes complete surgical removal of the lesion difficult. When possible, Mohs micrographic surgery

has the highest cure rates for this tumor.^{1,3} In this case, we opted for conventional surgery with wide surgical margins made by plastic surgery due to the large extent of the lesion. •

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Dowling-Degos Disease - Genetic and spectral disorder of reticular hyperpigmentation

Doença de Dowling-Degos: desordem genética e espectral de hiperpigmentação reticular

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ABSTRACT

Dowling-Degos disease (DDD) is a rare genodermatosis. Progressive reticular hyperpigmentation of flexural areas mainly characterizes the disease, which may be associated with a large spectrum of benign lesions and cutaneous neoplasms. It may cause psychosocial impairment due to the deformity caused by the lesions with significant aesthetic damage. We present two cases of male patients with hyperchromic macules, multiple comedones, epidermal cysts, cribriform scars on the face, cervical region, dorsum, anterior thorax, armpits, genital region, and disfiguring tumors, characteristic of DDD.

Keywords: Carcinoma; Genetics; Hidradenitis; Hidradenitis Suppurativa; Skin Pigmentation

RESUMO

A doença de Dowling-Degos (DDD) é uma genodermatose rara, caracterizada principalmente por hiperpigmentação reticular progressiva de áreas flexurais, que pode estar associada a um grande espectro de lesões benignas e neoplasias cutâneas. Pode ocasionar prejuízo psicossocial devido ao potencial de deformidade das lesões e dano estético significativo. Apresentamos dois casos de pacientes, ambos do sexo masculino, com máculas hiperocrômicas, múltiplos comedões, cistos epidérmicos, cicatrizes cribriformes em: face, região cervical, dorso, tórax anterior, axilas e região genital, além de tumorações desfigurantes, características da DDD.

Palavras-chave: Carcinoma; Dermatopatias; Genética; Hidradenite; Hidradenite Supurativa; Pigmentação da Pele; Síndrome

INTRODUCTION

Dowling-Degos disease (DDD) is a rare genodermatosis, with an autosomal dominant transmission, variable penetrance, and expressiveness, initially described by Jones and Grice in 1974.^{1,2}

It is a late-onset disease, after the second decade of life. Reticulated dyschromia, mainly on the face and flexural surfaces such as neck, armpits, elbow pit, submammary areas, and groins, characterize the condition.^{3,4} Concomitantly, lesions of the pilosebaceous unit may occur, such as comedones, epidermal cysts, abscesses, hidradenitis suppurativa, in addition to skin neoplasms, such as squamous cell carcinoma and keratoacanthoma.^{2,4} Pigmentation is progressive and symmetrical, usually extensive and asymptomatic, exacerbated by sun exposure.^{5,6,7}

Case report

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Differential diagnoses should be made with acanthosis nigricans, acropigmentation of Kitamura, Galli-Galli disease, dyschromatosis universalis hereditaria, and dyschromatosis symmetrica hereditaria.⁸ Such conditions may present clinical overlaps with each other, and some authors can consider them as different diseases, while others, the spectrum of the same disease.^{4,5,9}

The diagnosis is based on suggestive clinical characteristics associated with the findings on histopathological examination.^{7,8} Histopathology shows hyperpigmentation of the basal layer, filiform proliferation of the epidermis, sometimes similar to reindeer horn, in addition to hyperkeratosis and budding, arising from the hair infundibulum, featuring a follicular plug.^{1,3,6} Perivascular lymphohistiocytic infiltrate in the papillary dermis and horny pseudocysts can be observed, with a standard number of melanocytes.^{7,10} Genetic tests reveal mutations in keratin 5 (KRT5), protein O-glycosyltransferase 1 (POGLUT1), protein O-fucosyltransferase 1 (POFUT1), and PSENEN gene as causing the disease. Patients with the latter mutation present lesions of hidradenitis suppurativa.^{2,4}

There is no definitive treatment for DDD.⁷ The treatments are unsatisfactory, and topical hydroquinone, tretinoin, adapalene, and corticosteroids can be used, in addition to Er:YAG laser, aiming to reduce the risk of post-inflammatory hyperpigmentation.^{1,7} Isotretinoin would be an option since the condition promotes a change in keratinization.¹ Cystic and tumoral lesions should be treated with surgical excision.⁷

METHODS

We conducted a retrospective descriptive study of two cases diagnosed with Dowling–Degos disease at the Dermatology Clinic from November 2016 to May 2019.

CASE REPORT

Case 1

A 58-year-old man reported hyperpigmentation on the face, cervical region, and flexural areas for ten years, associated with the progressive appearance of multiple cysts, which drained purulent exudate, and a tumor on the buttock and lateral aspect of the right thigh. On clinical examination, the patient had predominant hyperpigmentation on the face and flexural areas, multiple open comedones, and epidermoid cysts distributed throughout the body and on the hips and buttocks, hidradenitis suppurativa-like lesions, and a keloid-like tumor (Figure 1). He denied similar cases in the family.

Case 2

A 59-year-old man presented suppurative lesions in the gluteal region associated with progressive growth tumor for one year. On clinical examination, he showed dermatosis characterized by multiple comedones spread throughout the body, associated with hidradenitis suppurativa-like lesions in the left buttocks, and vegetative and ulcerated tumor in the perianal and intergluteal region, whose biopsy diagnosed invasive squamous cell carcinoma (Figure 2). He denied similar cases in the family.



FIGURE 1: Patient presenting facial reticulated hyperchromia (A), associated with multiple open comedones and cystic lesions on the back (B). In the lateral region of the gluteus and right hip, there is a large, hardened tumor mass associated with fistulized lesions with purulent secretion (C)

DISCUSSION

DDD is a late-onset genodermatosis, usually in adulthood, which initially affects the armpits and groins, and later, the intergluteal and inframammary regions, neck, and trunk.¹ Both patients presented the condition in adulthood, and the lesions clearly show the preference for these locations. Although this disease predominantly affects women in a proportion of 2:1,¹ we present two male patients.



FIGURE 2: Patient presenting facial reticulated hyperchromia (A), associated with multiple open comedones and cystic lesions on the back (B). In the lateral region of the gluteus and right hip, there is a large, hardened tumor mass associated with fistulized lesions with purulent secretion (C)

The patients reported here presented not only the dyschromic manifestations of the syndrome, but also several alterations in the pilosebaceous unit spread throughout the body, from open comedones to hidradenitis, with a high impact on the quality of life, in general, and on self-esteem, in particular, including significant aesthetic changes.

The exuberance of the lesions in this study demonstrates why this disease is also called the “dark dot disease” (DDD) by some authors.¹ Despite not having inflammatory phenomena in their benign evolution, these lesions are deeply unsightly. In the

second patient, we also observed intergluteal squamous cell carcinoma, a neoplasm reported in association with this disease.^{2,4} In addition to both cases’ clinical aspect characteristics, a histopathological examination confirmed the diagnoses, which was compatible with the findings described in the literature.

Finally, it is essential to note that the present study was conducted at the Dermatology Clinic of a tertiary reference hospital in the country’s largest city. In three years of care, only two cases of DDD were evidenced, showing the rarity of this dermatosis in the general population. ●

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Nodular fasciitis in the forehead: a rare presentation

Fasciite nodular na fronte: uma rara apresentação

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ABSTRACT

Nodular fasciitis is a benign tumor resulting from the reactive proliferation of fibroblastic or myofibroblastic cells presenting rapid growth and rich cellularity. In adults, the extremities' involvement is more frequent; however, other regions can be affected. In this report, we present the case of a 40-year-old woman with a nodular lesion on the forehead. The histopathology confirmed the diagnosis of nodular fasciitis. The reported case highlights its rare location and alerts the clinical dermatologist in its inclusion among the differential diagnoses of tumor lesions on the face.

Keywords: Facial Dermatoses; Fasciitis; Forehead

RESUMO

A fasciite nodular é um tumor benigno, decorrente da proliferação reativa de células fibroblásticas ou miofibroblásticas de rápido crescimento e rica celularidade. Em adultos, o acometimento das extremidades é mais frequente; entretanto, outras regiões podem ser acometidas. Neste relato, é apresentada paciente feminina de 40 anos, com lesão nodular na fronte, com diagnóstico de fasciite nodular confirmado à histopatologia. O caso relatado procura destacar sua rara localização e alertar o dermatologista clínico para a sua inclusão entre os diagnósticos diferenciais das lesões tumorais na face.

Palavras-chave: Fasciite; Face; Testa

INTRODUCTION

Nodular fasciitis (FN) is a benign tumor derived from the proliferation of fibroblasts and myofibroblasts that can affect hypodermis, muscle, and fascia. In 1955, Konwaler described the FN as pseudosarcomatous fasciitis or infiltrative fasciitis, because of its strong resemblance to sarcoma.¹ The etiology of the FN is still uncertain, but there are reports of previous trauma in about 10 to 15% of cases.² Clinically, the lesion presents as a solitary, fast-growing nodule measuring 1 to 5 cm and more frequently affecting the extremities of the upper limbs. Some presentations can be confused with sarcoma due to its rapid growth, rich cellularity, and high mitotic activity, being of fundamental importance the distinction of the FN with malignant neoplastic diseases.

Case report

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CASE REPORT

A 40-year-old hypertensive female patient with a history of onset of tumor in the forehead for 4 years, with progressive increase and mild local pain. On physical examination, the tumor of the patient had a 2 cm diameter, mobile, fibroelastic consistency, well-delimited, and not adhered to deep planes on the left forehead (Figure 1). With the diagnostic hypotheses of schwannoma, lipoma, and epidermoid cyst, and due to unavailability of ultrasonography exam (USG), fine needle aspiration (FNA) was performed, which showed fusocellular mesenchymal proliferative lesion suggestive of FN (Figure 2). We chose to perform surgical excision, with the removal of a 2cm-diameter tumor, with a light yellow color, well-delimited and parenchymal consistency (Figure 3). In the histopathological examination, the presence of mesenchymal proliferation with discrete atypias was noted (Figure 4), which, associated with FNAB and clinical presentation, confirmed the diagnosis of FN in the absence of immunohistochemistry.

DISCUSSION

NF is a benign fibroproliferative disease of unknown etiology that affects men and women between the ages of 20 and 40. Reports of spontaneous remission and frequent location on bony prominences suggest the etiological hypothesis of previous trauma at the site of lesion.² In adults, the extremity involvement



FIGURE 1: Movable nodule, about 2 cm in the left forehead, without involvement of the suprajacent skin

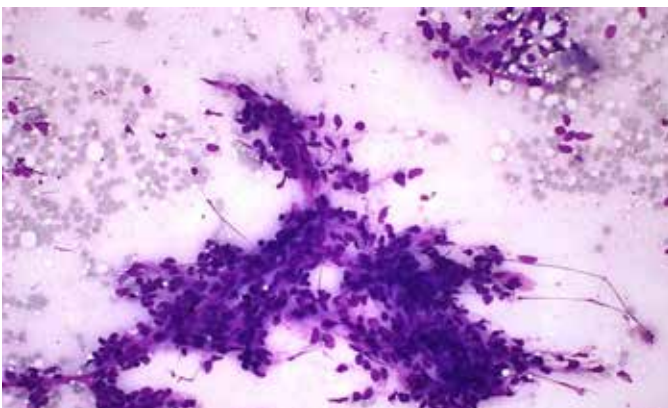


FIGURE 2: PAAF: presence of agglomerated fusel cell mesenchymal cells



FIGURE 3: Complete excision of the lesion, with removal of a pale yellow nodule, about 1.5 cm and parenchymal appearance on palpation

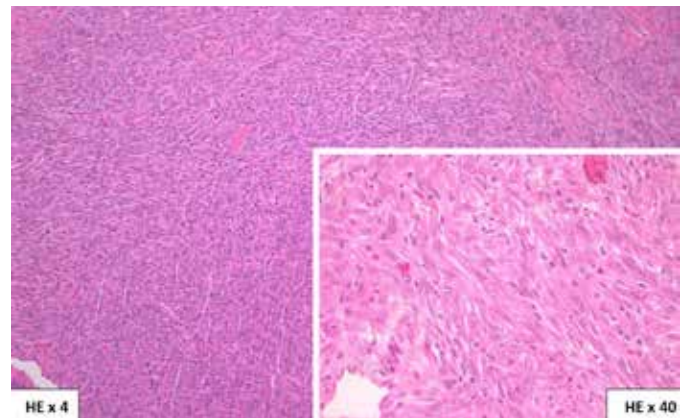


Figure 4: Histopathology: presence of mesenchymal proliferation with discrete atypia

is more frequent in the upper extremities (43%), followed by the trunk (25%) and lower extremities (22%), while only 10% of the FN occurs in the face and neck.^{3,4} Most cases of NF in the face and neck occur in children.⁵

Clinically, FN manifests as a tumor lesion, well delimited, about 2 to 5 cm in size, with subcutaneous nodular growth fast and self-limited, and may present painful sensitivity at the site. The main differential diagnoses include pyogenic granuloma, cysts, lipoma, dermatofibroma, neurofibroma, and sarcoma.^{6,7} Because it is infrequent, it is usually a neglected entity in the evaluation of benign tumor lesions, with other diagnostic hypotheses raised prior to FN. Many cases are usually confirmed by histopathological examination.⁸

USG can be performed and may show well-defined, hypoechoic, dermal nodular lesions, with or without the presence of a heterogeneous hyperechoic center, and may therefore make a differential diagnosis with malignant nodular lesions. Consequently, anatomopathological examination is necessary in these cases.⁹

Histopathology demonstrates a well-circumscribed, fascial or intramuscular subcutaneous nodule with a star-like appearance. A proliferation of rounded and fusiform fibroblast

and oval nucleus myofibroblasts with thin chromatin and prominent nucleolus is seen. In newly-onset lesions, the cells are loosely arranged in an edematous and myxomatous stroma, while the older lesions show hyalinized collagen bundles.¹⁰ Fibroblasts and myofibroblasts react positively to vimentin and specific muscle actin and some cells are CD68 positive.¹¹ The histological differential diagnosis includes fibrosarcoma and malignant fibrous histiocytoma.

The treatment consists of complete surgical excision of the lesion, with a variable recurrence rate according to the literature, probably due to the incomplete excision of the lesion.

Other types of treatment may be considered conforming to the location of the lesion, such as CO₂ ablative laser and intralesional infiltration with triamcinolone.¹²

The main objective of this study was to highlight the unusual location of the case and alert the dermatologist for the diagnosis and inclusion of NF as a differential diagnosis of other facial tumors, in order to define the best therapeutic approach of the lesion, avoiding diagnostic errors and possible local recurrences. ●

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Mohs micrographic surgery in the treatment of penile cancer

A cirurgia micrográfica de Mohs no tratamento do câncer de pênis

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RESUMO

O câncer de pênis é um tumor raro, mas que apresenta grande impacto na qualidade de vida dos pacientes. No Brasil, a incidência é de 5,7 casos/100.000 homens/ano, representa 2% de todos os tipos de câncer que acometem homens no país e, em 2015, culminou em 402 mortes. O tratamento tradicional é a penectomia total com uretostomia perineal e consequente perda da manutenção das funções sexuais e urinárias normais. Para a preservação da função peniana, a cirurgia poupadora de órgão é preferida quando possível e a cirurgia micrográfica de Mohs conFigure-se em uma importante alternativa cirúrgica.

Palavras-chave: Carcinoma de Células Escamosas; Cirurgia de Mohs; Neoplasias Penianas

ABSTRACT

Penile cancer is a rare tumor that has a significant impact on patients' quality of life. In Brazil, the incidence is 5.7 cases/ 100,000 men/year, representing 2% of all types of cancer affecting men in the country and, in 2015, it culminated in 402 deaths. The traditional treatment is total penectomy with perineal urethrostomy and the consequent loss of normal sexual and urinary functions. For the preservation of penile function, organ-sparing surgery is preferred when possible, and Mohs micrographic surgery is an essential surgical alternative.

Keywords: Carcinoma, Squamous Cell; Mohs Surgery; Penile Neoplasms

INTRODUCTION

Penile cancer is a rare tumor worldwide, but it has a great impact on patients' quality of life. According to a Colombian study of systematic review and meta-analysis carried out in 2017, the incidence of penile cancer worldwide is 0.84 cases per 100,000 men/year, with the highest incidence rate found in Romania: 7.6 cases per 100,000 men/year. Brazil approaches the upper mark, with an incidence of 5.7 cases/100,000 men/year.¹

According to the National Cancer Institute (INCA), the malignancy corresponds to 2% of all types of cancer that affect men in the country and that, in 2015, culminated in 402 deaths.² The most common sites of involvement in decreasing order are: the glans, the foreskin, and the penile shaft³. Traditional treatment is partial or total amputation of the penis associated with urethral reconstruction and consequent loss of ability to main-

tain sexual intercourse and normal urinary functions.³

In a survey conducted by Opjordsmoen, S et al, men with penile cancer would choose the treatment with the lowest long-term survival to increase the chance of being sexually potent.⁴ Therefore, to preserve penile function, today organ-sparing surgery is preferred when this possibility exists.

We present a case of a 46-year-old patient with moderately differentiated squamous cell carcinoma in the glans, who opted for surgery with margin control by cutting and freezing intraoperative to preserve the male organ.

CLINICAL CASE

A 46-year-old male patient with phototype III attended the dermatology clinic referred by the urology team for evaluation of a hardened, ulcerated consistency plaque on the glans for 1 year. He had a history of moderately differentiated invasive squamous cell carcinoma (SCC) in the glans 10 years ago, approached with partial penectomy and inguinal and pelvic emptying by laparoscopy, with one positive lymph node. He denied risky sexual behavior. After partial penectomy, the patient got married and had three children. On physical examination, he presented hypochromic stains on the penile glans and a hardened erythematous-hypochromic plaque with central ulceration on the ventral face of the glans (Figure 1). A biopsy was performed on the lesions that showed moderately differentiated SCC with an infiltrative growth pattern, with no identified angiolymphatic invasion and penile intraepithelial neoplasia. Computed tomography of the abdomen and pelvis showed no evidence of lymph node metastasis. In view of the findings of a new malignant lesion restricted to the glans, the tumor staging was T1aN0M0, with imprecise clinical limits. To preserve sexual function, Mohs Micrographic Surgery (CMM) was chosen in a joint approach with the urology team. We marked the clinical limits of the lesion (Figure 2), antisepsis, field placement and local anesthesia with Klein's solution. Excision of the visible tumor was performed with a scalpel angled at 90°, a stage known as Debulking (Figures 3 and 4). Then, the surgical defect was enlarged by 2mm, deeply and peripherally, with resection of part of the distal urethral mucosa, configuring the 1st. CMM stage (Figure 5). In this stage, before the complete removal of the fragment, cross-sectional markings were made (Nicks), which served as a reference in the perilesional tissue for correct mapping of possible margins to be enlarged. The excised part was divided into four fragments that were placed in gauze with standardized orientation, stained and named A1, A2, A3, and A4, for making the histological slide by freezing and topographic map (Figures 5 and 6). After histopathological analysis, tumor-free margins and foci of penile intraepithelial neoplasia reaching the edges of fragments A3 and A4 (figures 6 and 7) were found. Primary closure was performed (figure 8) after defining the subsequent clinical treatment of the other lesions identified at the anatomical pathological examination.

DISCUSSION

Penile cancer affects mainly men in the sixth and seventh decades of life, and may also affect younger men.



