Review Articles

Literature review: auricular disorders Part 2 – benign neoplasms

Revisão da literatura: afecções auriculares, parte 2: neoplasias benignas

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ABSTRACT

Knowledge, diagnostic ability and surgical skills for approaching diverse cutaneous conditions in the auricular region are parts of a dermatologist physician's daily routine. In the second part of this review article, will be approached the benign neoplasms of the auricular region, describing their particularities and ideal methods of treatment.

Keywords: Ear; Nevus; Cysts; Keloid

RESUMO

Faz parte do dia a dia dos dermatologistas o conhecimento, a capacidade de diagnóstico e a habilidade cirúrgica para várias afecções da pele na região da orelha. Na segunda parte deste artigo de revisão, serão abordadas as neoplasias benignas da região auricular, descrevendo-se suas particularidades e métodos ideais de tratamento.

Palavras-Chave: *Orelha; Nevo; Cistos; Queloide*

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CYSTS

Cystic lesions of the auricula are preferentially located in the space between the cartilage and the perichondrium, in the skin and subcutaneous tissue.

Endochondral pseudocyst of the auricula

Cartilaginous cyst that presents as a fluid collection between the cartilage and the perichondrium, resulting from chronic traumas of low intensity. In this case, the perichondrium not only detaches from the cartilage, but can also produce another cartilaginous tissue within which there is formation of the liquid content that originates the auricle pseudocyst. This cyst of firm consistency requires surgery: it must be incised, drained and curetted or cauterized using electrocoagulation or caustic substances, so that its walls can coapt.[1]

Epidermal cysts

They occur due to the proliferation of epidermal cells that produce keratin within the dermis. In some cysts it is possible to observe a central point corresponding to the obstructed pilosebaceous orifice that eliminates malodorous keratinous

material. They are usually well defined, mobile, non-adherent, superficial and of various sizes. Sometimes they become infected or inflamed. Treatment consists of complete excision, including the pilosebaceous orifice and the capsule.[2]

Pilar cysts

They occur preferentially in the scalp, but also arise in the retroauricular area or earlobe, while preauricular cysts (Figure 1) are generally congenital and might be associated with the preauricular fistula (Figure 2). They contain keratin in their core and are less common than the epidermal variant. Treatment includes the complete removal of the cyst and fistulous tract.

Multiple steatoma (sebocystoma)

A condition of autosomal dominant genetic nature, characterized by the presence of multiple cysts containing sebum. Lesions range from a few millimeters to two or three centimeters, yellowish, with an orifice through which material drains. Treated with exeresis.

Figure 1: Congenital pre-auricular cyst

Figure 2: Preauricular fistula

Milia *en plaque* is an entity characterized by an agglomerate of miliums in the form of erythematous and / or edematous plaques, with or without comedones, located in the retroauricular or periorbital region. They predominate in women and might regress spontaneously. Therapeutic options include manual extraction, topical tretinoin, electrocauterization, cryotherapy, excision and photodynamic therapy.[3]

Other cystic lesions: Sweat glands tumors, dermoid cysts, myxoid, eruptive velum cyst, milium and even follicular keratosis and perforating folliculitis can be found.

ORGANOID NEVI

Nevus verrucosus

It is a congenital lesion characterized by hyperplasia of epidermal structures. It arises as well-defined irregular papules or plaques, often multiple or generalized (ichthyosis histrix), [4] linear or unilateral (nevus unius lateralis) and sometimes inflamed (inflamed linear verrucous nevus). Differential diagnosis is performed *vis à vis* linear lichen planus, lichen striate, incontinentia pigmenti in its verrucous phase, linear psoriasis and linear porokeratosis. Nevus verrucosus may be accompanied by skeletal lesions, and organic, urologic and angiomatous malformations, as well as hypoplasia of underlying structures, constituting the epithelial nevus syndrome.

Treatment depends mainly on the location and extent of the lesions. Except for total or partial excision, other procedures – such as dermabrasion, electrocoagulation, electrodissection, cryosurgery and chemosurgery – offer only temporary relief, since there is a trend for the lesion to recurr.