FIGURE 1: Erythematous hypochromic plaque with inaccurate boundaries and ulceration in the center, ventral glans face, on the left



FIGURE 2: Image showing the marking of the clinical limits of the tumor lesion

The mutilating potential of therapeutic surgical approaches permanently and significantly affects the quality of life of these patients. Recent studies emphasize that the histopathological characteristics of these tumors and the absence of lymph node involvement are more significant prognostic factors than the aggressiveness of surgical treatment.⁵ The most common histological variant is SCC, which corresponds to more than 95% of cases.³ Organ preservation can be achieved through non-surgical therapies such as external radiotherapy, brachytherapy, and topical immunomodulators. However, these are limited options in more advanced cases and have several undesirable consequences.⁶ All patients must be circumcised before considering conservative non-surgical treatment.³ The option for radiotherapy involves high rates of radiation, with proportionally high rates of urethral stenosis, fibrosis, and penile necrosis, and has higher failure rates than partial penectomy.⁷ Topical immunomodulators, such



FIGURE 3: Debulking



Figure 5: 1st stage of Mohs micrographic surgery showing deep and peripheral margins enlargement



FIGURE 4: Tumor sample

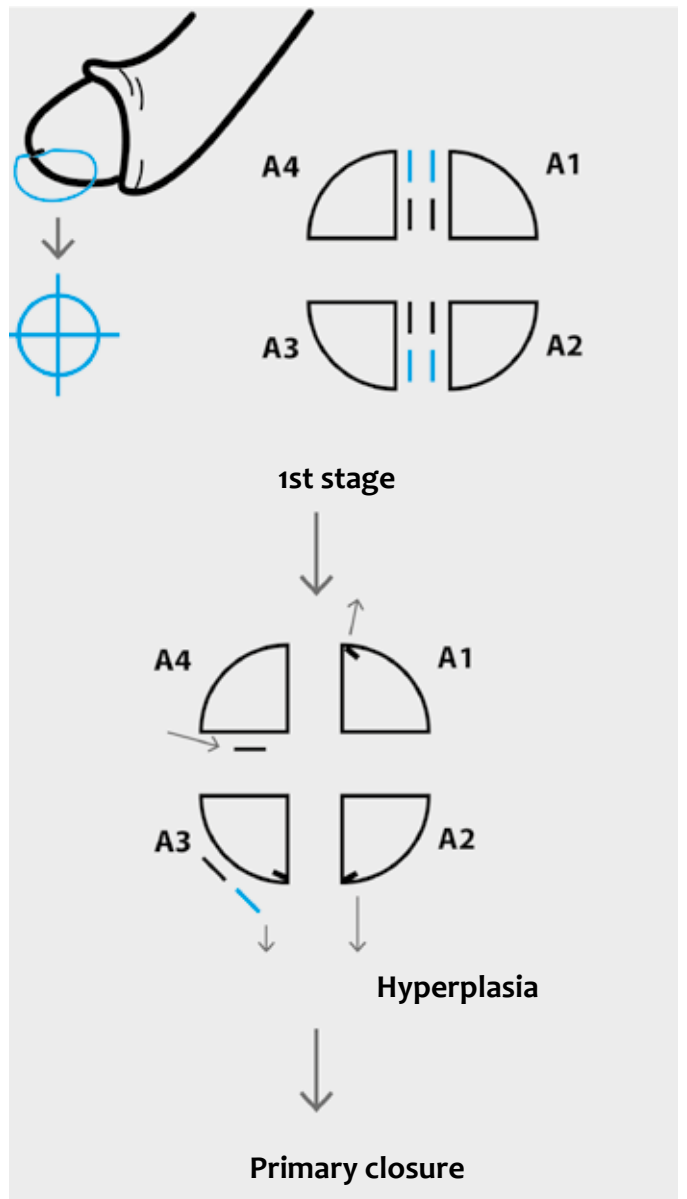


FIGURE 6: Topographic map of Mohs micrographic surgery

as Imiquimod 5% or 5-fluouracil, are effective for carcinomas in situ, have modest cure rates (63%), require daily application for 6-8 weeks, and can make it difficult to identify recurrent disease.⁸

Genital preservation surgeries aim to completely excise the primary tumor and perform local reconstruction, if necessary, to preserve the patient’s reproductive and urinary functions.⁹ Genital preservation surgeries are indicated for tumors in situ / Ta / T1 and some well or moderately differentiated T2 tumors and selected cases of T3 stage.^{10,11} The main disadvantage of these procedures is the higher local recurrence rate, which requires quarterly monitoring in the first two years, every six months from the 3rd to the 5th year, and annual until the 10th postoperative year.³

Among the surgical options, we have laser surgery, Mohs micrographic surgery, circumcision, local wide excision, “glans resurfacing”, glandectomy, partial penectomy, and total penectomy with preservation of the urethra. Mohs micrographic surgery uses intraoperative microscopic evaluation to ensure complete tumor excision with maximum preservation of normal perilesional tissue. It has excellent cure

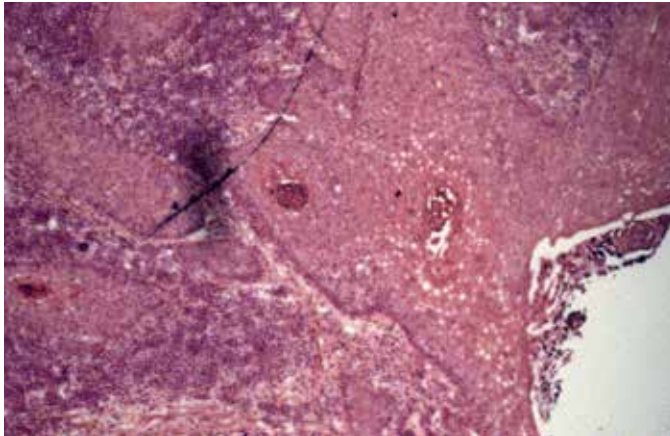


FIGURE 7: Cut and freeze blade photo: Proliferation of atypical squamous cells, with formation of projections that infiltrate the dermis, coexisting hyperkeratosis, dyskeratosis, and chronic lichenoid inflammatory activity



FIGURE 8: Primary closure of surgical defect

rates for SCC from other locations, but the literature shows recurrence rates of 26–32% when located on the penis.^{12,13,14}

However, a study shows that with the strict maintenance of oncological surveillance and consequent re-approaches when necessary, the overall survival rates are excellent and the rates of progression are low.¹⁵ The same study considers that Mohs micrographic surgery should not be limited to the approach of low-grade, small, and superficial tumors as was suggested in the older literature, but that it should be used as a treatment strategy for tumors with urethral involvement, saving patients from total or partial penectomies.¹⁵ The recurrence rate found

in this study was 11.1% and the re-approach of these patients with new Mohs micrographic surgery showed a final cure rate of 100%, both in cases of SCC in situ (mean follow-up of 72.5 months) and invasive SCC (mean follow-up of 77 months). In view of the great psychosocial impact of aggressive penile cancer treatment and the possibility of management with preservation of sexual and urinary functions associated with good overall survival rates, we believe that the best approach is one that allows the maintenance of quality of life despite the need for more frequent and prolonged follow-ups. ●

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Trilobate flap for nasal reconstruction: optimizing results

Retalho trilobado para reconstrução nasal: otimizando resultados

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ABSTRACT

The trilobate flap can be used for the reconstruction of mid-size defects of the lower nasal portion. The third lobe determines the rotational arch's increase, reducing the flap tension and improving the final aesthetics. We report the case of a patient submitted to the procedure with good aesthetic and functional results.

Keywords: Dermatologic Surgical Procedures; Mohs Surgery; Surgical Flaps

RESUMO

O retalho trilobado pode ser usado para a reconstrução de defeitos de tamanho médio da porção nasal inferior. O terceiro lóbulo determina o aumento do arco rotatório, reduzindo a tensão do retalho e melhorando a estética final. Relatamos o caso da uma paciente submetida ao procedimento com bom resultado estético e funcional.

Palavras-chave: Cirurgia de Mohs; Retalhos Cirúrgicos; Procedimentos Cirúrgicos Dermatológicos

INTRODUCTION

Nasal reconstruction is always challenging for the dermatological surgeon. Resection of skin neoplasms usually causes losses of nasal substance. There are many alternatives for skin coverage, and skin flaps are the best option. However, the choice of the best technique must be considered individually to achieve the best functional and aesthetic results, with minimal distortion of the anatomy. We report the case of a young patient with a significant nasal defect after tumor resection and rotation of a trilobed flap.

Case report

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CASE REPORT

The patient was diagnosed with basal cell carcinoma at the transition from the nasal tip to the nasal dorsum. The incisional biopsy showed a tumor of approximately 1.2 cm (Figure 1). After undergoing tumor resection through Mohs micrographic surgery, the patient presented a defect of approximately 1.6 cm (Figure 2). After intraoperative analysis of the best surgical technique, we chose to perform a transposition flap with a variation of the bilobed flap technique (Figure 2). The immedi-



FIGURE 1: BCC on the nasal tip and preoperative marking of the limits



FIGURE 2: Excision by Mohs micrographic surgery and trilobed flap marking with methylene blue



Figure 3: Immediate postoperative period



Figure 4: Late postoperative period: 60 days

ate postoperative period showed little distortion of the anatomy (Figure 3). Reassessment was performed 90 days after surgery, with excellent aesthetic and functional results (Figure 4).

DISCUSSION

Partial-thickness nasal defects at the nasal tip present unique reconstructive challenges. The literature cites the bilobed flap and the extended glabellar flap as the first options for these types of defects for nose tip reconstruction.^{1,2}

The dorsal nasal flap is a widely used reconstruction technique that preserves the distal nasal contour as a sliding flap, with rotation and advancement elements, accessing the reservoir of loose nasal skin and dorsal glabellar. The repair occurs using long suture lines, hidden along the nasofacial groove and the glabella, but always with an oblique distal nasal suture line at the standing cone repair site.^{2,3}

The trilobed flap recently described and detailed by Albertini and Hansen, is an elegant technique to address the nasal tip's lower nasal defects. This flap further expanded the application of transpositional flaps for nasal repair that were previously repairable only by interpolation or grafts.³

This technique presents as a good option in case of small and medium defects in the distal region of the nose as it allows greater mobility with the use of tissue from the upper nasal portions.

This flap consists of the incorporation of the third lobe into the bilobed flap. The third lobe allows a total rotating arc of 135° to 150° with lobes separated by an external rotating angle of 45° to 50° oriented perpendicularly to the free margin, which determines an increase in the rotational arc, facilitating orientation and decreasing the tension perpendicular to the alar edge.³

The flap is projected around this pivot point, drawing arches and creating lobes of equal or almost equal size. The final flap design, including exact lobe size and length, is altered to a small degree on a case-by-case basis, based on various factors: swelling of the tissues at the time of reconstruction, elasticity, stiffness of the nasal skin, and surrounding scar.^{4,5}

Due to the greater tissue mobility obtained with the multilobed flap, there is no need to extend the skin incisions superior to the glabella. The natural upper incisions (and the true reservoir) are found in the upper lateral wall and the medial nasal skin.⁴

This flap depends on the upper nasal portion. Therefore, patients with thin nasal skin, previous radiation, or previous nose surgery can benefit from other reconstructive options.^{3,5}

Unlikely results with difficult healing, nasal asymmetry, especially alar, reduced airflow by anchoring in the ipsilateral internal nasal valve^{3,6} and tip elevation are possible complications. However, they are usually avoided with proper patient selection, flap design, and surgical execution.⁴

The trilobed flaps create a flap with predictable tissue movements and outcomes and can be used for defects in the nasal tip with excellent functional and aesthetic results. Selection of the type of flap for each defect remains crucial to the success of reconstruction. ●

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Tunneled island flap for reconstruction of eyebrow defect

Retalho em ilha tunelizado na reconstrução de defeito na sobrancelha

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ABSTRACT

Facial surgeries are often challenging for surgeons due to their complex anatomy, aesthetic and functional importance. Interventions in the cosmetic subunit that comprises the eyebrows can be particularly difficult. This case report aims to demonstrate the use of island flap in this region. We performed the excision of melanocytic nevus, with a good outcome and absence of postoperative complications. This technique is commonly used for facial reconstruction; however, the literature describes only a few cases using this method for eyebrow reconstruction.

Keywords: Eyebrows; Reconstruction; Surgical Flaps

RESUMO

Cirurgias na face conFigurem-se, muitas vezes, em desafios para o cirurgião em virtude da complexa anatomia, importância estética e funcional. Intervenções na subunidade cosmética que compreende as sobrancelhas podem ser particularmente difíceis. O objetivo deste trabalho é demonstrar o uso do retalho em ilha nesta região. Foi realizada exérese de nevo melanocítico, com bom resultado final e ausência de complicações pós-operatórias. Esta técnica é comumente utilizada para reconstrução facial, porém existem poucos casos descritos em que foi utilizada para reconstrução de sobrancelhas.

Palavras-chave: Reconstrução; Retalhos Cirúrgicos; Sobrancelhas

INTRODUCTION

The reconstruction of facial defects is a challenge, considering its cosmetic importance and the scarcity of adjacent skin in some face areas. Skin flaps represent a modality frequently used in reconstructive surgery of the face. Island flap is one of the reconstruction options that can produce good results in this region due to this type of flap's characteristics.¹

There are few reports in the literature on eyebrow reconstruction using the tunneled island flap. This study presents a description of the performance of this technique.

METHODS

A female patient presented with a congenital melanocytic nevus, occupying two thirds in the lateral of the right eyebrow (Figure 1). We marked the site to be incised on the scalp based on the region's anatomy and palpation of the temporal artery. In the area of the frontal branch of the temporal artery, we drew the flap with the shape of the future eyebrow, with correct measurement for a perfect fit (Figure 2). A subcutaneous tunnel was then made through which the island flap passed and was positioned in the eyebrow region (Figures 3, 4, and 5). The flap was sutured with simple stitches of 6.0 mono nylon thread. The scalp incision was sutured with 4.0 mono nylon thread (Figure 6).

RESULTS

The tunneled island flap's final result evidenced the absence of scarring between the scalp and the eyebrow. There were no postoperative complications or distortion of the anatomy, with only the need to periodically trim the hairs (Figures 7 and 8).



FIGURE 1: Congenital melanocytic nevus on the eyebrow



FIGURE 2: MARKING THE ISLAND FLAP



FIGURE 3: Surgical defect after excision of the tumor



FIGURE 4: Subcutaneous tunnel through which the flap passes



FIGURE 5: Flap positioned in the eyebrow area

DISCUSSION

The eyebrows, essential subunits of the upper third of the face, play an aesthetic role in facial expression and eye protection and require care in reconstructions to achieve a natural result.² Total or partial loss of eyebrows has negative physical and psychological consequences.³ It can result from heat or radiation burns, avulsion injuries, alopecia areata, leprosy, or surgical excision of neoplasms.⁴



FIGURE 6: Suture with simple stitches

Reconstruction after eyebrow deformity can be challenging, as donor sites must have hair follicles, texture, and sebaceous quality similar to that of the recipient area.⁵ The technique must be individualized, so in the case of thinner eyebrows, it can be performed using a composite graft. For thicker eyebrows, the island flap in the superficial temporal artery territory would have greater applicability,⁶ representing a good option when the defect is in the lateral region of the eyebrows.⁷ Therefore, planning of the flap area with the exact dimensions of the defect to be filled is required to avoid anatomical distortions.

Different reconstruction techniques have been described worldwide, such as cosmetic tattoos, hair grafts, temporoparietal fascia flaps, scalp flaps, and tunneled island flaps.⁸ The tunneled island flap is a technique that incises a skin island and creates a subcutaneous tunnel between the flap and the defect. The flap is mobilized and transferred through this tunnel, superior to the defect. It is useful when you want to avoid any skin incisions between the donor site and the defect, thus creating a subcutaneous tunnel in the forehead to allow the transfer of a skin island from the scalp to the eyebrow.⁹

The island flap has two fundamental characteristics: the donor skin is an island, highlighted on all sides of the surrounding epidermis and dermis; and a subcutaneous pedicle is maintained, ensuring vascularity and allowing some mobility to a nearby receptor area.¹⁰

These flaps are extremely perfused, mobile, and the tunnel created provides the most direct path to the primary defect. Also, they require only a single surgical time, thus eliminating the morbidity of subsequent procedures. However, in addition to the risks common to all surgical procedures, such as hemorrhage, infection, and necrosis, these flaps are technically challenging, and the dissection of the pedicle is slow, as it has to be performed carefully.¹

CONCLUSION

The tunneled island flap is a technique commonly used for facial reconstruction. However, only a few cases used this technique for eyebrow reconstruction. In our case, we obtained an aesthetically satisfactory result, achieving an appearance sim-



Figure 7:
Final result



FIGURE 8: Final result (side view). There is no scar between the scalp and the eyebrow

ilar to that of the contralateral eyebrow, absence of scar between the scalp and the eyebrow, and preservation of the motor and sensitive innervation of the forehead. Dermatological surgeons and others professionals qualified in surgical procedures in this area can reproduce this technique with the same results. ●

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Perianal Buschke-Lowenstein tumor: report of two cases treated with 25% podophyllin ointment

Tumor de Buschke-Lowenstein perianal: relato de dois casos tratados com podofilina em vaselina sólida 25%

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ABSTRACT

Penile cancer is a rare tumor that has a significant impact on patients' quality of life. In Brazil, the incidence is 5.7 cases/ 100,000 men/year, representing 2% of all types of cancer affecting men in the country and, in 2015, it culminated in 402 deaths. The traditional treatment is total penectomy with perineal urethrostomy and the consequent loss of normal sexual and urinary functions. For the preservation of penile function, organ-sparing surgery is preferred when possible, and Mohs micrographic surgery is an essential surgical alternative.

Keywords: Podophyllin; Condylomata Acuminata; Carcinoma, Verrucous; Neoadjuvant Therapy

RESUMO

O tumor de Buschke-Lowenstein (TBL) é variante extremamente rara do condiloma acuminado que, apesar de manifestar-se clinicamente por lesões de grandes proporções, apresenta comportamento biológico e características histológicas benignas. Existem diversas abordagens terapêuticas disponíveis, muitas delas apoiando-se em abordagens cirúrgicas extensas e mutilantes. Apresentamos dois casos de pacientes com TBL tratados com podofilina tópica, cujas respostas terapêuticas foram extremamente favoráveis em ambos os casos.

Palavras-chave: Podofilina; Condiloma Acuminado; Carcinoma Verrucoso; Terapêutica; Terapia Neoadjuvante

INTRODUCTION

The Buschke-Lowenstein tumor (BLT), also known as giant condyloma acuminata, is a rare variant of condyloma acuminata, comprising about 0.1% of cases.¹ It is caused by human papillomavirus (HPV) types 6 and 11.² The risk factors described are immunosuppression, pregnancy, alcohol and tobacco consumption, poor local hygiene, and Herpes simplex virus infection.³

Clinically, the lesion reaches massive proportions, with aggressive local characteristics, invading and causing deformity in the adjacent tissues, but without lymphatic, vascular, or neuronal invasion.⁴ The risk of degeneration for squamous cell carcinoma (SCC) ranges from 30% to 56%. At the same time, condylomata acuminata has a risk of only 2%.⁵ Histologically, BLT is distinguished from condylomata acuminata by its pro-

liferation and deep penetration into adjacent tissues, and from SCC, by the integrity of the basement membrane and inability to produce metastases.³

BLT has several therapeutic approaches, such as topical medications, cryosurgery, surgical excision, immunotherapy, chemotherapy, radiotherapy, and electrocoagulation. There is no consensus to guide the therapeutic decision, but the initial choice in many services is surgical excision.⁵

Surgery should be local resection, keeping margins free of residual disease. Surgical removal can cause extensive wounds, cicatricial strictures, and fecal incontinence. Abdominoperineal amputation is indicated when the sphincter apparatus is involved.⁶ Among the most used topical agents are podophyllin, which has an exfoliating, immunological, and antimetabolic action,^{5,6} and imiquimod, an immunomodulatory substance capable of enhancing the immune response to HPV.^{5,6}

CLINICAL CASE 1

A 25-year-old man with positive serology for HIV for eight months, using antiretroviral therapy (ART) with tenofovir, efavirenz, and lamivudine since his diagnosis, presenting CD4 cell count of 253/ μ L and undetected viral load.

He complained of an anal tumor for nine months, evolving in the last month with an increase in the lesion, associated with a foul odor and difficulty in hygiene. Upon dermatological examination, he presented a large tumor with a verrucous aspect, 30 cm in diameter, occupying the buttocks, perineum, and intergluteal sulcus, making it impossible to identify the anus (Figure 1).

The biopsy of the lesion ruled out SCC, which report concluded it was a condyloma acuminata. We prescribed sulfamethoxazole + trimethoprim, due to secondary infection, and topical treatment with podophyllin 25% in solid petroleum jelly started once a week, instructing the patient to remove the medication, washing it after six hours (Figure 1).

After 23 applications for six months, we observed almost complete regression of the lesion (Figure 2). After that, surgical resection and cauterization of the few remaining lesions were indicated. The histopathological evaluation of the resected piece did not present carcinoma, and the clinical evolution was favorable. The wounds healed after four weeks, with no signs of recurrence.

CLINICAL CASE 2

A 42-year-old woman, positive for HIV for six years, complaining of an anal tumor for 12 months. She withdrew ART after six months, with a significant increase in the lesion. The patient had 124/ μ L CD4 T lymphocytes and a viral load greater than 200,000/mL.

She was instructed to resume ART and started weekly treatment with podophyllin 25% in solid petroleum jelly. After the third application, the lesion showed a significant reduction. She received 12 applications over four months when the disappearance was complete (Figure 3). The biopsy performed at the first consultation revealed condyloma acuminata, with no signs of malignancy. Anal colposcopy and cytology of the anal canal were normal at the end of treatment. There was no recurrence.

DISCUSSION

BLT involving the perianal region is rare. A meta-analysis evaluating publications from 1958 to 2000 found only 51 cases. It is more frequent in men (2.7:1) with an average age of 43.9 years.⁷

There appears to be a complex interaction between HIV, HPV, and local mucosa's immunological mechanisms. HIV increases HPV transcription, and it causes a decrease in the number of macrophages, Langerhans cells, and CD4 T lymphocytes in the mucosa, with a consequent reduction in local immune control of HPV infection and increased proliferation of this virus.⁸

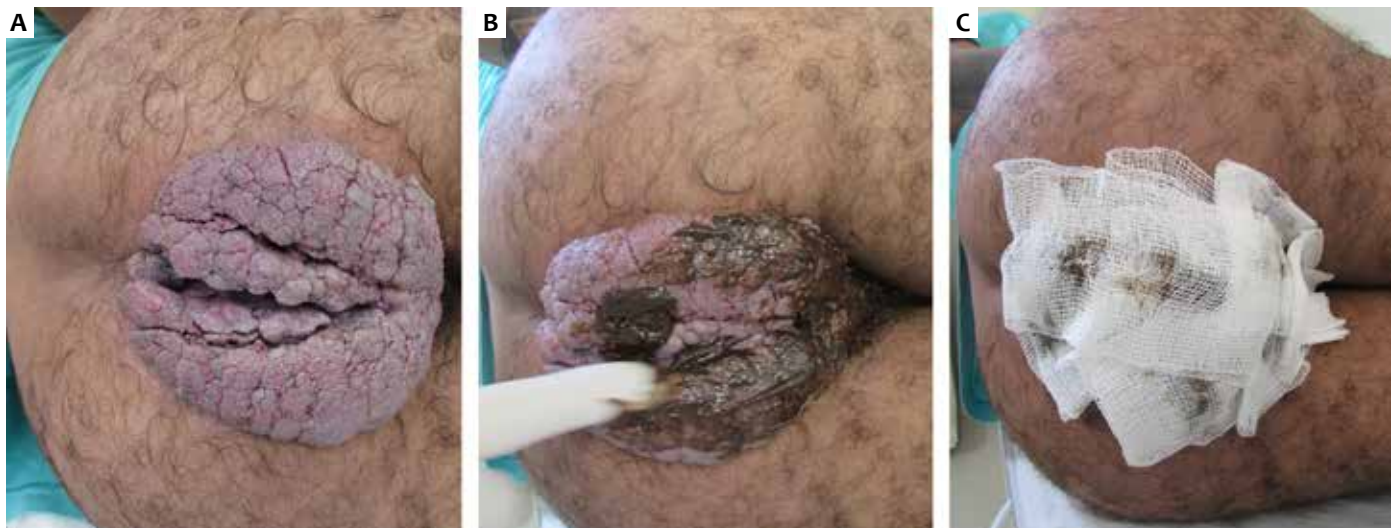


FIGURE 1: Buschke-Lowenstein tumor, case 01. (A) Clinical aspect of the lesion. (B) Application of 25% podophyllin in solid petroleum jelly to the lesion and (C) gauze protection before the patient puts on his clothes



FIGURE 2: Buschke-Lowenstein tumor, case 01. **(A)** Clinical aspect after 23 weekly sessions performed for six months. **(B)** Detail of the perianal region with spacing of the buttocks



FIGURE 3: Buschke-Lowenstein tumor, case 02. **(A)** Clinical aspect of the pre-treatment lesion and **(B)** after 12 weekly sessions performed for four months

Although resection is the most commonly performed procedure, topical treatment was instituted to reduce the lesions' size, thus facilitating resection and postoperative discomfort. Podophyllin 25% in solid petroleum jelly was used because it is easy to obtain and has low cost since the authors work in public hospitals and have experience with this medication in anogenital condyloma acuminata, including in children.^{1,5,6} This procedure causes remission of the lesions, avoids surgeries, and facilitates surgical procedures and postoperative recovery.

The patients are being followed up and periodically reassessed with dermatological and proctological examination. If there is no clinical lesion, cytology and anal colposcopy are performed. This follow-up strategy is necessary for surveillance of malignant lesions and early detection of clinical and subclinical recurrences.^{7,9}

CONCLUSION

Treatment with topical substances can be instituted for BLT cases, with the possibility of reducing lesions, facilitating operative treatment, and avoiding complications caused by healing. ●

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Case report

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Extensive Vulvar Keloid Post Multiple Treatments for Genital Condylomas: Case Report

Extenso queuloide vulvar pós-tratamentos para condilomas genitais: Case report

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ABSTRACT

Genital infection by Human Papillomavirus occurs 65% of the time after contact with contaminated skin and mucosa. Most people eliminate the virus through the immune system. Individual factors such as smoking, age, multiparity, prolonged use of hormonal contraceptives, sexual behavior, and infection by other Sexually Transmitted Diseases cause viral perpetuation and genital lesions, most of them asymptomatic. The most affected regions are anogenital. Treatment should be individualized. Excessive treatment can lead to scarring. We report a case of multiple treatments for removal of genital condyloma with consequent formation of extensive vulvar keloid.

Keywords: Condyloma acuminatum; Scar; Scar, Hypertrophic; Keloid

RESUMO

A infecção genital pelo papilomavírus humano ocorre em 65% das vezes após contato com pele e mucosa contaminadas. A maioria das pessoas elimina o vírus através do sistema imune. Fatores individuais, como tabagismo, idade, multiparidade, uso prolongado de contraceptivos hormonais, comportamento sexual e infecção por outras doenças sexualmente transmissíveis, ocasionam perpetuação viral e lesões genitais, em sua maior parte, assintomáticas. As regiões mais afetadas são anogenitais. O tratamento deve ser individualizado. O tratamento agressivo pode ocasionar distúrbios cicatríciais. Relatamos um caso de múltiplos tratamentos para retirada de condiloma genital com consequente formação de extenso queuloide vulvar.

Palavras-chave: Cicatriz; Cicatriz Hipertrófica; Condiloma Acuminado; Queuloide

INTRODUCTION

Human papillomavirus (HPV) is a very prevalent sexually transmitted disease (STD). Subtypes 6 and 11 cause genital warts and have a low potential for malignancy.¹ The risk of transmission of verrucous lesions from sexual contact is 65%. After contact with the virus, most people eliminate it. However, in some individuals, there is viral perpetuation, due to factors such as smoking, age, multiparity, prolonged use of hormonal contraceptives, sexual behavior, and infection by other STDs.² Subclinical forms and lack of knowledge about HPV manifestations increase the potential for transmission.^{3,4}

After contact with HPV, there are three forms of infection: latent (detection of viral DNA), subclinical (microscopic change), and clinical (visible, verrucous, and often in anogenital regions). Non-macroscopic manifestation and non-knowledge

of the lesions lead to potential transmitters.^{3,4} Diagnosis involves psychological care. The patient may feel guilty or punished, thus avoiding treatment, or performing it in an exacerbated way.²

The treatment is individualized and seeks removal of the lesion. Measures to reduce transmissibility⁴ should be targeted, followed by medications, surgeries, and immunotherapy.

Vulvar condylomas can be treated with trichloroacetic acid 70 to 80%, Podofylline (effect on lesions smaller than 10 cm²) or Imiquimod (antiviral and antitumor effect for limited lesions).⁸

Extensive and recurrent lesions are treated surgically.⁵ CO₂ vaporization is effective for extensive lesions of the lower genital tract with a resolution of up to 71%.⁶ Large lesions requiring histopathology are removed by surgical excision.⁷ Due to the high rates of recurrence (25%), multiple treatments can be applied, being detrimental because of the potential of disorders in the cicatricial processes. Healing is the new tissue in a region of solution of continuity.

Some factors interfere and form keloid scars (reddish, irregular, pigmented, and without respect to the region's previous margins).⁷ The pathophysiology of the keloid is still unclear; hyperproliferation of collagen and fibroblasts of the dermis is known to cause exacerbated scarring.⁸ Among the risk factors are race black/yellow, age between 10 and 30 years, female, hypertension, and excessive surgical manipulation. The sites most affected are upper abdomen, ear lobule, and sternal region. Such scars are free of dermal appendages, with a smooth and shiny appearance.^{8,9} We report a case of cicatricial hypertrophy after numerous treatments for excision of genital condyloma with formation of extensive vulvar keloid and physical and emotional sequelae for the carrier.

CASE REPORT

A 22-year-old white, nulligesta woman with on combined hormonal contraceptive use and male condom, without comorbidities, was admitted to a gynecological clinic with a history of extensive vulvar condylomatosis treated with chemical cauterization and thermokauterization. It evolved with itchy and painful vulvar lesion. The vulvar physical examination showed extensive fibrous plaques lesion, on the clitoris, 5 cm in diameter, extended to large lips, hardened and fixed, obliterating the clitoris (Figure 1). The diagnostic hypothesis of extensive vulvar keloid was confirmed by biopsy, suggesting sequelae of multiple treatments. Prescribed high-potency corticosteroid and simple vulvectomy schedule with local infiltration of large lips with injectable triamcinolone 20mg/ml. Resected lesion under locoregional anesthesia with no possibility of surgical wound suture due to localization and extension, leaving wound healing for second intention, under hygiene care and occlusive dressing with oily lotion, as seen in Figure 2. She remained two days hospitalized for analgesia and learning of care. On the 13th postoperative day: no complaints, satisfied, without pain and presenting a wound of good evolution. In the locality, good granulation of the surgical wound, without signs of infection and without formation of new keloid, decrease in the hardening of the labia



FIGURE 1: Extensive vulvar keloid formed from multiple treatments for genital condyloma in a female patient, with diagnosis confirmed by histopathological study



FIGURE 2: Immediate surgical result of the resection of the extensive vulvar keloid on the pubic hill and application of injectable Triamcinolone on the area of small labial lips, with a bloody wound for healing by second intention

majora and the obliteration of the clitoris. On the 25th postoperative day, wound with good evolution, decrease of diameter and depth of bloody area (Figure 3). On the 75th postoperative day, area of young tissue under the former surgical wound and improvement of skin thickening in large lip region (Figure 4),

local pruritus complaint, prescribed topical corticosteroid therapy and hydration. After 105 days, a totally epithelial bloody area and a 1.5 cm scar in the region, without relief, color discreetly pinker than the original skin, improved hardening around the clitoris and large lips, good individualization of small and large lips. Patient in clinical follow-up three years ago, with no signs of recurrence of keloid or condylomas and satisfied with final treatment results.

DISCUSSION

HPV is a sexually transmitted disease whose infection occurs by accessing the virus to the epithelial basement mem-

brane by microtraumas during sexual intercourse or virus entry into the cervix transformation zone.¹

Patients diagnosed with HPV infection are emotionally fragile, presenting fear, guilt, and anger, because the social stigmas of STDs.¹⁰ This can provoke a desire for punishment by avoiding treatment or by performing it exacerbated.³ The patient, on menacing and using oral hormonal contraceptive, risk factors for viral perpetuation, underwent treatment for vulvar condylomatosis with chemical cauterization and electrocauterization and evolved with extensive vulvar lesion.³ The electrocauterization destroys the tissue by heat, among its adverse effects are the cicatricial process, loss of pilification, retractions, and local hypochromia.⁸ The patient evolved, biopsy confirmed keloid tissue, requiring treatments for scar excision and personal satisfaction. Gomes et al states that spontaneous regression is possible and present in up to 30% of cases within three months of observation.¹⁰ Already the excessive treatment can be cause an abnormal cicatricial process. The patient presents as risk factors to age, female gender, and excessive surgical manipulation.^{5,6,8} It is questioned to what extent overlapping treatments would bring more benefits than harm.^{3,8}

CONCLUSION

HPV is present in most of the sexually active population, mostly eliminated by the immune system. Recurrence should be treated cautiously. Exaggerated manipulation of the region is a risk factor for inadequate scarring and formation of keloids. The psychological counseling necessary for many patients is emphasized. ●



FIGURE 3: Result of postoperative evolution on the 25th postoperative day of the resection of extensive vulvar keloid, in the healing process by second intention, with the aid of a dressing based on Dersani Hydrogel with Alginate®



FIGURE 4: Results of postoperative evolution on the 75th postoperative day of the resection of extensive vulvar keloid, in the healing process by second intention, with the aid of a dressing based on Dersani Hydrogel with Alginate®

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Case report

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Exuberant Proliferating Trichilemmal Tumor in a young person

Tumor triquilemal proliferante exuberante em jovem

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ABSTRACT

Proliferating Trichilemmal Tumor (PTT) is an uncommon benign neoplasm that usually affects adult women over 60 years old, preferably as a solitary nodule in the scalp. We report a case of PTT in a young woman presenting exuberant, asymptomatic scalp tumors 7 years ago, which recurred after surgical exeresis. The histopathology confirmed the diagnosis, and surgical excisions and clinical follow-up were performed. PTT appears to originate from the wall of trichilemmal cysts, from the isthmus of the hair follicle's outer root sheath. Usually, it has benign biological potential with a low risk of malignancy and high recurrence. The therapy of choice is surgery.

Keywords: Cysts; Neoplasms; Scalp

RESUMO

Tumor triquilemal proliferante (TTP) é uma neoplasia benigna incomum que acomete, habitualmente, mulheres adultas acima de 60 anos, preferencialmente como nódulo solitário no couro cabeludo. Relata-se um caso de TTP em mulher jovem, apresentando exuberantes tumorações assintomáticas no couro cabeludo, há sete anos, que recorreram após exéreses cirúrgicas. O histopatológico confirmou o diagnóstico, tendo sido realizadas retiradas cirúrgicas e acompanhamento clínico. O TTP parece originar-se da parede dos cistos triquilemais, provenientes do istmo da bainha radicular externa do folículo piloso. Geralmente, apresenta potencial biológico benigno, com baixo risco de malignização e elevada recorrência. A terapia de escolha é a cirurgia.

Palavras-chave: Cistos; Couro Cabeludo; Neoplasias

INTRODUCTION

Proliferating trichilemmal tumor (PTT) is an epidermal adnexal neoplasia formed by several cysts containing squamous epithelium with trichilemmal keratinization, in other words, the abrupt transition of a nucleated epithelial cells to an anucleated in the absence of a granular layer.¹⁻³ PTT usually appears as a dermal nodule or solitary subcutaneous scalp of elderly women.^{1,3-13} It was first described by Wilson-Jones in 1966, who named it a proliferating epidermoid cyst due to its clinical and histological resemblance to squamous cell carcinoma.^{1,3, 5,9-13} Since then, PTT has received several other names, such as scalp pilaris tumor, proliferating trichilemmal cyst, trichilemic pillar tumor, invasive pilomatrixoma, hydatid keratinous cyst, trichoclamidocarcinoma, giant hair matrix tumor, hairy scalp tumor, and others.^{1,3,6} It exhibits the ability to grow rapidly over

a given period of time, but the evolution to malignant PTT is uncommon.^{5,7,11,12} Therefore, it presents heterogeneous histological behaviors ranging from benign, locally aggressive growth with chances of causing invasions in the underlying tissue layers up to that time, with local recurrence metastatic potential and regional lymph node metastasis.^{3,10,11,12} PTT is a rare tumor that corresponds to 0.1% of benign cutaneous tumors and may form from a pre-existing or concomitant trichilemmal cyst or as an isolated lesion.^{5,11,12}

CLINICAL CASE

We report the history of a 24-year-old female patient from Uberlândia (MG), treated at the dermatology department of the Clinical Hospital of the Federal University of Uberlândia, complaining of painless nodules in scalp for seven years ago. It refers to progressive growth in recent months, followed by ulceration with output of bloody discharge. Reports recurrence of previously excised scalp lesions. She denies having suffered any kind of trauma.

Dermatological examination showed exophytic, painless, ulcerated tumor with sanguinolent exudation, measuring 10x5 cm in size, in the left parietal region (Figure 1), and several other smooth-surface tumors, soft consistency, measuring 1 to 3 cm in diameter, distributed on the scalp, some surrounded by alopecia (Figure 2). There was no palpable adenomegaly in the cervical and scalp. Two lesions were excised (Figure 3), whose pathological examination showed squamous cell proliferation in the dermis with formation of tumors with irregular multiple lobules of varying sizes, differentiating into large keratinocytes with dis-

crete atypia, abrupt keratinization with foci of calcification and absence of invasion of adjacent tissues (Figure 4).

Trichilemmal tumor, pilomatrixoma, protuberant dermatofibrosarcoma was clinically suspected, but the correlation between clinical and histopathological findings confirmed the diagnosis of PTT.

Surgical treatment of some lesions has been proposed, with wide margin exercises and simple interrupted suture with good healing results. Due to the recurrent nature of the lesions, the patient remains in outpatient clinical follow-up and other injuries are scheduled.

DISCUSSION

Trichilemmal cysts correspond to 20% of cutaneous cysts, the others classified as epidermoid cysts originating from trichilemmal or external root sheath of the hair follicle.^{1,11} PTT seems to originate from these lesions, presumably after inflammation or trauma, due to the fact that have areas with benign characteristics and others with malignant properties.^{1,5-12} It differs from the trichilemmal cyst because it is uncommon, larger, and histologically more atypical, and the malignant proliferative trichilemmal tumor is less aggressive.⁹ PTT occurs more frequently in women (79,5-87% of cases)



FIGURE 1: Exophytic, ulcerated, 10x5 cm tumor with sanguinolent exudation on the left parietal region



FIGURE 2: Soft, smooth, some ulcerated tumors, 1 to 3 cm, surrounded by an area of alopecia on the scalp



FIGURA 3: Extensive excision of two proliferating trichilemmal tumors on the scalp

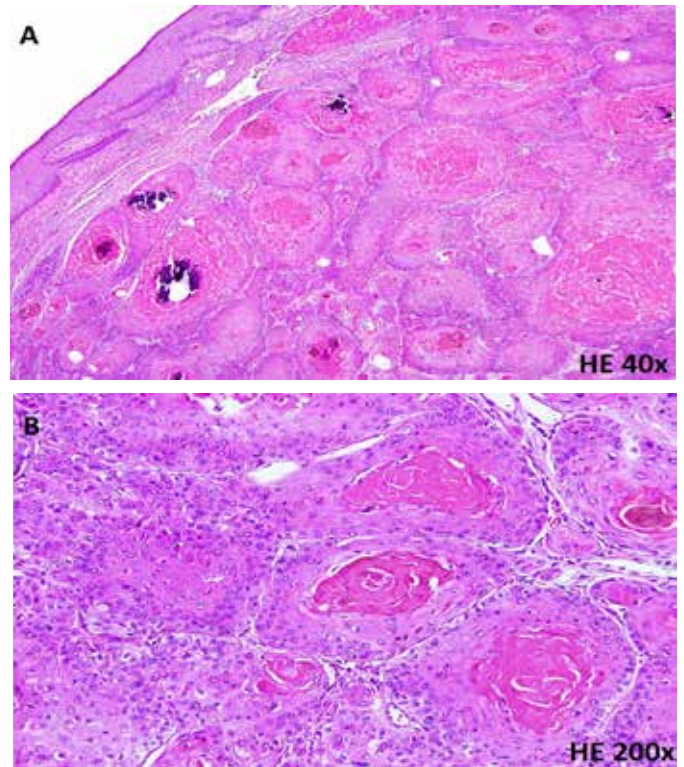


FIGURE 4: (A) Dermal tumor with multiple lobules varying in sizes, squamous cell proliferation with abrupt keratinization and foci of calcification. (B) Squamous epithelial lobules with discrete atypia associated with amorphous keratin mass and absence of inflammatory infiltrate

with ages ranging from 21 to 88 years, mean 62.4 years, despite reports in children under 18 years.^{1,3,5,10-12} Lesions are usually solitary nodules, smaller than 1 cm to 12 cm (average 3.3 cm), although there are citations of multiple nodules, sometimes ulcerated and larger than 25 cm.^{3,10-13} It often presents a history of slow and progressive growth from months to years, followed by fast growth and exophytic appearance.^{2,10,12} The preferred location is the scalp (85.4–90% of cases) and sometimes has alopecia.^{1,3,5,10-13} The remaining 10% are mostly on the back and more rarely elsewhere, such as frontal, nose, eyelids, lips, oral cavity, neck, trunk, genital, buttocks, upper and lower limbs, and skull base.^{1,3,5,10-12} The reported case manifested unusual features, such as the onset of PTT at 17 years, the presence of multiple nodules and some ulcerated lesions. Most of the PTT presents benign biological behavior, but with a chance of recurrence, in a period of six months to more than 10 years after surgery, especially in cases of cleft excision, and in very rare cases may have malignant biological potential.^{3,5,10-12} Local recurrence rates (3.7%) and lymph node metastasis (1.2%) are low, but when occur they may cause invasion into deep tissue planes by continuity, contiguity or spread, resulting in high

morbidity rates and even mortality.^{3,5,10-12} Some studies have shown the importance of researching Ki-67 immunoperoxidase, a protein responsible for cell proliferation, because the expression index correlates with the degree of malignancy of the tumor.^{7,8} The differential diagnosis of PTT includes squamous cell carcinoma, basal cell carcinoma, keratoacanthoma, pilomatricoma, sweat gland tumor, protuberant dermatofibrosarcoma, cylindroma, epidermoid cyst, malignant PTT, and angiosarcoma.^{1,4,11} Histopathological findings of squamous epithelium lobules, cells with abundant eosinophilic cytoplasm with abrupt keratinization, variable cytological atypia and absence of infiltration into adjacent stroma.^{1,4,12,13} PTTs are recurrent and potentially invasive, so, the therapy of choice is surgeries with a margins of 0.5 to 1 cm.^{5,7,9-12} Among surgical techniques, Mohs' micrographic surgery has a good indication, because its margins may to exceed visible clinical limits and thus reduce the risk of relapse and metastasis.^{5,9,11} Other treatments such as lymph node dissection, radiotherapy, and chemotherapy may be required in cases of metastatic dissemination or transformation to malignant PTT.^{5,11} Therefore, long-term follow-up is indispensable.^{7,10-12} ●

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Digital myxoid cyst: treatment by conservative compressive technique

Tratamento do cisto mucoso digital por técnica compressiva conservadora

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ABSTRACT

The digital myxoid cyst is a benign lesion of controversial origin, usually in the distal phalanx, close to the posterior nail fold. Clinical picture or imaging techniques, such as ultrasound and magnetic resonance imaging, can help diagnose the condition. It may present with discomfort to patients because it is often painful. There are several possible treatments, such as surgical excision and suturing, with or without grafting, cryotherapy, electrocoagulation, and curettage. However, it presents high recurrence rates. In this paper, we propose a simple, fast, and low-cost method for treating digital myxoid cyst, so far with low recurrence rates.

Keywords: Ambulatory Surgical Procedures; Dermatology; Nails

RESUMO

Cisto mucoso digital é lesão benigna de origem controversa que, em geral, ocorre na falange distal, próximo à dobra ungueal posterior, podendo apresentar coloração azulada e onicodistrofia. O diagnóstico pode ser realizado pelo quadro clínico ou por técnicas de imagem, como ultrassonografia e ressonância magnética. Gera incômodo aos pacientes por ser, muitas vezes, doloroso. Com vários tratamentos possíveis, tais como excisão cirúrgica e sutura, com ou sem enxerto, crioterapia, eletrocoagulação e curetagem, entre outros, apresenta altas taxas de recidiva. Neste artigo propomos método simples para tratamento do cisto mucoso, até o momento, com baixas taxas de recidiva.

Palavras-chave: Doenças da Unha; Procedimentos Cirúrgicos Ambulatoriais; Terapêutica

INTRODUCTION

Myxoid cyst is a benign tumor whose origin is still controversial. It may be caused by minor trauma that degenerates the connective tissue into mucoid material or would derive directly from the adjacent interphalangeal joint, damaged by osteoarthritis osteophytes, leading to the formation of a 5–20 mm in diameter rounded nodule. The cysts affect the fingers of the hands and feet, but especially the hands, and usually occur near the distal joints of the fingers.¹

The cysts can be painful and sometimes, when periungual, can produce groove or structural changes in the nail lamina that are reversible with treatment, which is evidence of the benignity of the lesion, since malignant lesions usually produce permanent onychodystrophy, due to the invasion of the matrix and not just compression.¹

The diagnosis is made by high-resolution ultrasound or magnetic resonance imaging, but can be made by transillumination with a small flashlight or by puncturing with a large needle, revealing the transparent mucous material inside.

The histologic exam presents a focal deposition of mucin in the dermis with surrounding fibrous stroma and no epithelial lining; mucin can be highlighted by the alcian blue and colloidal iron stains.

Several treatments are described in the literature^{2,3}, such as electrocoagulation and cryotherapy⁴, both with significant relapse rates and even surgery⁵, with higher cure rates, which is the first choice especially under the matrix¹. In this article, we will demonstrate a simple, safe and that has shown very satisfactory results technique.

METHODS AND RESULTS

After local asepsis, the cyst contents are emptied through a needle (40x1.2 mm/18 G11/ 2"). Digital nerve block may be performed, but the procedure usually generates little discomfort to the patient.

Depending on the diameter of the cyst (Image 1), the size of the syringe plunger rubber to be used should be chosen.

The rubber should be placed above the emptied cyst (Image 2) and with the aid of micropore fixed by compression, thus collapsing the walls of the cyst together (Image 3).

The secret to the success of the method is to tell the patient not to remove this bandage for at least 15 days, as this will allow the cyst to "heal", leading to the definitive collapse of the walls, preventing recurrence (Image 4).



FIGURE 2: Rubber embedded in the already emptied cyst



FIGURE 3: Micropore tape securing and compressing the cyst walls to one another



FIGURE 1: Assessment of the cyst diameter to choose the size of the rubber syringe plunger to be used

During a six months follow up, there were not relapses, except in those who lost the dressings early, not reaching the recommended time of 15 days. In these cases, relapse occurs in a matter of days.

DISCUSSION

In the treatment of the mucous cyst, we had seen recurrences even after procedures such as removal by saucerization followed by electrocoagulation of the internal base of the cyst, close technique cryotherapy for 45 to 60 seconds with a probe compressing the cyst walls, both techniques with high morbidity rate; or even surgical excision of the cyst, whether or not followed by local graft.



FIGURE 4: Final result with the final collapse of the digital mucous cyst

With higher recurrence rates for the first two methods and for simple excision and slightly lower for cases in which total skin grafting was applied under a Brown dressing.

This is evidence that optimal treatment does not depend

on tissue destruction, but rather on effective and lasting local compression that allows the inner walls of the cyst to reestablish and recap.

Despite the compression applied for a period of 15 days, we have not yet witnessed local necrosis. Even if this happens, it should not be something to fear, as the recommended procedures for treatment of myxoid cysts, such as electrocoagulation or cryotherapy, generate local necrosis.


CONCLUSION

Myxoid cyst is a benign lesion that should be treated conservatively.⁵ The method described above should be used because it is a simple, fast, inexpensive, and easy procedure, with no major complications observed so far, and can be performed in the office during the initial consultation of the patient. As a disadvantage, we point out the inconvenience that it is necessary for the patient to remain with the dressing for an extended period, which, if not performed, may impair the effectiveness of the method. ●

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Cuniculatum carcinoma: HPV 16 positive lesion with an abdominal graft surgical reconstruction.

Carcinoma cuniculatum: lesão positiva para HPV 16 com reconstrução cirúrgica com enxerto abdominal

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ABSTRACT

The cuniculatum carcinoma is a rare, slow-growth, local aggressive subtype of the squamous cell carcinoma, presented as a verrucous lesion with a low metastasis capacity. Its histopathology shows keratin-filled crypts, and its etiology is linked with HPV, especially 11 and 16 subtypes. The treatment of choice is surgical excision, followed by regular monitoring due to the high risk of local recurrence. We report a case of a woman with Cuniculatum carcinoma treated with surgery with perioperative frozen section procedure, graft, and good evolution.

Keywords: Carcinoma; Carcinoma, Squamous Cell; Foot

RESUMO

O carcinoma cuniculatum consiste em um subtipo raro do carcinoma espinocelular, que se apresenta como uma lesão verrucosa, de crescimento lento, agressividade local e baixa capacidade de produzir metástases. Sua histopatologia revela criptas preenchidas por queratina, e sua etiologia tem ligação com o HPV, principalmente os subtipos 11 e 16. O tratamento de escolha é a excisão cirúrgica, seguida de acompanhamento regular pelo elevado risco de recidiva local. Relata-se caso de paciente feminina, portadora de carcinoma cuniculatum, tratada pela cirurgia com congelamento perioperatório, realização de enxerto e boa evolução.

Palavras-chave: Carcinoma; Carcinoma de Células Escamosas; Pé

INTRODUCTION

Squamous cell carcinoma is the second most common type of skin cancer, behind only basal cell carcinoma.¹ Carcinoma cuniculatum (CC) is a verrucous carcinoma (VC), a rare and indolent form of squamous cell carcinoma (SCC), well differentiated. Slow growth, local aggressiveness, and low capacity to cause metastasis characterize it. It penetrates the deep tissues, forming crypts filled with keratin.^{2,3}

It was first described in 1954 by Aird *et al.*^{1,5} It is more common in men than in women (5:1). Its pathogenesis has not been completely elucidated, but the association with human papillomavirus (HPV), especially subtypes 11 and 16, has been described.^{5,6}

The treatment of choice is surgical excision with margin control, and the patient must be regularly monitored for the risk of recurrences, even when the surgical margins are free.⁶

Case report

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CASE REPORT

A 64-year-old woman, working as a sales clerk, sought medical care due to a lesion in the left plantar region two years ago, with a foul odor and associated local pain. The physical examination observed an exophytic lesion with a verrucous surface, 5 cm in diameter, with an infiltrated and ulcerated border (Figures 1 and 2). The image examination, X-ray of the left foot, showed a nodular image with a radiotransparent halo in soft tissues in the plantar region, without bone involvement (Figure 3).

A biopsy was performed based on the suggested hypotheses of VC, keratoacanthoma, common wart, and amelanotic melanoma. The result favored the diagnosis of VC. Excisional



FIGURE 3: X-ray of the left foot
Nodular image with radiotransparent halo in soft tissues in the plantar region



FIGURE 1: Exophytic lesion with verrucous surface, with infiltrated and ulcerated border in the left plantar region



FIGURE 2: Approximate image of the left plantar lesion

surgery was then scheduled with histopathological evaluation by perioperative frozen section. The report indicated a well-differentiated SCC, focally invasive, reaching the middle dermis, with no perineural infiltration or angiolymphatic invasion, and free surgical margins (Figures 4 and 5).

After the lesion's complete excision, skin grafting was obtained from the abdominopelvic fold (Figure 6), and a fragment was sent for HPV research, with a positive result for type 16.

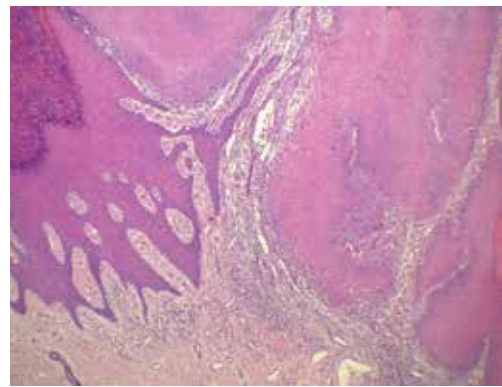


FIGURE 4:
Histopathological examination

Well-differentiated SCC, focally invasive, reaching the middle dermis (Hematoxylin & eosin 100x)

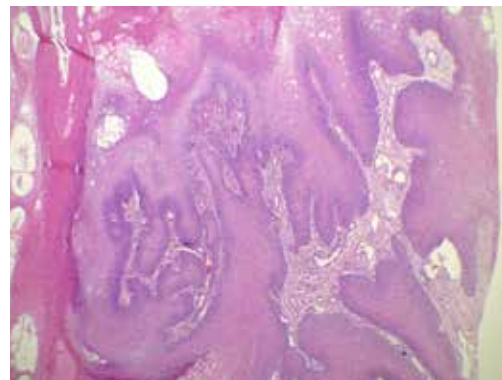


FIGURE 5:
Histopathological examination

Squamous cell proliferation with papillomatosis and dyskeratosis, high degree of cell differentiation, without atypias (Hematoxylin & eosin 100x)



FIGURE 6:
Immediate post
surgery with
abdominal graft

The patient evolved with good graft healing and subsequent return to daily activities. After 1.5 years of follow-up, we observed no lesion recurrence.

DISCUSSION

CC is a VC, a rare and indolent form of SCC. It usually has characteristics of a benign, verrucous tumor, with slow growth, more expansive than invasive evolutionary pattern, good cell differentiation, and low tendency to metastasis.^{2,7}

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Although its most common site is in the plantar region, it can occur in several other areas, such as the flank, legs, face, oral cavity, genital, and palmar region.⁴ It is called CC when it is located in the plantar area; Buschke-Lowenstein tumor, in the anogenital region; and Ackerman's tumor, when in the oral region.^{3,8}

It was first described in 1954 by Aird *et al.*^{1,5} It is more common in men than in women (5:1)⁵ – what makes the report of this case even rarer because it is a female patient. Its pathogenesis is unknown, but the association with human papillomavirus (HPV), especially subtypes 11 and 16, has been described,⁶ as identified in the case above.

Diagnosis is based on dermatological and histopathological examination.^{5,6} This carcinoma should be suspected when the patient has a recurrent lesion on usual therapy. The differential diagnosis is made with common wart and keratoacanthoma.^{6,9}

We observed scaly proliferation with papillomatosis and dyskeratosis in histopathology, a high degree of cell differentiation, without atypia. Keratin pseudocysts can be formed, sometimes with pus, generating a foul odor.³

The treatment of choice is surgical excision with margin control, and the patient must be regularly monitored for the risk of recurrences, even when free margins are obtained.^{6,7} In the case reported, we opted for surgery with a perioperative frozen section to control the margins and perform a graft on the patient's abdomen, with Brown dressing. The patient has been in follow-up for more than two years without recurrence so far. ●

Case report

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Squamous Cell Carcinoma excision and upper lip reconstruction with double advancement technique

Exérese de carcinoma espinocelular e reconstrução de lábio superior com técnica de duplo avanço

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ABSTRACT

Although several techniques have been described for upper lip reconstruction, functional reconstruction of total upper lip defects remains a challenge. We report a case of a significant size squamous cell carcinoma excised in the upper lip region using the double-advancement technique, with positive functional and aesthetic results.

Keywords: Carcinoma, Squamous Cell; Lip Neoplasms; Surgical Flaps

RESUMO

Embora várias técnicas tenham sido descritas para a reconstrução do lábio superior, a reconstrução funcional dos defeitos totais do lábio superior permanece um desafio. Neste caso, realizou-se a exérese de um carcinoma espinocelular de tamanho significativo, em região labial superior, por meio da técnica de duplo avanço, com resultados funcional e estético positivos.

Palavras-chave: Carcinoma de Células Escamosas; Neoplasias Labiais; Retalhos Cirúrgicos

INTRODUCTION

Over the years, several techniques for lip reconstruction with different levels of complexity have been developed, given the organ's peculiarity and functions. Each case is different from the other, with each patient's characteristics, anatomy, sex, comorbidities, and smoking. It is essential to consider the size of the lesion and its location in the different labial subunits. The main objectives are to maintain speech ability, adequate nutrition, and symmetry and aesthetics, since it is located on the face and directly related to its personal image.

This report aims to show the approach on a large and deforming lesion with a surgical technique that preserved the patient's lip and function.

CASE REPORT

A 59-year-old man, without pathological history, in his first dermatological consultation, with a history of a lesion on the upper lip with five months of evolution, presented difficulty in speech and suction movement. The clinical examination observed a tumor with raised erythematous borders, an ulcerated keratotic center with whitish areas, and a hyperchromic center in the upper lip's medial region measuring more than a third of the upper lip (Figure 1). Dermoscopy was limited by the keratotic component of the lesion, with few structures being observed. The rest of the facial skin did not have photodamage or other lesions suspected of malignancy.

Incisional biopsy and anatomopathological examination of the fragment were performed, describing follicular comedones and chronic inflammation. Due to the exuberant and rapidly progressing lesion, we opted for complete excision of the lesion and a new anatomopathological exam.

We performed total excision of the lesion with a 6 mm surgical limit and correction of the defect with bilateral advancement flap in the lateral subunits of the upper lip (Figures 2 and 3), with satisfactory results both aesthetically and functionally, preserving blood supply through the upper labial arteries, mucosa, and the orbicularis oris muscle of the mouth.

The result of the anatomopathological examination of the specimen with surgical limits was squamous cell carcinoma with peripheral and deep surgical limits free of neoplasia. The patient returned with an operative wound with good healing and satisfied with the preservation of the functionality of the lips (Figures 4 and 5).



FIGURE 1: Pre-surgical double advancement marking



FIGURE 2: Transoperative post-tumor excision



FIGURE 3: Demonstration of surgical plan

DISCUSSION

Techniques for upper lip reconstruction are poorly described in the literature since tumors in this region are uncommon, with only 5% of lip tumors, and the most common histological type is squamous cell carcinoma, as in the case.^{1,2}

The lip is divided into four subunits with two laterals and one medial at the top, the filter, and a single subunit at the bottom. The patient's lesion was in the medial subunit, progressing to the left lateral subunit. There is no mandatory technique for reconstructing the upper lip. Professionals must analyze the size and location of each defect and know its anatomical struc-



FIGURE 4: Seven-day postoperative



FIGURE 5: 45-days postoperative

ture. Thus, its division into units facilitates the reconstruction plan.³

With the flap with medial advancement of the cheeks, we managed to maintain tissue perfusion through the upper labial artery's blood supply and the perforating arteries subcutaneously in the pedicle, preventing necrosis.⁴ As neighborhood skin is used, it is possible to maintain texture, hair, and skin color.

As we performed the excision of part of the vermilion lip, we also promoted the vermilion mucosa's advancement to maintain the aesthetics and anatomical functionality.⁴ Small cutaneous branches of the infraorbital nerves are cut during the surgery, leaving the skin with reduced sensitivity, which is usually recovered spontaneously in the postoperative period.⁵

An option for surgery would be the renowned Abbe flap, described in the literature and performed more than 100 years ago. Nevertheless, we chose to perform a bilateral advancement flap to perform the surgery in just one surgical time, avoiding the need for patient collaboration to care for the surgical wound and the risks of being submitted to another surgical time.⁵

CONCLUSION

For upper lip defect correction, advancement flaps are a good option with satisfactory functional and aesthetic results. ●


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Case report

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Onychomatricoma, an ignored diagnosis: Case Report

Onicomatricoma, um diagnóstico ignorado: Case report

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ABSTRACT

Onychomatricoma is a rare tumor of the nail complex of unknown etiology. It has a higher incidence in women in the 5th decade of life. However, this occurrence may be mistaken due to data and information that have not yet been fully elucidated. This study aims to review the literature and report two cases of onychomatricoma. We included two male patients with different age ranges and different tumor presentations. This study shows that the rarity and incidence reported in the literature are questionable and that the onychomatricoma can be underdiagnosed.

Keywords: Dermoscopy; Onychomycosis; Pathology, Clinical

RESUMO

O onicomatricoma é um tumor raro do complexo ungueal, de etiologia desconhecida. Apresenta-se com maior incidência em mulheres na 5ª década de vida. Contudo, é possível que esta ocorrência esteja equivocada devido a dados e informações que ainda não foram seguramente esclarecidos. O propósito do presente estudo é fazer uma revisão de literatura e relatar dois casos de onicomatricoma. Foram incluídos dois pacientes do sexo masculino, com faixa etária distinta e apresentações diferentes do tumor. O entendimento deste trabalho evidencia que a raridade e a incidência relatadas na literatura são questionáveis, e que o onicomatricoma pode ser subdiagnosticado.

Palavras-chave: Dermoscopia; Onicomicosse; Patologia Clínica

INTRODUCTION

The onychomatricoma is a rare and benign tumor of the nail complex, first described in 1992 by Baran and Kint. The classic tetrad characterizes it: xanthonychia, subungual hyperkeratosis, splinter hemorrhages affecting the nail plate, longitudinal and transverse hypercurvature of the nail plate.^{1,2,3,4,5,6} Thus, underdiagnosed conditions, confusion with specific differential diagnoses, or even the lack of knowledge about the tumor by some dermatologists justify the rarity of this condition.

Its etiology is still unknown.^{5,6,7} However, recent studies are in a continuous investigation for this motivation, assessing genetic and environmental conditions, risk factors, profession, history of trauma, domestic and/or aesthetic habits, and even associated diseases, suggesting new hypotheses for this tumor's

cause.

The onychomatricoma affects mainly women, with a higher occurrence in the 5th decade of life.^{1,5,6,7,8} However, the present reported cases are of men, aged 42 and 64 years, respectively. This incidence may be wrong given specific data and information that have not yet been correctly clarified.

Most studies have related onychomycosis as a confounding factor.^{1,3,5,6} Thus, many patients are initially treated with antifungal drugs for years. It is possibly a justification for late diagnoses and, also, for underdiagnosed cases, reducing the percentage of reported cases and, consequently, the incidence rate, inducing to believe in the disease's rarity.

Clinical characteristics can suggest the diagnosis, and it is more easily evidenced when it appears classically, grouped in a tetrad.

Complementary diagnostic methods guide the diagnosis, such as dermoscopy, ultrasound, and anatomopathological study.^{1,2,5,6,7} Other more recent exams present innovative ways for diagnostic purposes, as pointed out in some reports, expanding the possibilities for unveiling new cases of this tumor.

Treatment is complete local excision performed as a curative mean.^{1,5,6} Concomitantly with therapy, the anatomopathological study of the skin fragment is conducted to clarify the disease. There is a possibility to develop nail dystrophy after the surgical procedure, making this explanation necessary to the patient before the surgical approach. Therefore, it is plausible that some patients refuse to undergo surgery due to this disorder, reducing the number of confirmed cases.

This study aims to conduct a literature review with to expand the knowledge of dermatologists on the tumor, demonstrating various techniques to determine the disease, and reporting two cases of onychomatricoma with unequal presentations in men with different age groups, who obtained their diagnoses by different methods.

METHODS

Two men with onychomatricoma, with different age groups, and different tumor presentation locations, were included. They obtained their diagnosis by the tumor's clinical manifestation; however, the complementary methods to determine the diagnosis were unequal. Case 1 presented with nail onychodystrophy in the left hallux. After performing an anatomopathological examination, it showed compact hyperkeratosis, agranulose and acanthosis compatible with onychomatricoma. In the second case, the third finger's dermoscopy revealed splinter hemorrhage, xanthonychia, subungual hyperkeratosis, and longitudinal hypercurvature of the nail plate, which constituted the classic tetrad. Still, radiography of the right hand was performed, showing no bone changes, and corroborating the diagnosis of neoplasia.

CASE REPORT

Case 1

A 42-year-old man, skin phototype V, military, with hypercholesterolemia, presented at the Dermatology Service to be

followed up for lichenoid dermatitis. The clinical examination observed no lesions suggestive of the pathology that motivated the medical consultation. However, onychodystrophy (Figure 1) and subungual hyperkeratosis of the left hallux were found.

The patient mentioned previous trauma to the same toe ten years ago and reported mild pain when putting on his shoes, without other symptoms. He also explained that he had performed direct mycological examination and nail plate culture seven years ago, with a positive result for *Candida* sp, with topical antifungals being prescribed, with no success.

We performed radiography of the left foot, which did not reveal bone involvement. Based on this data, the possibility of a benign tumor of the nail complex was suspected. Then, we decided to perform an excisional biopsy (Figure 2), sending the material to the pathologist to confirm the diagnosis of onychomatricoma.

Case 2

A 64-year-old man, skin phototype III, driver, with benign prostatic hyperplasia, systemic arterial hypertension, and peripheral arterial disease, presented to the Dermatology Service to assess an asymptomatic lesion located in the 3rd right finger, which appeared ten years ago, with a history of previous trauma.

He reported previous empirical topical treatments for onychomycosis, with no success. The clinical examination found subungual hyperkeratosis and longitudinal hypercurvature on the nail plate.

Dermoscopy observed the presence of xanthonychia, splinter hemorrhages, subungual hyperkeratosis, and longitudinal hypercurvature on the nail plate (Figure 3), suggesting the clinical and dermoscopic diagnosis of onychomatricoma (Figure



FIGURE 1: Onychodystrophy of the left hallux

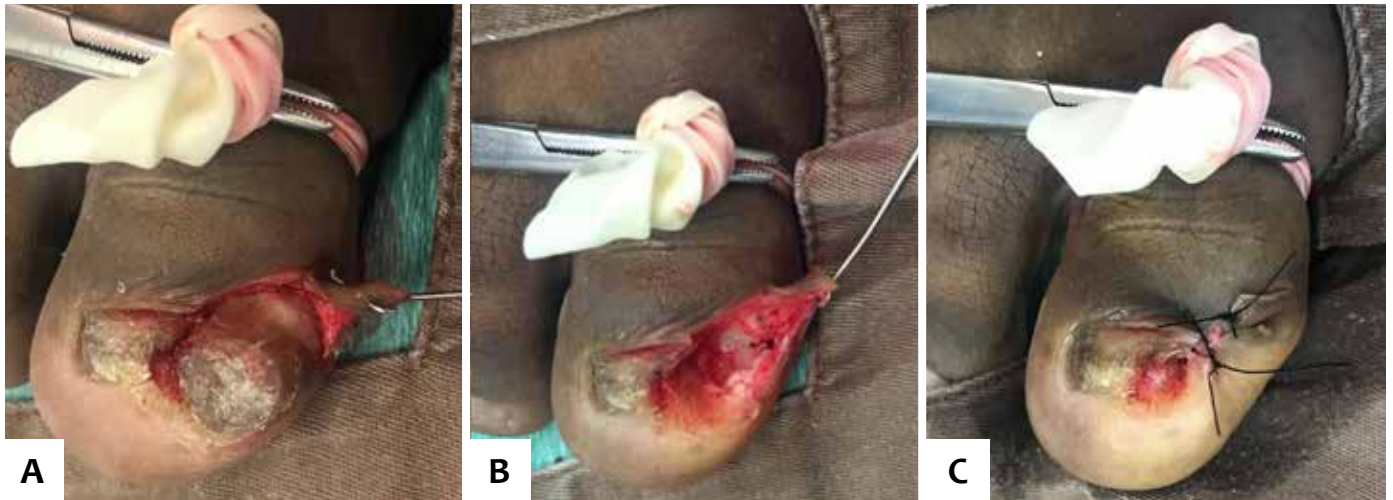


FIGURE 2: Excisional biopsy of the left hallux onychomatricoma: a) tumor seen during the operation, b) removal of the lesion, c) closure after surgical exploration

4). Radiography and computed tomography of the right hand were also performed, showing no bone involvement, and corroborating the tumor condition's determination.

Subsequently, we performed the histopathological study by excisional biopsy (Figure 5), revealing in the microscopic examination discrete projections of the epidermal cones in the superficial dermis amid hyalinized collagen, mild solar elastosis, and vasodilation.

Altogether, the clinic, dermoscopy, imaging exams, and anatomopathological study help define the condition as onychomatricoma.

DISCUSSION

First described in 1992 by Baran and Kint,^{1,2,3,4,5,6,9} onychomatricoma is a rare and benign fibroepithelial neoplasm of the nail complex.^{1,2,3,4,5,6,7,9,10,11} The plurality of studies points to the nail matrix as the original site of the tumor. However, Mello et al. suggest its beginning in cell differentiation areas with matrix cells (metaplasia), which may be the portions of the proximal fold or the nail bed. Therefore, the hesitation in determining this neoplasm's genesis also implies a lack of knowledge on the subject.

The onychomatricoma so far presents uncertain etiology.^{1,3,4} However, predisposing factors have been documented, such as onychomycosis and a history of previous trauma.^{1,5,6} In contrast, the study by Kallis and Tosti states that the tumor is the causative factor of onychomycosis due to the cavitations formed by the digitiform projections inside the nail plate, making the environment favorable for the invasion of fungi.

Nail mycosis is an extremely relevant point when addressing the onychomatricoma theme. Onychomycosis is reported from different perspectives: as a pathology caused by the tumor; as a precursor agent; as a disturbing factor for diagnosis; and, also, a complicating factor of the neoplasia.



FIGURE 3: Subungual hyperkeratosis and longitudinal hypercurvature of the nail plate of the third right finger are observed

Due to similar clinical and dermoscopic characteristics, the tumor is possibly initially diagnosed as onychomycosis and often treated as such,^{1,2,4,7} delaying its diagnosis and complicating the onychomatricoma condition. It is possible that, in some cases, the pathologies can indeed coexist, which contributes even more to the percentage of underdiagnosed cases of the tumor.

Some current studies (Table 1) describe trauma cases before the onychomatricoma, both accidental due to crushing⁶ or

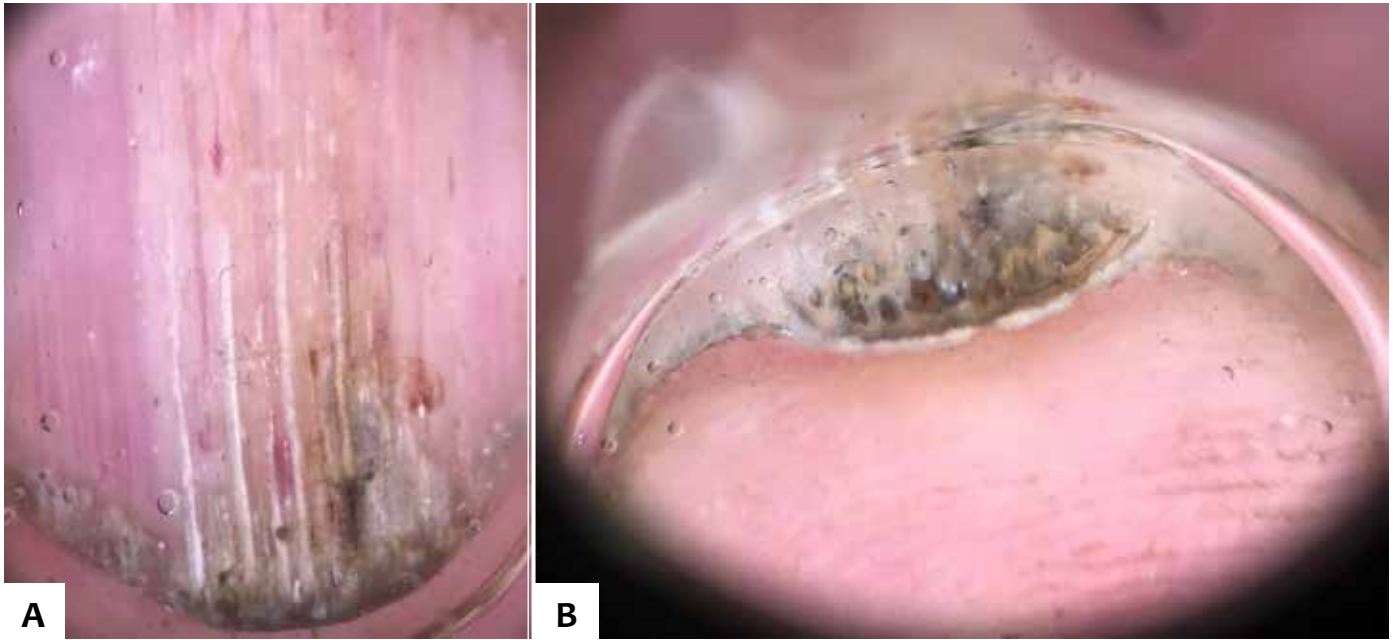


FIGURE 4: Dermoscopy: **A)** Xanthonychia and splinter hemorrhages are identified, **B)** Subungual hyperkeratosis, longitudinal hypercurvature of the nail plate and cavitations of the third right finger



FIGURE 5: Excisional biopsy of the third right finger: **A)** removal of the onychomatricoma, **B)** visualization of the intraoperative tumor, **C)** closure after surgical procedure

even due to small and repeated bruises, as in the case of a tool-maker.³ Such information allows us to conclude that the type of trauma is separated from the tumor's occurrence and that the profession can be considered a risk factor.

In the present study, there was a history of previous trauma ten years ago in both cases. However, in other studies, patients deny previous trauma. There is also a single report of a four-year-old girl with onychomatricoma but without a history of pre-existing trauma,⁶ so it is hypothesized that this patient was predisposed due to onychomycosis.

Predisposing factors must be continually evaluated during anamnesis to collect a greater amount of data for medi-

cal research purposes, correctly defining information about the tumor.

The tumor is more prevalent in Caucasian women, with a peak incidence in the 5th decade of life,^{1,3,4,5,6,7,9} although some studies indicate that the development of the onychomatricoma has no preference for sex.^{7,10,12}

In the reports of this study, both patients are men aged 42 and 64 years, respectively. The patient in case 1 has skin phototype V, making it another different data for the research statistics.

Most of this information may be mistaken, as analyzed in Table 1. This mistake probably occurs due to socio-cultural and environmental factors, such as aesthetics, care for personal health,

TABLE 1: Epidemiological data on onychomatricoma in the reference literature

LITERATURE	SEX	AGE	SITE	ONYCHOMYCOSIS	TRAUMA	“COLOR OR RACE”	TIME TO DIAGNOSIS
Rushing <i>et al</i> [5]	Men	66y	Left hallux	-	No	White	4 years
Kallis <i>et al</i> [3]	Men	35y	Right thumb	Yes	No	White	2 meses
Kallis <i>et al</i> [3]	Men	47y	Left Hallux	Yes	No	White	2 years
Mello <i>et al</i> [7]	Men	36y	Left Hallux	Yes	Yes	White	4 years
Tavares <i>et al</i> [6]	Woman	61y	Left Hallux	No	No	White	30 years
Joo <i>et al</i> [2]	Woman	27y	Right index finger	-	No	White	8 years
Kamath <i>et al</i> [4]	Men	60y	Left Hallux	Yes	No	Brow	2 years
Madi <i>et al</i> [8]	Men	23y	Right thumb	No	Yes	White	1 years
Zou <i>et al</i> [14]	Woman	31y	Right index finger	No	No	White	4 years
Charfi <i>et al.</i> [9]	Woman	46y	Hálux direito	-	No	White	2 years
Park <i>et al</i> [11]	Men	47y	Hálux direito	Yes	No	White	5 years
Tambe <i>et al</i> [12]	Woman	50y	Right ring finger	No	No	White	6 years
Kamath <i>et al</i> [4]	Men	73y	Hálux esquerdo	-	Yes	White	10 years
Kamath <i>et al</i> [4]	Men	53y	Right third toe	-	Yes	Brow	1 year
Kamath <i>et al</i> [4]	Woman	80y	Left Hallux	-	Yes	Brow	2 years
Kamath <i>et al</i> [4]	Men	59y	Right little finger	-	Yes	Brow	2 years
Kamath <i>et al</i> [4]	Woman	45y	Right little finger	Yes	Yes	Brow	3 years
Kamath <i>et al</i> [4]	Men	60y	Left Hallux	-	Yes	Yellow	20 years

profession, and even underdiagnosed cases or cases mistakenly treated as onychomycosis.

Recent information exposes that fingers are more commonly affected than toes, with 63% and 36% prevalence, respectively, and the disease may affect only a single finger or simultaneously.^{3,7,9,10} Nevertheless, diagnostic errors are common due to confusion with nail mycosis, and the true incidence in the lower extremity remains to be determined.

As shown in Table 1, it is possible to assess a disagreement between the literature's statistical data and the present reports' information. For this reason, the reported uncertainty may reveal to us how much more research is still needed.

The tumor extends digitiform projections of the matrix that penetrate the “nail plate”, causing thickening of the nail plate, xanthonychia, splinter hemorrhage, transverse and longitudinal hypercurvature, characterizing the clinical tetrad.^{1,2,4,5,6,7,9,10,11,12}

Other findings may be present, including longitudinal groove, subungual hematoma, verrucosity of the proximal nail fold, erythema and edema of the proximal fold, longitudinal melanonychia, onychodystrophy, and dorsal pterygium.^{3,4,5}

The onychomatricoma contains several differential diagnoses, such as onychomycosis, which represents 50% of nail disorders; nail and periungual tumors: digital fibrokeratoma and subungual fibroma; Bowen's disease; common subungual wart;

osteochondroma; squamous cell carcinoma; longitudinal melanonychia, and subungual exostosis.^{1,2,4,6,7,12}

Fibrokeratoma and nail fibroma are considered the primary tumors for differential diagnoses. In the longitudinal sections of the onychomatricoma, the lesion resembles the first, and the stroma located in the lunula can also suggest the second. However, the presence of digitiform projections excludes these diagnoses.⁶

The diagnosis is based on the classical tetrad signs and additional methods, such as dermoscopy, ultrasound, and histopathology, the latter being considered the gold standard.^{1,2,3,4,5,7,9} Other studies include, as complementary exams, magnetic resonance imaging, radiography, nail clipping, and confocal microscopy.^{1,4,5,9,10}

After surgical exploration, there is a possibility of permanent nail dystrophy. Therefore, this probable event should be informed to the patient and communicate that the tumor has an indolent nature and without any malignant potential, being possible to maintain only clinical follow-up if it is asymptomatic. Thus, the patient can develop an understanding to evaluate the need for surgery together with the dermatologist.

Given the above, some patients choose not to perform the procedure. Countless cases may be ignored and not documented, contributing to reducing the percentage of its incidence.

It is possible to see perforations in the distal portion of the nail plate, hemorrhagic streaks, and longitudinal white grooves corresponding to the nail plate's channels in dermoscopy.^{1,2,4,7,9,10}

X-ray examination shows no underlying bone involvement linked to the onychomatricoma.^{3,7,10} In the present reported cases, the radiography did not reveal erosion or bone remodeling, reinforcing the suspected diagnosis of the tumor.

The ultrasound examination reveals a hypoechoic tumor lesion affecting the nail matrix and a hyperechogenic area corresponding to the digitiform projections.^{1,4,7,9,10}

Magnetic resonance imaging points to the nail matrix with low signal uptake, while the distal digitiform projections show high uptake.^{1,4,9,10} This imaging method should be considered for dystrophic nails with negative mycological tests. So far, there are no other lesions reported in the literature that have the same presentation as this tumor, becoming specific.⁹

Another possible exam to be performed is nail clipping, which corresponds to the plate's distal cut, studied histologically, revealing peculiar characteristics such as thickening of the nail blade, cavitations filled with serous material, and periphery with a thin layer of epithelium.⁴ Nail clipping presents itself as an easy and minimally invasive resource, aiding in the diagnosis, and excluding associated fungal infections.

Confocal microscopy was used in four cases, and it seemed useful in the preoperative diagnosis of the onychomatricoma, thus being another available method.⁹

Histologically, the onychomatricoma is a fibroepithelial tumor comprising two different zones: the proximal and the distal. The first, located below the posterior nail fold, is characterized by having deep epithelial invaginations occupied by overlapping nail protrusions, in the shape of V. The second, still a distal zone, corresponding to the lunula, contains epithelial originating from the matrix epithelium, which proliferates and causes perforations in the nail plate.^{1,3,6,7,10}

Treatment consists of complete surgical excision, including the entire proximal matrix, to avoid local recurrence.^{1,4,5,6,7,10,12} The long-term prognosis seems favorable since there is only one case of recurrence reported so far.

After anesthesia, an avulsion of the nail plate is performed, allowing the visualization of the tumor projections that will be removed.^{4,7} For symptomatic tumors, tangential excision is preferable to avoid onychodystrophy. However, there is a risk of incomplete removal.⁶ It is plausible that this sequelae will be permanent if the tumor is larger than 3mm.¹³

Mohs surgery allows tumor distancing with minimal removal of the affected nail matrix and, thus, minimizes the potential aesthetic and functional impacts of the surgery.¹⁰ It is possible that, in the future, this technique will be performed in most cases, encouraging greater medical demand for both diagnosis and treatment.

CONCLUSION

In short, the onychomatricoma remains a rare tumor, but statistical data on its epidemiology is doubtful.

Its tendency to be asymptomatic and the misdiagnosis of onychomycosis favor diagnostic difficulties. There is a need to raise the rate of clinical suspicion when a single nail is affected or if topical antifungals do not achieve therapeutic success, or even if there is a history of trauma.

The histopathological examination must confirm the diagnosis when the classical tetrad does not reveal the tumor. However, the variety of complementary tests currently used facilitates diagnostic suspicion. It is essential to report that more recent methods are being introduced, minimizing the aesthetic impact and contributing to patients' acceptance of research.

The gold standard for treatment is complete surgical excision. However, knowledge of appropriate surgical techniques is essential to prevent tumor recurrence and possible onychodystrophy.

We reported two cases of onychomatricoma to expand the disease's knowledge; after all, it is still a diagnostic challenge.

This study allowed a new question: is the diagnostic difficulty due to the rarity of the onychomatricoma, or is it due to the dermatologists' insufficient knowledge? ●

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Surgical approach to Köenen tumor: a case report and literature review

Abordagem cirúrgica do tumor de Köenen: relato de um caso e revisão da literatura

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ABSTRACT

Koenen tumors are nail fibromas included among the main diagnostic criteria of tuberous sclerosis. Elongated and thin papules, firm, with smooth surface and broad base characterize these lesions, which are slightly erythematous and present varying sizes. Koenen tumors commonly affect toenails and result in aesthetic and functional concerns associated with deformities, pain, and bleeding. Several therapeutic options are described for Koenen tumors; however, there is a lack of consensus in the literature regarding standard treatment. We report the case of a patient who underwent surgery with excellent outcomes. Additionally, we discuss the advantages and disadvantages of each therapeutic modality.

Keywords: Dermatologic Surgical Procedures; Fibroma; Nails; Tuberous Sclerosis

RESUMO

Os tumores de Köenen são fibromas ungueais que constituem um dos principais critérios diagnósticos da esclerose tuberosa. Caracterizam-se por pápulas alongadas e afiladas, firmes, de superfície lisa, base alargada, levemente eritematosas e de tamanhos variados, mais comuns nas unhas dos pododáctilos. As lesões são consideradas um problema estético e funcional, podendo ocasionar deformidades, dor e sangramento. Quanto ao tratamento, existem diversas opções, entretanto não há consenso na literatura sobre a melhor delas. Relatamos um caso tratado cirurgicamente, com ótimo resultado, e descrevemos as vantagens e desvantagens de cada modalidade terapêutica atual.

Palavras-chave: Esclerose Tuberosa; Fibroma; Procedimentos Cirúrgicos Dermatológicos; Unhas

INTRODUCTION

Also known as Bourneville-Pringle epiloia or phacomatosis, tuberous sclerosis (TS) is a rare, multisystemic neurocutaneous disorder that can develop with hamartomas of skin, central nervous system, kidneys, and lungs.¹

In the context of this autosomal dominant disease, it is known that half of the affected families are linked to chromosome 9q34, with inactivating mutations of the tumor suppressor genes of the hamartin protein (TSC1), and the other half to chromosome 16p13. It causes inactivating mutations of genes tumor suppressors of tuberin protein (TSC2). The hamartin-tuberin complex is an essential tumor growth inhibitor, and its absence triggers the loss of inhibition on cell proliferation and migration.²

Among the most common skin changes are facial angiofibromas, fibrotic plaques (Shagreen patch), periungual fibromas (Koenen's tumors), hypochromic macules in the shape of leaves (ash leaves), and frontal fibrous plaque.^{1,2} Many patients have

Case report

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nail fibroma as the only cutaneous manifestation of the disease, so their presence in the dermatological physical examination should raise the disease's clinical suspicion.³

In addition to Koenen's tumor (KT), the literature describes other nail disorders in TS, such as subungual hyperkeratosis, reddish and whitish longitudinal leukonychia, hemorrhagic splinters, and longitudinal grooves. It is believed that the presence of reddish and/or whitish longitudinal ridges, in addition to nail fibroma, greatly increases the suspicion of TS.⁴

Other significant findings include seizures, intellectual disability, subependymal nodules, retinal, and other organ hamartomas.² Although surgical excision is preferable for nail fibroma,⁵ other therapies are available. However, few studies compare the efficacy and disadvantages of different therapeutic modalities.

CASE REPORT

A 38-year-old woman presented progressive nail growths on her fingernails and toenails since the second decade of life. It caused local bleeding after minimal trauma, pain, and deformities, making it impossible to wear closed shoes and manipulate objects. She denied comorbidities and reported a father with TS.

The nail lesions presented as elongated and tapered papules, firm, with a smooth surface, broad base, slightly erythematous, and varied sizes, in all toes and some fingers (Figures 1-3). Regarding location, most had implantation in the proximal fold and a small part in the lateral folds. Some nail plates showed longitudinal lines and canalicular depressions of variable extension, with some thin whitish ridges (Figure 1). In the limbs, there were multiple hypochromic spots in the shape of a leaf (ash leaves), in addition to homochromatic lenticular lesions, confetti-like. On the face, we observed flat, normochromic, isolated, and coalescent papules, especially in the malar and dorsal nasal regions, suggesting angiofibromas. In the abdominal and dorsal area, she had hypochromic stains, measuring up to about 20 cm.

The anatomopathological examination of nail lesions was compatible with digital fibrokeratomas; on the face, with angiofibromas; and the hypochromic spots on the chest, with



FIGURE 1: Koenen's tumors on the nails of the left foot: before surgery. In nail plates, longitudinal lines and canalicular depressions are observed



FIGURE 2: Koenen's tumors on the nails of the left hand: before surgery



FIGURE 3: Koenen's tumors on the nails of the right hand: before surgery

achromatic nevus. Computed tomography (CT) of the skull showed dysplasias and cortical fibromas, without other changes. Therefore, given skin and nail lesions, complementary exams, and family history, we established the TS diagnosis.

We opted for surgical excision of nail lesions, removing four tumors in each surgical period. The nails of the hands and the left foot were approached. Fibromas located in the posterior fold were excised by folding the nail plate through two parallel incisions at the posterior and lateral folds' junction. Once folded, the origin of the tumor implantation was visualized, proceeding

to their detachment, with the aid of dental spatulas, until its total removal. In the postoperative period (Figure 4), the patient followed the guidelines regarding rest, cleaning, and dressing, progressing satisfactorily, with good aesthetic and functional results (Figures 5-7). The patient is followed up to continue the surgical treatment.

DISCUSSION

KTs are nail hamartomas mainly in the toes, more common in women, and appearing mostly after the second decade of life. They can progressively increase in size, compressing the nail matrix, and causing changes such as longitudinal nail depressions.¹ They are classified as peri or subungual, the first being the most common.⁶ It is a benign cutaneous manifestation of TS, present in about 50% of patients.⁷ Trauma and pressure exerted by the shoes are triggering factors, which justifies the feet's most common location.³

The various treatment options include surgical excision, CO2 laser, shaving followed by phenolization,⁷ topical rapamycin,⁸ and even nail amputation.² There are divergences in the literature about the best therapeutic option.⁷ Each modality has advantages and disadvantages, and, regarding the recurrence rate after the proposed treatments, there are no consistent data in the literature. However, it is believed that techniques that preserve the matrix may have significant recurrences.⁷

To choose the ideal treatment, one must consider the location of the tumor (hands or feet), age of the patient, main complaint (pain, discomfort, or cosmetic alteration), the life cy-



FIGURE 5: Left hallux nail: third month after surgery



FIGURE 6: Nail of the third finger of the left hand: eight months after surgery



FIGURE 4: Nails of the second and third fingers of the right hand: third postoperative day

cle of the tumor (primary or recurrent), number of lesions, and comorbidities.⁷

Surgical excision of the lesions is effective and inexpensive, not requiring specific devices. This treatment is preferable in patients with low surgical risk, recurrent tumors, extensive and multiple, especially in the feet. The aesthetic result is usually satisfactory.⁷ Given the above, we chose the surgery in our case due to the minimal surgical risk, the presence of large and multiple lesions, especially on the feet, in addition to nail deformities and the need for functional resolution.



FIGURE 7: Right hand nails: eight months after surgery

Regarding the other therapeutic modalities, there is the CO₂ laser, which directs bundles of light waves until the complete vaporization of the tumor. It has a low risk of bleeding, short duration (about 10 seconds per lesion), in addition to a satisfactory aesthetic result and excellent healing. It may be a proper choice for patients with surgical contraindications, small and moderate lesions, especially in the hands.^{4,5}

Shave excision followed by phenolization involves removing the protruding portion of the tumor and applying phenol to the lesion base. This method allows the preservation of the matrix and the nail plate, being more used in patients with high surgical risk, prioritizing the aesthetic result, and patients with small tumors and preferably located in the hands.⁷ Possible complications are necrosis, infection, and deformities of the fold and nail plate, which are rare.⁹ We did not choose this option due to the high risk of worsening nail dystrophy.

For aggressive, recurrent KT's with a high degree of morbidity, amputation of nail devices and reconstruction by full-thickness skin graft presents the possibility of an effective result.²

Another option is rapamycin, which belongs to the class of immunosuppressants. This medication inhibits mTOR (mammalian target of rapamycin), suppressing vascular growth factors, and destroying tumor cells. Its topical use shows promising results for the treatment of subungual fibroma. It is considered safe and well-tolerated, with rapid lesion involution. The recommendation is to use it twice a day, providing the clinical picture's initial improvement in just two months and the total disappearance of the lesions after six months.⁸

CONCLUSION

So far, there is no gold standard treatment in the literature for TK.⁷ The decision must be based on the advantages and disadvantages of each method. We report the case of a patient successfully treated for fibroma after surgical approach, with healing, excellent aesthetic and functional results, without complications, and without recurrence of the lesions in the first eight months of follow-up. ●

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Undifferentiated pleomorphic sarcoma: a case report

Sarcoma pleomórfico indiferenciado: Case report

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ABSTRACT

Undifferentiated sarcomas are neoplasms of soft tissues derived from the mesoderm. We report a case of undifferentiated pleomorphic sarcoma, its dermoscopy, staging, and therapeutic approach.

Keywords: Dermoscopy; Sarcoma; Soft Tissue Neoplasms

RESUMO

Os sarcomas indiferenciados são neoplasias de tecidos moles derivados do mesoderma. Relatamos um caso de sarcoma pleomórfico indiferenciado, sua dermatoscopia, seu estadiamento e sua abordagem terapêutica.

Palavras-chave: Dermoscopia; Neoplasias de Tecidos Moles; Sarcoma

INTRODUCTION

Sarcomas are tumors derived from the mesoderm. They are divided into two types: bone and soft tissue sarcomas, which include skin tumors.¹ Soft tissue sarcomas affect one to five individuals/ 100,000/ year, mainly over 55 years old.² Its main subtypes are gastrointestinal, liposarcoma, and leiomyosarcoma.² There are two types of soft tissue sarcomas that most commonly appear on the skin: undifferentiated pleomorphic sarcoma (previously called storiform-pleomorphic malignant fibrous histiocytoma) and myxofibrosarcoma (formerly called myxoid malignant fibrohistiocytoma).¹ Cutaneous sarcomas usually present as skin-colored nodules or subcutaneous masses, with rapid growth, most commonly in the limbs.¹

There is a histological grade (G) for soft tissue sarcomas (FNCLCC-France) in which three parameters receive a score: differentiation (1 to 3), mitotic index (1 to 3), and tumor necrosis (0 to 2). The sum of the numbers results in GX, G1 (2 to 3), G2 (4 to 5), and G3 (6 to 8). The American Cancer Society (ACS) has also established a TNM staging for this group of tumors depending on their topography: head and neck, trunk and extremities, thoracic and abdominal visceral organs, and retroperitoneum.³

Undifferentiated pleomorphic sarcoma (UPS) is the fourth most important soft tissue sarcoma. Regarding its epidemiology, it affects mostly men (two thirds) in relation to women, in the age group of 50 to 70 years (most affected age group).⁴ It is more frequent in the lower limbs, but 10% to 15% occur in the head and neck.¹ Regarding its evolutionary trend, it presents a local recurrence of 19% to 31%, and a metastasis index of 31% to 35% (with a frequency of 90% in the lungs, 85% in the bones, and 1% in the liver). Its five-year survival rate is 65% to 70%. The recommended treatment is surgical with 2 cm margins and adjuvant radiotherapy in selected cases (however, it is difficult to obtain free margins through surgery). For metastatic disease, chemotherapy is indicated (albeit with poor results) with anthracyclines (16% to 27% response) associated or not with ifosfamide. Doxorubicin is palliative. New studies have shown greater benefit with eribulin mesylate.⁵

CASE REPORT

An 88-year-old woman presented a nodular lesion with progressive growth in the left clavicle region for three months. Dermatological examination showed a violet, ulcerated tumor, with friable necrosis, 11 cm in the longest axis, in the left infraclavicular area (Figure 1). Polymorphic vessels characterized the dermoscopy without other specificities (Figure 2). We performed the lesion's incisional biopsy, whose anatomopathological examination revealed infiltration of the deep portion of the sample by large cell neoplasia, with a predominant fusiform shape, with cell nuclei of varying sizes and shapes, with irregular chromatin. Numerous mitosis figures, compatible with fusocellular and pleomorphic sarcoma, infiltrated the reticular dermis, histological grade 3 (Figure 3). The immunohistochemical panel showed a negative result for the expression of all the antigens surveyed (AE1/AE3, p63, 34BetaE12, protein S-100, SOX-100, melan-A, AML, desmin, CD3, and CD34), compatible with undifferentiated sarcoma. After anatomopathological examination, we opted for excision of the lesion with wide surgical margins and closure by primary intent of the surgical wound (Figure 4). We performed screening for lung, lymph node, and abdominal metastases with imaging tests, which was negative. The patient is being followed up, with no signs of local recurrence (Figure 5).

DISCUSSION

For TNM staging of this case, we adopted the American Joint Committee on Cancer (AJCC) 2018 reference for the trunk and extremities' soft tissue sarcomas. Table 1 represents the



FIGURE 1: Clinical image



FIGURE 2: Dermoscopy

T, and Table 2 shows the final staging (note that the G classification described in the introduction is considered). In both tables, the case reported is highlighted.

In the reported case, immunohistochemistry was essential since clinically, and even histopathologically undifferentiated pleomorphic sarcoma makes a differential diagnosis with:

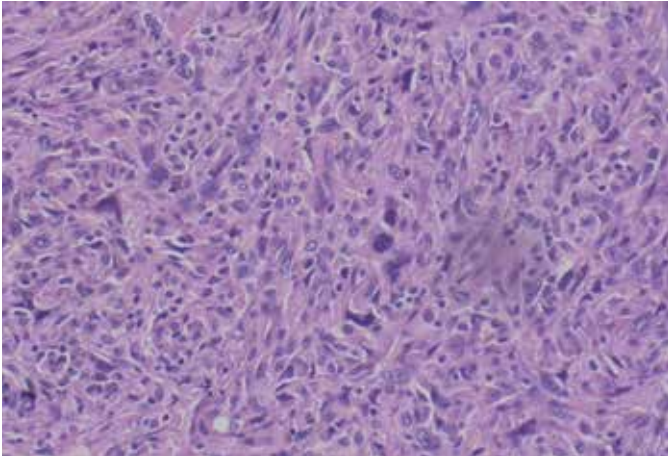


FIGURE 3: Histopathology



FIGURE 5: Final result



FIGURE 4: Wound after exeresis

- Melanoma (desmoplastic; frequent positivity for S100, HMW-MMA; rare positivity for SMA, desmin, factor XIIIa, laminin, and type IV collagen; and generally negative for HMB-45, Gp100, melan A/Mart1, tyrosinase, and transcription factor of microphthalmia).⁶

- Spindle cell squamous cell carcinoma (positive for AE1/AE3, vimentin, and CAM5.2, and vimentin).⁷

- Dermatofibrosarcoma protuberans (positivity for CD34 and vimentin; negativity for neuron-specific enolase, HMB-45, and S100 protein).⁸

- Atypical fibroxanthoma (exclusion diagnosis, negative for cytokeratins to exclude spindle cell carcinoma, spindle cells, S-100, and melanogenic markers such as melan-A and HMB-45 to exclude melanoma; desmin, actin, and H-caldesmon to

exclude leiomyosarcoma; and p63 and p40 to exclude squamous cell carcinoma).⁹

- Leiomyosarcoma (positivity for smooth muscle actin, desmin, and S100).¹⁰

- Undifferentiated pleomorphic sarcoma (positive for vimentin, factor XIIIa, CD68, CD10, CD34, CD 99; and negative for keratin, muscle, and melanocytic markers).¹¹

CONCLUSION

Table 1: Final TNM

T	Values (cm)
1	Until 5
2	5 to 10
3	10 to 15
4	Higher than 15

Table 2: G staging

Staging	TNM and G
IA	T1N0M0G1
IB	T2-4N0M0G1
II	T1N0M0G2-3
IIIA	T2N0M0G2-3
IIIB	T3-4N0M0G2-3
IV	N1M1

The case presented enriches the dermatologist's memory regarding the possible differential diagnoses among the rare mesenchymal tumors. We emphasized how essential the role of histopathology and immunohistochemistry is, reviewing the game

of markers with their diagnostic correlations in this mini-challenge and bringing the opportunity to review the staging and the conduct of soft tissue sarcomas. ●

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Oculofacial tumor aggressiveness due to moderately differentiated squamous cell carcinoma under systemic immunocompetence

Agressividade tumoral óculo-facial por carcinoma epidermoide moderadamente diferenciado sob imunocompetência sistêmica

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ABSTRACT

Squamous cell carcinoma (SCC) represents the second most common type of skin cancer. SCC originates from the atypical proliferation of the cells of the epidermis's spinous layer and is more frequent in men over 50 years of age, with a low skin phototype and history of sun exposure. We describe the case of a 61-year-old man, previously healthy, with a lesion presenting fast and exuberant growth on the face. The anatomopathological and immunohistochemical exams confirmed the diagnosis of moderately differentiated SCC.

Keywords: Carcinoma, Squamous Cell; Neoplasms, Squamous Cell; Skin Neoplasms

RESUMO

O carcinoma epidermoide (carcinoma escamocelular, carcinoma de células escamosas ou CEC) representa o segundo tipo de neoplasia cutânea mais comum. O CEC origina-se da proliferação atípica das células da camada espinhosa da epiderme e é mais frequente em indivíduos do sexo masculino, maiores de 50 anos, de fototipo baixo e com história de exposição solar. Descrevemos o caso de um homem de 61 anos, previamente hígido, apresentando lesão de crescimento rápido e exuberante na face, cujo anatomopatológico e imuno-histoquímica comprovaram tratar-se de CEC moderadamente diferenciado.

Palavras-chave: Carcinoma de Células Escamosas; Neoplasias Cutâneas; Neoplasias de Células Escamosas

INTRODUCTION

Non-melanoma skin cancer is the most common type of cancer in the world and Brazil. Squamous cell carcinoma (SCC) is the second most frequent subtype. While most patients present a good prognosis, some cases may have worse outcomes.¹ Recent epidemiological data have shown an increase in its incidence worldwide, which impacts public health.² Thus, early diagnosis and treatment are essential. We present an unusual case of moderately differentiated SCC, with a fast and exuberantly aggressive local evolution in a patient without evidence of immunosuppression.

CASE REPORT

A 61-year-old man, retired farmer, presented an erythematous papule infiltrated with a central crust in the right malar region. He was monitored for multiple previous non-melanocytic skin neoplasms and had no evidence of immunosuppression or other comorbidities. The patient was referred for biopsy under the diagnostic hypothesis of keratoacanthoma or SCC. Histopathology showed actinic damage and sebaceous hyperplasia, not presenting neoplastic cells. The patient returned after three months for follow-up, showing exacerbated growth of the lesion, exulcerated tumor, local pain, and loss of ipsilateral vision (Figure 1). The new histopathological exam evidenced moderately differentiated squamous cell carcinoma (Figure 2). Computed tomography (CT) of the face showed a large expansive, infiltrative lesion, with regional involvement of the skin and subcutaneous tissue, superficial irregularities and ulcerations, orbit's impairment, zygomatic process, orbital floor, and projection to the maxillary sinus. There was also evidence of proptosis of the eyeball, surrounded by the lesion. We observed no swollen lymph node enlargement in the cervical region. The patient was staged as T4aN0M0 and urgently referred to the institution's Head and Neck Surgery Service and briefly submitted to a surgical procedure. We performed an orbital exenteration with total maxillectomy and partial parotidectomy, with reconstruction using a microsurgical flap of the rectus abdominis muscle, with good postoperative evolution (Figure 3) and anatomopathological exam indicating lesion-free margins.

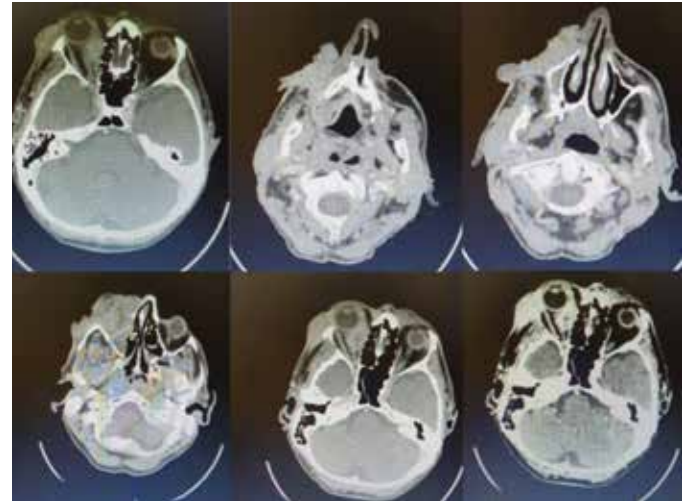


FIGURE 2: Computed tomography showing skin and subcutaneous infiltration, with superficial ulcerations; orbital impairment, zygomatic process, orbital floor and projection to the maxillary sinus; proptosis of the eyeball, surrounded by the lesion



FIGURE 1: Moderately differentiated SCC on the face: large ulcerated tumor, affecting the ocular region



FIGURE 3: Surgical outcome

DISCUSSION

SCC originates from the atypical proliferation of cells in the spinous layer of the epidermis. It may present clinically as a papule, nodule, or plaque lesion and may also present associated hyperkeratosis and ulceration. Professionals should consider actinic keratoses, basal cell carcinomas, attachment tumors, amelanotic melanomas, and keratoacanthoma as differential diagnosis,

as in our case.² Well-differentiated SCC can be clinically and histopathologically similar to keratoacanthoma, and, therefore, it is crucial to rule out this possibility in contexts of rapid tumor growth. In the present case, histopathology and invasiveness of deep structures confirmed that it was SCC.

Epidemiologically, SCC occurs predominantly in men over 50 years old, with a low skin phototype and sun exposure history. It is the most frequent tumor in transplanted individuals, being two to three times more common than basal cell carcinoma. The natural history of squamous cell carcinomas varies from slow to fast, with accelerated growth being observed more frequently in immunosuppressed patients. In the context of immunosuppression, SCC is often multiple and tends to be more aggressive.⁶

Among the most common causes of systemic immunosuppression, in addition to organ transplants (58%), there are systemic inflammatory diseases (16%).⁴ In kidney transplant patients, treatment with immunosuppressants combined with ultraviolet radiation causes a deficiency in the systemic and local immune response. These patients have a numerical decrease in lymphocyte subtypes (CD4 and CD8), lymphocytes that express the interleukin-2 receptor on the skin, and Langerhans cells. Thus, there is less expression of MHC-II, compromising local immunological competence, and favoring the appearance of pre-neoplastic and neoplastic lesions.⁵

As in other types of malignant neoplasms, local leukocyte populations are protective against cancer development and also play a role in the “sculpture” of the high-intensity tumor, leading to many causes of immunogenicity and tumor progression. UV radiation and chronic viral transport may represent unique risk factors for SCC development, and the local immune system plays a key role in modulating the response to both.⁶

The staging of squamous cell carcinomas considers the tumor volume measured by diameter and depth. It also considers lymph node metastases and distant metastases. High-risk characteristics for metastatic SCC include impairment greater than 2 mm in thickness, Clark’s level greater than or equal to IV, perineural invasion, anatomical location (ear, lips, and mucous membranes are at higher risk), origin in scarring, immunosuppression, and differentiation degree (worse prognosis is associated with poorly differentiated and undifferentiated).² Our case presents a patient with negative staging for lympho-hematogenous dissemination, but with extreme local aggressiveness, ultimately affecting one of the ocular devices.

The first-line therapy for SCC is complete surgical excision with histopathological control of the margins.³ The literature recommends margins of 4 mm to 10 mm, depending on the clinical characteristics suggestive of high risk of recurrence or not – although there is no consensus. In low-risk tumors, professionals may choose curettage with electrocoagulation and cryosurgery. Adjuvant radiotherapy is recommended in patients with negative margins after surgery in case of perineural involvement, and few drugs have been used successfully in adjuvant or neoadjuvant therapy for metastatic SCC.³

This case presented an uncommon rapid growth, aggressiveness, and tumor volume of the moderately differentiated SCC, mainly due to the absence of a factor that compromises his systemic immune status, highlighting the potential local immunomodulation by UVR.

Surgical management proved to be complicated and followed the principle of Oncodermatology therapy, with the neoplasia’s wide excision. ●

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Condylomata acuminata in childhood treated with 5% imiquimod cream: case report

Condiloma acuminado na infância tratado com creme de imiquimode a 5%: Case report

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ABSTRACT

DNA-virus, which belongs to the papillomavirus group, causes Condylomata acuminata. In children under 3 years of age, transmission tends not to be due to sexual abuse but rather vertically. There is no consensus for the treatment of anogenital warts in children. The chosen method is individualized for each patient, preferring less traumatic treatments, with fewer sequelae. Although there are still some restrictions on the imiquimod use in children under 12, studies have already shown this therapy's success in this age group. We report a case treated with 5% imiquimod cream with the resolution of pre-existing lesions.

Keywords: Condylomata Acuminata; Papilloma; Warts

RESUMO

O condiloma acuminado é causado pelo DNA-vírus, que pertence ao grupo papovavírus. Em menores de três anos, a transmissão tende a não ser por abuso sexual, mas, sim, de maneira vertical. Não existe consenso para o tratamento das verrugas anogenitais em crianças. O método escolhido é individualizado para o paciente, sendo a preferência por tratamentos menos traumáticos, com menor seqüela. Embora ainda existam algumas restrições ao uso do imiquimode em menores de 12 anos, estudos já mostram o sucesso desta terapêutica nesta faixa etária. Será descrito um caso tratado com imiquimode creme a 5%, com resolução das lesões preexistentes.

Palavras-chave: Condiloma Acuminado; Infecções por Papillomavirus; Verrugas

INTRODUCTION

Condylomata acuminata is caused by the DNA-virus, which belongs to the papillomavirus group, also known as HPV.¹ It is associated with viral types 6, 11, 30, 42, 43, 44, 45, 51, 52, 54, and it is possible to exist more than one subtype in a lesion.² Physical examination characterizes it as vegetating lesions, with sessile papules (seen in the raphe of the penis), and pink, moist projections resembling a cauliflower. These lesions are in the genital and perigenital region and may affect the urethral, rectal, or vaginal areas. Studies show that the association between HPV and sexual abuse increases directly with age, especially after five.³

Most studies show that HPV infection in children does not necessarily mean sexual contact, and other forms of contamination should be considered.⁴ A survey conducted with 42 prepubertal children with anogenital condyloma showed that 28.6% of them acquired HPV by vertical transmission, 7.1% resulted from the autoinoculation of warts located on the hands, and only 4.8% were associated with sexual abuse.³

The treatment of *Condylomata acuminata* in children requires greater attention, as ablative procedures and laser surgeries are painful and require general anesthesia in the vast majority. Among other treatment options is topical imiquimod 5%, even though its use in children has not yet been fully defined, unlike in the adult population that responds well to therapy, with a favorable resolution of lesions.⁵

CASE REPORT

One year and six months old male patient presented to consultation with his companion (mother), who has referred perianal and scrotum lesions on the patient for six months, with a progressive increase in number and size. Physical examination revealed erythematous papules in the scrotum and inguinal region, others confluent forming a plaque with a vegetative aspect, with a pink color, affecting the perianal area (Figure 1). The patient was born by normal delivery, at term, and without complications. The mother had a history of ovarian cancer and denied genital or extragenital lesions. The father and two brothers were also assessed, with no evidence of suspicious lesions. Serologies (syphilis, HIV, hepatitis B and C) were requested from the patient and parents, all negative. They were referred to the coroner to rule out sexual abuse. The patient was referred to the pediatric surgeon for evaluation and biopsy. While waiting for the consultation with the professional, we decided to start imiquimod cream 5%, twice a week. In three weeks, we observed a significant improvement of the vegetative and papular lesions. As an adverse event, we perceived moderate erythema (Figure 2). The treatment was conducted for seven weeks (Figure 3).

DISCUSSION

Condylomata acuminata appears in 10% of cases of sexually transmitted infection (STI). When found in childhood, there must be a suspicion of sexual abuse.³ Anogenital warts cases are progressively increasing in children, but it appears to be due to an increase in lesions in adult women.⁴

When found in the age group of children under three years of age, transmission tends to be in other ways. The most common is vertical (through the birth canal) since some authors described an incubation period of one to three years, or it can occur an ascending infection. There are cases of autoinoculation and heteroinoculation and, also, transmission by fomites. Even though it is not the most common form in that age group, sexual abuse must not be ruled out in any case of *Condylomata acuminata*.^{3,4} If sexual abuse is suspected, it should be investigated. If confirmed, the Guardianship Council has to be contacted with social assistance. The physical examination must be complete, trying to exclude warts in other regions.³



FIGURE 1: Erythematous papules in the perineum region, scrotum, inguinal region, associated with the vegetating plaque in the perianal region



FIGURE 2: Presence of hyperemia in the perianal, inguinal region, and penis body after three weeks of treatment with imiquimod

There are several types of treatments in cases of *Condylomata acuminata*, including surgical excision and CO₂ laser, in addition to topical treatments using imiquimod cream 5%. The recommendation is to apply the medication in the affected area three times a week, for 16 weeks, generating irritation and inflammation, with lesions' resolution. Another topical treatment is fluorouracil cream 5%, with recommended daily use until irritation appears. Podophyllin 25% is another option, applied to the lesions and washed after four hours, but is contraindicated



FIGURE 3: Total lesion regression after seven weeks of treatment with imiquimod

in children and pregnant women.⁶ Other options are chemical cauterization with trichloroacetic acid 50–70%, cryosurgery, and shave removal with electrocoagulation of the base.⁷

All these treatments are effective and safe in the adult population. However, when treating condyloma in children, the ideal would be that therapies present low cost, effectiveness and cause no trauma. Nevertheless, all these characteristics are not yet available to the entire pediatric population. The most used treatments are chemical or mechanical destruction of the lesions through cryocauterization, electrocauterization, or loop diathermy, all of which are painful and, in the great majority, requiring general anesthesia. Some studies show efficacy and safety with topical treatment with imiquimod cream 5% in children un-

der 12, even though its use in this age group has not yet been fully defined.^{7,8} The method chosen is individualized for each patient, with a preference for less traumatic treatments, which will evolve with fewer sequelae.⁸

Imiquimod is a topical immunomodulator, agonist of toll-like receptors 7, which can mediate the innate and cellular immune response and stimulate interferon (IFN) production and cytokines that lead to the destruction of collagen. Also, it acts on antigen-presenting cells, showing HPV antigens with greater effectiveness to CD4+ T lymphocytes. In addition to inhibiting HPV replication by 90%, thus decreasing viral load, it prevents recurrences by releasing cytokines, tumor necrosis factor-alpha, and IFN-alpha.¹

The effect of imiquimod mimics the normal immune response, increasing the release of IFN, which is antiviral, preventing proliferation and angiogenesis, also increasing the amount of messenger RNA from the CD4 + T lymphocytes at the site. It may still have the effect of anti-HPV immune memory, thus significantly reducing the recurrences of lesions.¹

When using this topical medication in children, these patients will have no disadvantages such as pain. Also, they can be treated in their own homes, being a medication with few adverse events. Among adverse events, it is possible to occur erythema, itching, burning, erosion, and greater sensitivity at the application site. Although quite unusual, patients may also experience systemic symptoms such as fatigue, fever, myalgia, central and peripheral nervous system changes, and gastrointestinal symptoms.⁵

In one study, it was possible to observe the success in treating children aged six months and 19 months after applying imiquimod for three and eight weeks, respectively. Despite having erythema around the lesions frequently, no other adverse events of importance occurred in these cases, as in our case.⁵

This case of Condyloma acuminata in children, which achieved therapeutic success with the use of imiquimod cream 5%, proved that this therapy is a good alternative to conventional treatments that, in most cases, have associated pain and even sequelae and trauma to children. ●

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Case report

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Earlobe transposition: a simple flap in the reconstruction of full-thickness surgical defect of anti-tragus

Transposição de lóbulo de orelha: um retalho simples para reconstrução de defeito cirúrgico de espessura total do antítrago

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ABSTRACT

Skin cancer frequently occurs on the pinna. The three-dimensional conformation and the limited availability of redundant regional skin make the reconstruction of this anatomical area challenging. We report the reconstruction of anti-tragus using an earlobe transposition flap.

Keywords: Carcinoma, Squamous cell; Ear auricle; Mohs surgery; Skin neoplasms; Surgical flaps

RESUMO

O câncer de pele ocorre com frequência na região do pavilhão auricular. A conformação tridimensional e a disponibilidade limitada de pele redundante regional tornam a reconstrução desta área anatômica desafiadora. Relata-se a reconstrução de antítrago utilizando-se um retalho de transposição de lóbulo de orelha.

Palavras-chave: Carcinoma de células escamosas; Cirurgia de Mohs; Neoplasias cutâneas; Pavilhão auricular; Retalhos cirúrgicos

INTRODUCTION

The pinna is a place with a high incidence of skin cancer. The literature estimates that about 16% of skin tumors occur at this location.¹ The pinna is formed by several subunits endowed with concavities and convexities that make its shape peculiar, requiring a detailed surgical reconstruction after the excision of tumors. We describe the reconstruction of a full-thickness defect of the antitragus using an earlobe transposition flap after a Mohs micrographic surgery (MMS).

CASE REPORT

A 72-year-old patient presented two well-differentiated squamous cell carcinomas (SCCs), located (Figure 1) in the region of the tragus and the antitragus. Both tumors were treated using the MMS technique, with free margins obtained in the first stage. After the closure by primary intent of the tragus re-



FIGURE 1: Nodules infiltrated in the region of tragus and antitragus, with previous histological diagnosis of well-differentiated infiltrative SCC



FIGURE 2: A. Transpositional flap with closure by primary intent of the secondary defect. B. Ten days postoperative



FIGURE 3: Surgical defect of 1.5 cm x 1.4 cm, affecting the total thickness of the antitragus. Design of the earlobe transposition flap

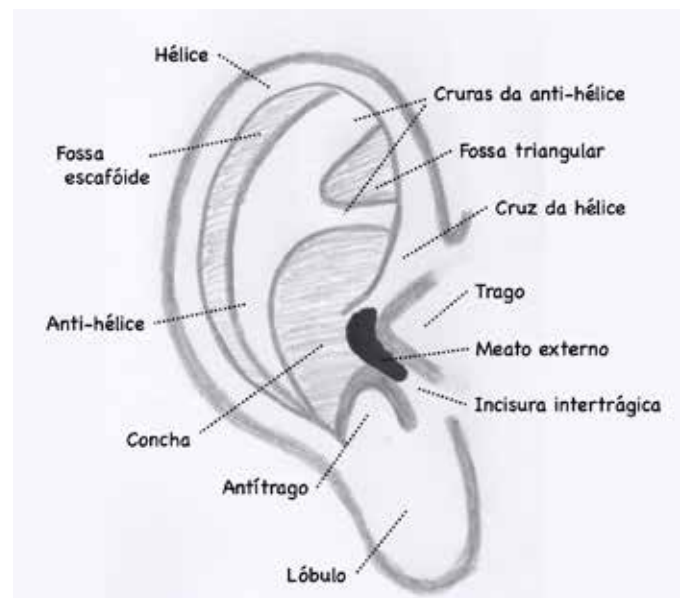


FIGURE 4: Anatomical subunits of the auricle

gion, a surgical defect of 1.5 cm x 1.4 cm, compromising the sub-unit of antitragus, was still present. An earlobe transposition flap was planned (Figure 2), with its redundancy in the pivotal region, purposely left to recreate the contour and projection present in the antitragus' natural anatomy (Figure 3).

DISCUSSION

About 12 subunits or anatomical points of interest form the pinna (Figure 4).² Regarding the antitragus, the literature on its reconstruction is scarce. A literature review found three articles describing complex techniques for its reconstruction, with good cosmetic results. A case report written by Chadha, Grob, and Soldin³ described an “open book” flap in the reconstruction of a defect involving antitragus, conchal bowl, and antihelix in

a case of SCC. The lesion was removed, and the surgical defect repaired using a tunneled transposition flap of the preauricular region in a single time. Finally, Gonzalez-Sixto et al.⁵ described a series of four reconstructed ear defects with a chondrocutaneous advancement flap by V-Y advancement. One of them used the flap to reconstruct the antitragus.

Primary and secondary wound closures are also simple closure options for this subunit, although earlobe elevation may occur as a consequence. The limited availability of redundant skin and the frequent extrapolation of anatomical subunits by tumors in this region make the pinna reconstruction challenging.^{6,7} The earlobe is one of the few ear sites with sagging skin, thus being useful for use as a donor site in flaps. ●

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Use of Google Glass in cheiloplasty: the surgeon's perspective

Uso do google glass na queiloplastia: a perspectiva do cirurgião

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ABSTRACT

Wearable technology has come to change the way people see and experience the world. Google Glass is one of these technologies created by Google Inc., Mountain View, California. This device has many uses, including in the medical field. One of these uses is the possibility of providing viewers with the surgeon's view during surgery. Surgeries with a small operative field, like cleft lip surgery, or cheiloplasty, should benefit from Google Glass. In our institution, we recorded the first cleft lip surgery using Google Glass to give our impression of the Glass.

Keywords: Education, Medical; Innovation; Technology

RESUMO

Tecnologia “de vestir” chegou para mudar a forma como as pessoas veem e experimentam o mundo. Google glass é uma dessas tecnologias e foi criada pelo Google Inc. California Mountain View. Este dispositivo tem muitas possibilidades de uso, inclusive na área médica. Dentre estes usos, está a viabilidade de fornecer aos espectadores a visão do cirurgião durante a cirurgia. Cirurgias com pequeno campo operatório, como queiloplastia, devem beneficiar-se com o uso do google glass. Em nossa instituição, gravamos a primeira queiloplastia com google glass com a intenção de dar a nossa impressão dos óculos da Google Inc.

Palavras-chave: Educação Médica; Inovação; Tecnologia

INTRODUCTION

Technology is increasingly present in our lives, and in the medical field, it is no different. At every moment, we face new devices in the clinical routine and in the surgical room that change the way we interact with the world.

Better known as “wearable” technology, Google glass is one of these technologies, and is already used in the medical field.¹⁻⁴ Created by Google Inc. California Mountain View, this device provides the possibility of seeing the surgeon during the surgery in real-time or recording the procedure for later review. Davis and Rosenfield made the first description of this technology's use in a surgical procedure in 2013,^{5,6} and their team demonstrates the difficulties and perspectives of this technology.

Surgeries with a small surgical field, such as cheiloplasty, benefit from the use of Google glass. Also, in teaching units, the difficulty in monitoring the surgery steps by residents and interns present in the room and outside the surgical field, due to the reduced visual field and the need to stop the surgery at each

Case report

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step, dramatically increases the time of the procedure.

Our institution recorded the first cleft lip surgery with Google glass to give our impression of this technology in reduced visual field surgeries.

OBJECTIVE

This study aims to assess the use of a new technology in cheiloplasty, with a small surgical field, and use the technology for the improvement and learning of the team.

METHODS

We chose a patient with an incomplete pre-incisive foramen fissure after obtaining informed consent with a recording authorization term. The absence of fast internet for streaming video limited the use of glasses only for recording. The patient was placed in Rose position and a Millard cheiloplasty was performed. After the surgery, we showed the video to residents.

RESULTS

The image was clear and made it possible to see the procedure and structures without any doubt. At first, its use may be strange for the surgeon, but over time it becomes habitual.

The procedure was conducted with the resident team for learning. The presence of the team during the surgery was essential. However, after the procedure with the recorded video, the surgical steps could be discussed and demonstrated with a detailed view, which generated fixation and understanding of



FIGURE 2: During surgery



FIGURE 3: First person view through Google glass



FIGURE 1: Surgeon using Google glass

the technique.

DISCUSSION

There are many benefits to using “wearable” technology like Google glass.⁷ The functionality that allows communication between doctors and the exchange of experiences or consultation during the surgical procedure is beneficial. However, the absence of fast internet is a limiting factor in countries like Brazil. Communication problems are a prevalent cause of errors in surgery, and this technology can reduce these errors.

The primary applicability for this technology that we see at the moment is in the surgical teaching, as in cheiloplasty, which has a limited surgical field, and the surgeon can share in

real-time the procedure steps. As far as we know, this was the first cheiloplasty performed with Google glass, and we were able to take the opportunity to review the surgical field with the entire team.

However, ethical issues and the lack of fast internet streaming video can be some limitations. Other features, such as remote supervision and even consultation for minor procedures, could also be used.

CONCLUSION

The use of Google glass in surgeries with a reduced visual field is very feasible, and residents can enjoy and share first-person learning from the attending physician. However, the full potential of this technology has yet to be discovered. The possibility of performing surgical procedures and teaching or discussing from a first-person view is excellent for surgeries with a small field or procedures performed in educational institutions. In the current moment of social isolation that we live in, it is an option to continue teaching. ●

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Treatment of Necrobiosis Lipoidica in the left forearm with association of Intense Pulsed Light and Erbium-Yag Laser 2940nm

Tratamento de necrobiose lipoídica no antebraço esquerdo com associação entre luz intensa pulsada e laser Erbium-YAG 2940nm

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ABSTRACT

Necrobiosis Lipoidica (NL) is a granulomatous dermatosis, mainly affecting people with diabetes due to collagen degeneration. Although there are different treatments, all have little response. Among the options, the Intense Pulsed Light (IPL) and the Laser Erbium-YAG enhance the production and remodeling of the collagen treating the atrophy, besides reducing the erythema by the coagulation of blood vessels. We report a case of a 24-year-old woman with type I Diabetes Mellitus who underwent NL treatment on her left forearm in seven sessions with the association of IPL and 2940 nm Erbium-YAG laser. We observed improvement in central atrophy and erythema.

Keywords: Necrobiosis lipoidica; Diabetes mellitus; Lasers; Laser therapy; Intense pulsed light therapy; Collagen

RESUMO

A necrobiose lipoídica (NL) é uma dermatose granulomatosa, que acomete principalmente diabéticos devido à degeneração do colágeno. Embora existam diferentes tratamentos, todos apresentam pouca resposta. Dentre as opções, a luz intensa pulsada (LIP) e o laser Erbium-YAG permitem aumentar a produção e remodelamento do colágeno tratando a atrofia, além de diminuir o eritema pela coagulação de vasos sanguíneos. Relata-se caso de mulher, 24 anos, com diabetes mellitus tipo I, submetida a tratamento de NL no antebraço esquerdo em sete sessões com a associação entre LIP e laser Erbium-YAG 2940nm. Observou-se melhora da atrofia central e do eritema.

Palavras-chave: Colágeno; Diabetes mellitus; Lasers; Necrobiose lipoídica; Terapia a laser; Terapia de luz pulsada intensa

INTRODUCTION

Necrobiosis Lipoidica (NL) is a relatively rare chronic granulomatous disease that affects more adults, women, and diabetic patients.^{1,2} It may occur associated with autoimmune thyroiditis, obesity, dyslipidemia, systemic arterial hypertension, rheumatoid arthritis, sarcoidosis, and inflammatory bowel disease.² Its etiology is unknown and may be the result of hypoxia due to microangiopathy and abnormal glucose metabolism by fibroblasts, resulting in collagen degeneration.^{1,2}

Clinically, it manifests as a circumscribed oval plaque, with an erythematous raised border and an atrophic center

with telangiectasia on the surface. It is typically located in the pre-tibial region,^{1,3} but can also be found on the face, trunk, and extremities.³ There is a possibility of ulceration related to overweight, hypertension, or trauma.^{1,2,3}

Histopathological examination shows collagen disorganization and degeneration, with T cells' inflammatory infiltration in the dermis and hypodermis.³

NL therapy usually consists of topical, intralesional, or oral corticosteroids. But there are several other therapeutic options such as topical retinoid and calcineurin inhibitor, PUVA, fumaric acid esters, antimalarials, anti-TNF-alpha, cyclosporine,^{1,2} pentoxifylline, intravenous immunoglobulin, skin graft, acetylsalicylic acid, dipyridamole, hyperbaric oxygen therapy, nicotinamide, benzoyl peroxide, photodynamic therapy, thalidomide, bovine collagen, clofazimine,¹ and mycophenolate mofetil.³

Intense pulsed light (IPL) treatment is based on its ability to increase collagen production (types I and III), reorganize it, and coagulate the blood vessel,³ improving the lesion's texture, and erythema. The treatment with fractional ablative lasers, which use water as their absorbing chromophore, would have a more selective action, with remodeling and increased collagen fibers production (types I and III) reducing the atrophy.

Although treatments with different types of laser and IPL appear to be promising in approaching NL, the pulsed dye laser (PDL) has case reports with conflicting results.⁴ A study reports a case of NL successfully treated with IPL after 13 sessions.³ The literature does not describe the treatment with Erbium-YAG 2940nm laser.

This paper reports a case of NL treated with IPL and Erbium-YAG 2940nm laser in a patient with long-term diabetes mellitus.

CASE REPORT

A 24-year-old woman with type I diabetes mellitus presented an oval, atrophic, yellowish plaque, with raised edges, telangiectasia, and slight flaking for two years. The lesion had 4.5 cm in diameter on the left forearm (Figure 1). The patient used injectable insulin glargine and aspart. After a clinical diagnosis of Necrobiosis Lipoidica (NL), she started the treatment with clobetasol propionate cream daily on the lesions' edges. After 45 days of unsuccessful treatment, we proposed a combined treat-

ment of IPL and Erbium-YAG 2940nm laser (Etherea® Platform). Table 1 shows the parameters used, and Table 2 describes the intervals between sessions.

The distal half was treated with IPL, and the proximal half with IPL and Erbium-YAG 2940nm laser in the first three sessions. Subsequently, the same treatment was instituted throughout the lesion.

The improvement in erythema was progressive, already noticed after the first session. After observing that there was no significant clinical difference between the portions of the lesion in the fourth session, we chose to conduct only IPL. The entire lesion was treated with IPL associated with Erbium-YAG 2940nm laser from the fifth session onwards. There was an improvement in central atrophy, less visible vessels, decreased thickness and erythema of the border (Figure 2), in addition to no further progression of the lesion. The patient is undergoing clinical follow-up.

DISCUSSION

NL remains a poorly understood disease, and conclusions about treatment have weak scientific evidence. Most cases use high-potency topical corticosteroids, and our patient received them in monotherapy at diagnosis and as adjunctive therapy in the fourth and fifth sessions. Nonetheless, it can aggravate atrophy and telangiectasia.



FIGURE 1: Pre-treatment clinical lesion presenting as an oval, atrophic, yellowish plaque, slight flaking, with raised edges and telangiectasia, 4.5 cm in diameter, in the left

TABLE 1: Scheme of treatment performed with the association of ILP and Erbium-YAG 2940nm laser in each session.

Session	IPL (nm filter, fluency mJ / cm ² , ms)			LASER ERBIUM-YAG 2940nm (Tip mtz, mode // mJ / cm ² , ms)
	1 ^a Pass	2 ^a Pass	3 ^a Pass	Single Pass
1 to 3	540, 16, 15	540, 16, 10	580, 18, 20	100, single // coagulation 45, 5
4 5	580, 18, 20	540, 17, 15	540, 17, 10	-
6	580, 18, 40	580, 18, 20	-	100, double // ablation 15, 300; coagulation 45, 5
7	580, 20, 40	580, 20, 20	-	



FIGURE 2: Left forearm lesion six months after end of the treatment with clinical improvement of erythema and atrophy

In addition to traditional treatments, IPL and different types of laser can be used, aiming to decrease inflammatory activity and improve atrophy and telangiectasia.

Two cases already used the pulsed dye laser to treat NL, without success in a few sessions,^{1,4} but with a good response regarding the reduction of symptoms and the lesion's size in six sessions.⁴ It indicates that in NL, the treatment can be more prolonged, with more sessions, to obtain better results.

The fractional ablative Erbium-YAG 2940nm laser stimulates the production of organized type I, III and VII collagen, and elastin, with increased tropoelastin.⁵ These changes would be responsible for the improvement in skin texture, in addition to the reorganization of collagen. These facts may be

TABLE 2: Intervals between sessions	
Interval (session)	Time (weeks)
1° and 2°	9
2° and 3°	7
3° and 4°	6
4° and 5°	5
5° and 6°	8
6° and 7°	13

responsible for the improvement of the patient's atrophy. There are no cases described with this type of treatment.

It is believed that IPL promotes a reduction in telangiectasia through its action on hemoglobin, anti-inflammatory and antiproliferative activity,⁶ acting in the production and organization of collagen fibers.³

The association of IPL and Erbium-YAG 2940nm laser allows clinical improvement with fewer sessions compared to the use of IPL alone. The production and reorganization of collagen improve the atrophy, moving the vessels away from the dermis in relation to the epidermis. In contrast, tropoelastin enhances the elasticity of the skin (which was aimed at using the combined technologies). IPL also acts by coagulating more superficial vessels, thus reducing erythema. We described an unprecedented case of NL treatment with associated use of IPL and Erbium-YAG 2940nm laser, with a moderate response after seven sessions, which remained six months after the end of the treatment. ●


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Extra-abdominal desmoid fibromatosis: the importance of Mohs micrographic surgery in a rare and recurrent tumor

Fibromatose desmoide extra-abdominal: a importância da cirurgia micrográfica de Mohs em um tumor raro e recidivante

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ABSTRACT

We report the case of a 13-year-old girl with a diagnosis of extra-abdominal desmoid fibromatosis, a neoplasm of benign origin but uncommon in the soft tissues. This study aims to expose the rarity of the tumor and its challenging therapeutic approach due to its high frequent local recurrence rates. For the treatment, Mohs micrographic surgery was performed in two stages to obtain a free margin.

Keywords: Fibromatosis, Aggressive; Mohs Surgery; Neoplasms

RESUMO

Apresenta-se caso de paciente do gênero feminino, 13 anos, com diagnóstico de fibromatose desmoide extra-abdominal, neoplasia dos tecidos moles, de origem benigna, porém incomum. Nosso objetivo é expor este relato pela raridade do tumor bem como por sua desafiadora abordagem terapêutica e pelas altas taxas de recidiva local frequentes nesses tumores. Para o tratamento, realizou-se cirurgia micrográfica de Mohs em duas fases para obtenção de margens livres.

Palavras-chave: Cirurgia de Mohs; Fibromatose Agressiva; Neoplasias

INTRODUCTION

Extra-abdominal desmoid fibromatosis is a rare, benign neoplasm, originating in fibroblastic cells and growing in a variable pattern. It can occur in almost any part of the body. Despite not presenting malignant behavior, such as metastases, the desmoid tumor has a high capacity for growth and local invasion. With few cases described in the literature, its surgical treatment is practically a consensus.^{1,2,3} The Mohs micrographic surgery (MMS) approach considerably reduces the chances of frequent recurrence of these tumors, estimated at approximately 11% to 64%, depending on the treatment used.⁴ We report a pediatric patient's case, with this neoplasm located on the lower lip, submitted to excision by micrographic surgery and followed for two years, without recurrence.

CASE REPORT

A 13-year-old girl presented a history of a lesion on the lower lip beginning seven months ago, with slow and progressive growth. She denied any symptoms or previous trauma at the site. The patient had a personal history of juvenile idiopathic arthritis (JIA), and she was using methotrexate, folic acid, and etanercept. The dermatological examination showed an erythematous-violaceous tumor, with a nipple-like surface and a well-defined fibroelastic consistency of approximately 2.5 cm on the lower lip, on the left. An excisional biopsy was performed, revealing fibromyxoid fusocellular mesenchymal proliferation, with an associated inflammatory cell component, on anatomopathological examination. The immunohistochemical panel showed diffuse and intense positivity of beta-catenin and vimentin and negative ALK-1. These findings pointed to the diagnosis of extra-abdominal desmoid fibromatosis.⁵

We opted for the excision of the lesion by Mohs micrographic surgery, revealing compromised margins in the first stage of the procedure and free margins after the surgical defect's enlargement. The patient was followed every three months in the first year and every six months in the second, with no clinical sign of recurrence of the lesion.

DISCUSSION

The desmoid tumor or desmoid fibromatosis consists of fibroblastic proliferation, originating in the soft tissues, with benign behavior. McFarlane first described it in 1832, and Muller coined the term desmoid five years later, based on the myofibroblastic cells that constitute these tumors. It has an estimated frequency of 3% of all soft tissue tumors. It most commonly affects individuals between 15 and 60 years old, being rare in the pediatric age group and after the fourth decade, with a slight preference for women. Its most frequent form of presentation is intra-abdominal, accounting for about 70% of cases.^{6,7,8}



FIGURE 1: Nodosidade eritemato-violácea, de superfície mamilonada e consistência fibroelástica

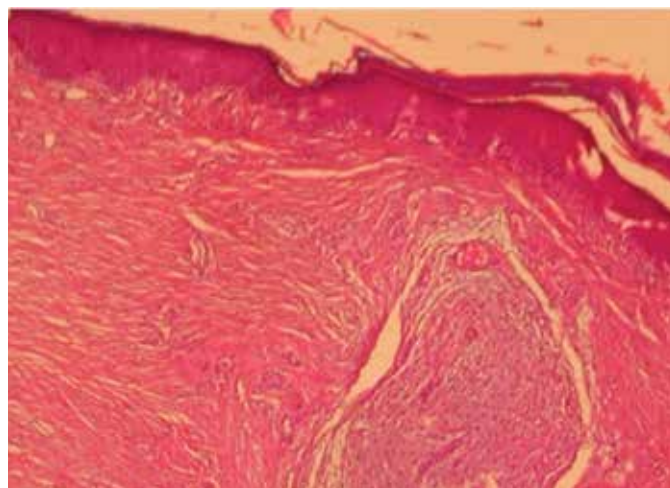


FIGURE 2: Proliferação mesenguimal fusocelular fibromixoide



FIGURE 3: Defeito cirúrgico após excisão com controle de margens operatórias. Realizada sutura com aproximação primária das bordas aproximação primária

The vast majority of cases, around 90%, are sporadic. Somatic mutations of the beta-catenin protein, which makes up the CTNNB1 gene, may boost this tumor's development.⁹ In addition to the genetic characteristics, other endocrine and physical factors, such as trauma, play an essential role in the disease's etiology.

In its rare, extra-abdominal form, its appearance in the extremities' soft tissues is more frequently observed, with half of the cases originating in the limbs, 43% in the chest, and only 7% in the head and neck region. However, any anatomical area is subject to its appearance.^{10,11,12} Our report stands out for its atypical presentation, both by the age group and by its very unusual location.

These tumors are characterized by presenting different degrees of aggressiveness with unpredictable biological behavior. Their natural history ranges from indolent and self-limited lesions to infiltrative lesions and with rapid local proliferation. The symptomatology depends on the presentation site and adjacent



FIGURE 4:
Paciente avaliada após
1 ano do procedimento
com bom resultado
funcional

structures. The majority is asymptomatic; however, due to its significant regional growth, it can compress or compromise other structures, organs, or the functionality of the affected region.^{13,14}

The diagnosis is made by anatomopathological examination that shows bundles of spindle cells, elongated, in the middle of stromal collagen, with variable vascularization. The cells are usually small, with clear cytoplasm and pale nuclei, showing no atypia or mitoses. Immunohistochemistry is positive for muscle cell markers, such as vimentin, desmin, and smooth muscle actin. In electron microscopy, spindles cells look like myofibroblasts.

The most common cases, intra-abdominal, usually require imaging methods to complement the diagnosis.⁷

According to most authors, the ideal treatment has not yet been established, although surgery is the first therapeutic option. The main challenge is the fact that, despite being histologically benign, they have high recurrence rates. Most of the cases described in the literature consist of intra-abdominal tumors where conservative treatment can be considered. However, there are few publications of lesions outside the abdominal cavity and

in pediatric patients. Nevertheless, when conventional deciding for local resection, it must contain a wide margin to avoid the risk of incomplete excision.⁶ Other therapeutic options include chemotherapy, hormone therapy, and radiation therapy.

In the case reported, the surgical approach, especially the treatment with MMS, was strictly necessary, considering the patient's age, the unfavorable anatomical location in a prime area, and the relevant percentage of these tumors' recurrence. Two surgical stages were necessary to obtain the free margins.

CONCLUSION

Health professionals must remember the desmoid tumor as a differential diagnosis of soft tissue tumors, and anatomopathological examination is essential for its diagnostic confirmation. The effectiveness of MMS is already widespread and should be indicated for the removal of tumors with potential for recurrence and in risk areas where tissue preservation is essential, as in our case. This approach was fundamental in healing, ensuring margin control and excellent aesthetic and functional results for the patient. ●

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Dermoepidermal grafting obtained by shave excision of papule formed post punch grafting in vitiligo: improvement of the cobblestone pattern

Enxerto dermoepidérmico obtido por remoção da pápula formada pós-enxertia com punch em vitiligo: melhora do padrão em paralelepípedo

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ABSTRACT

Vitiligo is an acquired dyschromia that affects 0,1-3% of the world population and can cause significant psychosocial distress in patients. Surgical management became essential in the last years. Mini-punch grafting is one of the cheapest and most straightforward approaches to all surgical treatments available. A limitation of this technique is the possibility of cobblestone-like repigmentation. We describe a new technique with a dermoepidermal grafting obtained by shave excision of papule formed post punch grafting in vitiligo. It could improve the cobblestone pattern in the donor site and the repigmentation in both donor and recipient areas.

Keywords: Dermatology; Hypopigmentation; Vitiligo

RESUMO

O vitiligo é uma discromia adquirida que afeta 0,1-3% da população mundial e pode causar sofrimento psicossocial significativo nos pacientes. O manejo cirúrgico tornou-se importante nos últimos anos. O minienxerto é uma das abordagens mais baratas e fáceis entre todos os tratamentos cirúrgicos disponíveis. Uma limitação desta técnica é a possibilidade de repigmentação em forma de paralelepípedos. Descreve-se uma nova técnica com enxerto dermoepidérmico obtido por remoção da pápula formada pós-enxertia com punches no vitiligo. Essa técnica pode melhorar o padrão em paralelepípedos no local doador e a repigmentação em ambas as áreas, doadora e receptora.

Palavras-chave: Dermatologia; Hipopigmentação; Vitiligo

INTRODUCTION

Vitiligo is an acquired depigmenting disease of polygenic inheritance, which affects 0.1-3% of the world population.^{1,2} It can cause significant psychosocial distress in patients. Among the treatments, mini punch grafting (MPG) proved to be one of the cheapest and easiest surgical approaches. A limitation of this technique is the possibility of cobblestone-like effect repigmentation.² We describe a new technique that can improve the appearance of repigmentation.

CASE REPORT

A 69-year-old woman with Fitzpatrick skin phototype V presented achromatic macules in the left frontotemporal and cervical region and upper limbs for 15 years. She underwent topical treatment with phototherapy with ultraviolet B and medium-potency corticosteroids, with improvement only in the lesions on the limbs. The condition remained stable for 12 years, when an autologous skin graft was performed, under local anesthesia, using the left cervical region as the donor site. We used a 2 mm punch at three points and implanted the material in areas of the same size and depth in the left frontotemporal region at 0.5 cm intervals. After one year, due to the presence of papules (cobblestone-like repigmentation) at the graft site, we decided to perform a new surgical intervention. The papules were incised close to the skin, and the domes were implanted at points in the left frontotemporal region (Figure 1). There was repigmentation of the recipient area in six months and aesthetic improvement of the donor site (Figure 2).

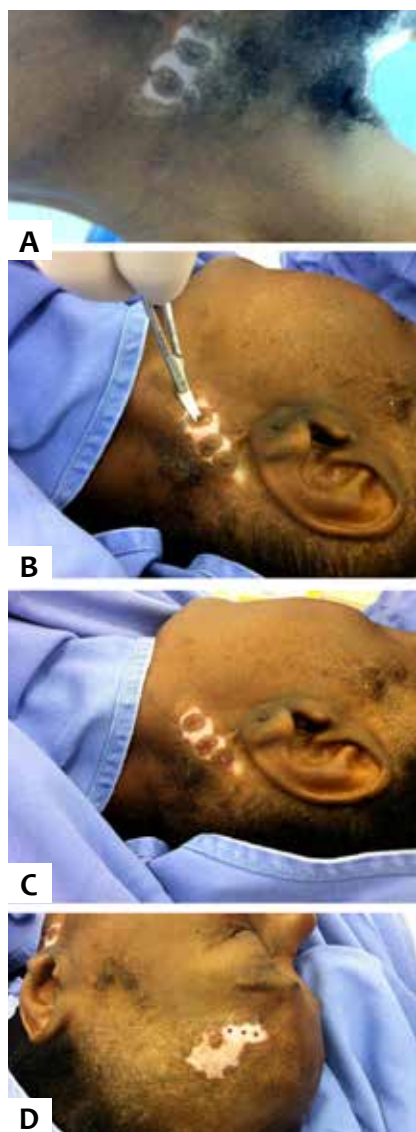


FIGURE 1:
Dermo-epidermal skin graft
A) Left cervical region: healing with a cobblestone-like effect;
B) Shaving of dermo-epidermal area;
C) Donor site after shaving;
D) Recipient site: left frontotemporal



FIGURE 2:
Result after six months
Satisfactory result after six months:
A) Donor site;
B) Recipient site

DISCUSSION

Vitiligo is a common acquired depigmentation disorder. The literature has proposed several theories about this disease's etiopathogenic mechanisms, including autoimmune, oxidative stress, neural, and viral theories.^{1,2} Behl and Batia first described the dermo-epidermal graft in 1973.³ The surgery aims to introduce melanocytes into depigmented vitiligo lesions. The mini graft proved to have the best applicability among the surgical techniques due to its simplicity, without the need for specialized instruments, and at low cost. Repigmentation rates are usually higher than 65% of cases.^{1,2,3,4} An important limitation of this technique is the possibility of cobblestone-like effect in repigmentation, present in about 30% of cases.² To avoid this effect, it is recommended to use a punch of up to 1.5 mm and a pigmented donor site similar in thickness to the recipient site's location. Some studies suggest benefits in making a recipient site 1 mm deeper and 0.5 mm narrower than grafts from donor sites.^{1,5}

As the presence of the cobblestone-like effect after the graft is not uncommon, this study proposed a new technique with the dermo-epidermal graft obtained by removing the papule formed post-mini punch grafting. The aesthetic result and the repigmentation were satisfactory in the donor and recipient sites. Larger samples are needed to support this technique. ●

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Association of fractional CO₂ laser and Q-switched Nd:YAG 1064 nm laser for tattoo removal

Associação entre o uso de laser de CO₂ fracionado ablativo e laser Q-switched Nd:YAG 1064nm para remoção de tatuagem

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ABSTRACT

Q-switched lasers are widely used for tattoo removal. A 48-year-old-woman with a black tattoo on the left thigh was submitted to ten sessions of Q-switched Nd:YAG 1064nm laser associated with fractional CO₂ laser, performed every two months. Six months after treatment end, we observed almost complete tattoo removal, besides improvement in skin texture. Q-switched lasers promote tattoo removal by a photoacoustic phenomenon. The association with fractional ablative lasers is an option for intensely pigmented tattoos or for tattoos with several colors, which are hard to remove.

Keywords: Laser Therapy; Lasers; Tattooing

RESUMO

Lasers Q-switched são os mais utilizados para remoção de tatuagens. Paciente feminina, de 48 anos, passou por consulta para remoção de tatuagem preta na coxa esquerda. Foram realizadas dez sessões de laser Q-switched Nd:YAG 1064nm associado ao laser de CO₂ fracionado com intervalos de 60 dias entre elas. Seis meses após a última sessão, houve remoção praticamente completa do pigmento, além de melhora da textura da pele. Lasers Q-switched removem tatuagens por meio do fenômeno fotoacústico. A associação com lasers fracionados ablativos é opção para tratamento de tatuagens com grande quantidade de pigmento ou em tatuagens com muitas cores, de difícil remoção.

Palavras-chave: Lasers; Tatuagem; Terapia a Laser

INTRODUCTION

The search for effective tattoo removal methods that do not result in the formation of unsightly tattoos when removing the pigment requires constant dermatologist improvement to deliver the best possible results. The tattooing art is ancient and consists of injecting pigment particles into the dermis to ensure a permanent tattoo. Approximately 5% of all people who get tattoos will want to remove them at some point in their lives.¹ Currently, Q-switched lasers are the most used for tattoo removal.²

CASE REPORT

A 48-year-old woman underwent consultation due to a black tattoo on her left thigh. The tattoo consisted of a completely black rectangular area, made to correct the skin graft donor area's coloration. According to the patient, the chosen color should simulate the skin tone of her skin phototype IV. However, the responsible tattoo artist made a mistake in selecting colors

Case report

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and inadvertently used black (Figure 1). The patient also presented scars on her thigh, knee, and right leg due to an accident. We decided to treat the areas with an ablative fractional CO₂ laser and Acroma Q-switched laser Nd:YAG 1064nm.

METHODS

We conducted a total of ten sessions. Before each session, topical and infiltrative anesthesia of the area to be treated with lidocaine 1% combined with vasoconstrictor diluted with saline (1:1) was performed due to the extent of the lesion and not to exceed the maximum dose of topical anesthetic considered safe.

From the first to the tenth session, the Acroma Q-switched Nd:YAG 1064nm laser, Etherea MX platform, Vydence Medical®, was associated (in the first session starting with the Acroma 1064nm laser, 5mm spot size, 900mJ in the first pass and 1200mJ in the second pass; the second session used the 3mm spot size with 600mJ energy, followed by the increase of energy to 900mJ in the third session, and to 1200mJ from the fifth session onwards), to the ablative fractional CO₂ laser (Sculptor, Vydence Medical®, 120mm spot size, fractional scanning mode, random, 90mJ energy, 0.3s interval, density 50MTZ/cm², X: 20mm Y: 18mm). From the second to the eighth session, the 800mm spot size was also associated (scanning type brush, interval stacking off, energy 80-100mJ frequency 150Hz, X: 10mm Y: 10mm).

The ablative fractional CO₂ laser was applied immediately after the Acroma Q-switched Nd:YAG 1064nm laser. The interval time between sessions was approximately two months, and the treatment started in November 2018.



FIGURE 1: The tattoo consisted of a completely black rectangular area, located on the left thigh

RESULTS

Six months after the last session, we observed practically complete pigment removal (Figure 2). Also, there was repigmentation of the area where the tattoo was performed, initially hypochromic, improvement of the skin surface texture, and thinning of the scars' thickness. The patient is satisfied with the results achieved.

DISCUSSION

So far, Q-switched lasers are the primary method available for tattoo removal, as they allow you to target the tattoo pigment and selectively remove it without damaging the surrounding tissue.^{3,4} The theory of selective photothermolysis, introduced by Anderson and Parrich in 1983, suggests that the interaction between skin and target chromophore allows selective destruction, even through heat, without destruction of adjacent tissues.⁵ However, for selective photothermolysis to occur, in addition to the appropriate wavelength, the pulse duration time must be shorter than the thermal relaxation time of the target structure.⁶ Thermal relaxation time (TRT) is the time required for the target structure to lose half the temperature rise at which it was heated by the laser, this time being variable for each chromophore.⁶ If the laser pulse duration exceeds the thermal relaxation time, damage to adjacent tissues will occur, evolving with burns and scarring.⁶ The average size of the tattoo pigment is 0.1µm, and the thermal relaxation time is 10ns, so the choice for Q-switched lasers, whose pulse duration is around nanoseconds.⁶ Depending on the color of the tattoo pigment, different wavelengths are indicated. For black tattoos, the length of 1064nm is the most suitable. Also, it allows greater penetration into the dermis, presenting the least risk of hypochromia and epidermal damage.³



Figure 2: Six months after the last session, virtually complete removal of the pigment is observed; there was repigmentation of the area where the tattoo had been performed, initially hypochromic, and improvement of the skin surface texture

The mode of operation of Q-switched lasers in the treatment of tattoos is through the photoacoustic phenomenon; when reaching the target (pigment contained in tattoos), the laser causes shock waves that make the target vibrate and “explode”, thus occurring the destruction of the pigment.⁷ The destroyed or altered pigment is then removed by phagocytosis by macrophages.⁶

The association with fractional ablative lasers is an option for treating tattoos, especially in those with a large amount of pigment, as in the case described, or in tattoos with many colors that are generally difficult to remove, as described in the article by Vanarase and collaborators.⁸ The study compared the isolated use of Q-switched laser with the use associated with CO₂ laser in

the ultra-pulsed mode to remove black tattoos. As a result, there was superiority in tattoo removal in the group that associated the two lasers, without increasing the rate of adverse events.⁸ Also, the association between lasers results in fewer sessions.⁸

CONCLUSION

The association between ablative and Q-switched fractional lasers in tattoo removal is an option to be considered in selected cases, given the difficulty in completely removing some of them. Professionals can still use the association only in the final sessions, when the pigment is already sparse, as there is the additional effect of assisting in the tattoo's healing. ●

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Case report

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Full thickness cross-shaped excision for rhinophyma in a patient with multiple comorbidities

Técnica de excisão em cruz para rinofima em paciente com múltiplas comorbidades

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ABSTRACT

Rhinophyma is a disfiguring and progressive disease of the nose with high prevalence. Despite being considered a benign condition, many patients have sought curative treatments due to aesthetic deformity and social stigmatization. There are several surgical techniques described in the treatment of this disease. This report aims to present a case that required an unusual therapeutic approach for the treatment of rhinophyma, given the patient's multiple comorbidities. The procedure was performed with no complications in the intra and postoperative period. The patient maintains an outpatient follow-up with a good long-term aesthetic result.

Keywords: Dermatologic Surgical Procedures; Rhinophyma; Rosacea

RESUMO

Rinofima é uma doença desfigurante e progressiva do nariz, com alta prevalência. Apesar de ser considerada benigna, muitos pacientes têm procurado tratamentos curativos devido a deformidade estética e estigmatização social. Existem diversas técnicas cirúrgicas descritas para o tratamento desta doença. O objetivo deste relato é apresentar um caso que necessitou de abordagem terapêutica pouco utilizada para tratamento de rinofima devido às múltiplas comorbidades do paciente. O procedimento foi realizado sem complicações no intra e pós-operatório, e o paciente mantém seguimento ambulatorial com bom resultado estético final a longo prazo.

Palavras-chave: Procedimentos Cirúrgicos Dermatológicos; Rinofima; Rosácea

INTRODUCTION

Rhinophyma is a disease of the nose characterized by hypertrophy of the sebaceous glands and proliferation of blood vessels and connective tissue.¹ Phymatous changes classically affect the lower two-thirds of the nose, and may also occur in the chin, forehead, or ear.¹ It is considered the most severe expression of the final stage of rosacea.¹

Clinically, rhinophyma manifests with an enlarged nose with irregular texture, enlarged pores, and telangiectasia.² These findings correspond, in histopathology, to sebaceous hyperplasia, infundibular dilation, and surrounding lymphohistiocytic infiltrate.² In more advanced stages, the nasal contours are distorted,

and there is a loss of demarcation between its subunits, which can compromise the respiratory airways.^{2,3}

Despite being considered a benign disease, many patients have sought curative treatments. Surgical methods are preferred over clinical treatment since the condition has a significant cosmetic deformity and social stigmatization.² The literature describes different surgical techniques for the treatment of rhinophyma, including the use of cold scalpels, electric scalpels and handles, and the CO2 laser.⁴

This report aims to present the cross-shaped excision technique for treating rhinophyma in a patient with multiple comorbidities. The surgery was performed without complications in the intra and postoperative periods, and long-term outpatient follow-up showed good aesthetic results.

CASE REPORT

A 68-year-old man presented progressive nasal enlargement with an irregular surface, enlarged pores, and telangiectasia (Figure 1). He reported aesthetic discomfort and social damage. We then opted for surgical treatment. As he presented several comorbidities, such as heart transplantation using immunosuppressants and antiplatelet agents, abdominal aortic aneurysm, and arterial hypertension, it was necessary to choose a quick surgical approach, with little bleeding and with a lower risk of complications. A vertical incision excised the excised skin at the nasal tip and dorsum and a horizontal cut in the alar sulcus, in a cross-shape, to reduce the hypertrophic tissue and the nose (Figures 2 to 4). It was followed by primary closure of the lesion, initially with internal suture approaching the surgical wound's edges and, after, external suture. There were no immediate complications or recurrence of the condition during the 36-month follow-up (Figure 5).



FIGURE 1: 68-year-old men with overall nose enlargement

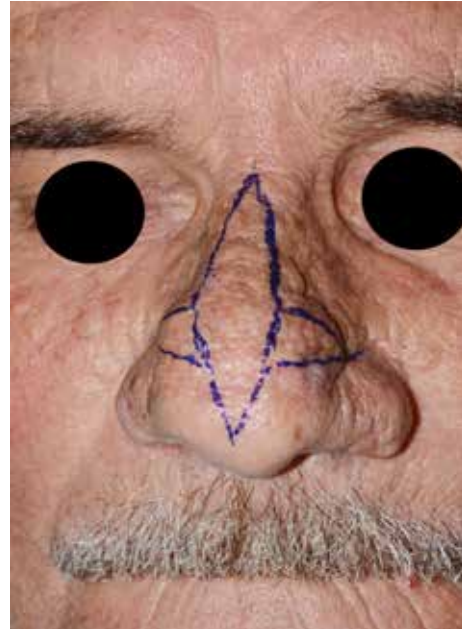


FIGURE 2: Surgical plan for nasal reduction



FIGURE 3: Excision of all layers in a cross-shape

DISCUSSION

Friedrich Dieffenbach presented the first descriptions of surgical treatment for rhinophyma in 1845. He excised the phymatous skin by making a vertical incision in the nasal tip and a horizontal incision in both nasal alar grooves, subsequently proceeding to the lesion's primary closure.^{4,5} This cross-shaped resection technique to reduce nasal volume is a quick procedure, with little bleeding and low risk of complications, with the advantage of removing part of the hypertrophic tissue, providing a



FIGURE 4: Internal suture to approach the edges of the surgical wound



FIGURE 5: Result after 36 months of primary synthesis

good aesthetic result for the patient and, most importantly, providing a fast recovery.⁵

The use of scalpels, cold or electric, and a tangential excision knife (for shaving) for superficial decortication of the rhinophyma and healing by secondary intention corresponds to a fast, low-cost, and relatively easy technique. However, it presents the main limitations of the excessive bleeding and, consequently, worse visualization of the surgical field and difficulties in modeling the affected region, besides care with dressings and successive returns.⁶ Since the reported patient had a previous history of platelet anti-aggregation and immunosuppression therapy by heart transplantation, we couldn't suspend the medication; thus this technique was not the first treatment choice.

The CO2 laser, on the other hand, is a good therapeutic option, when available, with adequate hemostasis and precision. However, it requires specially trained staff, prolonged time to perform the procedure, and has a high cost.^{6,7}

The risk of recurrence is variable and has been described in some series of cases, with good short-term cosmetic results but varying percentages according to the treatment performed

and the follow-up time.⁴ A series of 70 patients, published in 2016, pointed to a recurrence rate of 38% with the cold scalpel technique and secondary wound closure after 54-month follow-up.⁴ The reappearance of the phymatous changes occur by maintaining the pilosebaceous units, which provide the basis for reepithelization.⁴ In the case described, part of the hypertrophic tissue was excised in all its thickness, which could be a contributing factor for reducing long-term recurrences. When performed in two stages, a few months apart, the entire cosmetic unit's mechanical abrasion helps in camouflaging the surgical scar.⁵ Because it is not frequently used, there are no studies showing the rate of rhinophyma recurrence in patients undergoing the technique used in this report.

CONCLUSION

Cross-shaped excision and primary closure are a great option with very satisfactory results in patients with multiple comorbidities. A simple, safe, efficient, and not usually reported surgical technique for the treatment of rhinophyma. ●

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