Nevus sebaceous of Jadassohn

It is usually present from birth in the form of papules, sometimes shiny or grooved superficial plaques, eventually developing into single or multiple thick, verrucous or yellowish papillomatous lesions on the scalp and / or auricular region.

It should be treated with surgical excision due to the possibility of progression into papillary syringocystadenoma, basal cell carcinoma and other benign and malignant neoplasms.

Nevus comedonicus

Comedo-like papules (central part with a dark brown corneal plug) that arise grouped, in a unilateral or linear manner. They can progress into inflammatory reactions and scarring. Treatments comprise total or partial excision, topical retinoic acid or systemic isotretinoin.

Other nevi

Eccrine, apocrine, conjunctival and lipomatous nevi are rare, especially in the auricular region.

Eccrine nevi are well-defined areas with a history of localized hyperhidrosis or an orifice that drains liquid or mucoid substance and / or skin color plaques. Apocrine nevi may be associated with sebaceous nevi in the form of papules, nodules, or cysts. Conjunctival and lipomatous nevi are present from birth as nodules, plaques or tumors, usually of skin color, irregular and

tending to grow as the child develops. They can be treated with primary excision and suture or in partial exeresis.

BENIGN TUMORS

Epithelial tumors that may occur in the auricular region are: seborrheic keratosis, dermatosis papulosa nigra, verrucous dyskeratoma and acanthoma fissuratum. The first is frequent in the elderly, while the second often occurs in the dark skinned and Oriental Asian individuals. They usually arise in the periauricular regions.

Seborrheic keratosis is the most common benign tumor of the external ear, arising in the dermatological examination as a verrucous papule with adherent and greasy scale, more rarely in circumscribed plaques, ranging from light to dark brown in color. Can also be multiple, resulting from epithelial cells proliferation. It spreads with age, reaching the entire ear, including the external auditory canal. Treatment options are trichloroacetic acid, cryotherapy, and mild electrocoagulation.[5]

Pigmentary nevi

Pigmentary lesions of importance to the facial area that are rarely located in the auricular regions, are the nevus of Ota (nevus fuscoceruleus ophtalmomaxillaris), blue nevus and pigmented melanocytic nevi[6,7] (Figure 3).

Figure 3: Melanocytic nevus

Keloids

Normal scars usually increase in volume up until the second month, while hypertrophic ones increase for six months and progressively recede. In contrast, keloids present an anomalous healing process, increasing during 12 months or more, showing no signs of involution.

These lesions are very common in the auricles, especially in the lobule (Figure 4), resulting from trauma to the dermis with consequent fibrous proliferation. Its precise etiology is still unknown, however vascular-tissular, immunological, antigenic and interleukin related factors appear to be involved in its formation mechanism.[8] There is predilection for the dark skinned and Oriental Asian origin individuals, being equally distributed between genders and more common in individuals under 30 years of age. The most frequent sites of occurrence are: the deltoid, pre-sternal, preclavicular, scapular, pubic, nape and ears regions. Keloids are histologically characterized by wide bands of eosinophilic collagen. They can emerge after any type of trauma, however in the ear they are most often linked to the use of earrings. It arises as a cicatricial reaction that extends beyond the area of the trauma and does not recede with time. Generally pruritic, they consist of firm nodules with smooth surface. Lesions van be erythematous when occurring in occur in Caucasian individuals; hyperchromic or darkened and bright when occurring in dark skin or Oriental Asian origin individuals. Sometimes they present local heat or pain to pressure.

Therapeutic approaches for keloids are diverse. In the initial inflammatory phase, the most appropriate choice is intralesional injection with corticosteroids, occlusive dressings, silicone gels or sheets, or pressure earrings.

Figure 4: Keloids

In more stable phases, cryosurgery, radiotherapy, [9] intralesional injections with bleomycin sulfate[10] and 5-fluorouracil are indicated, among other possibilities.

There are studies describing the use of retinoids, interferon, methotrexate, D-penicillamine, colchicine and botulinum toxin, although without encouraging results.

Surgical removal, indicated in elevated and stable lesions, should always be followed by some of the therapeutic options mentioned above, due to the fact that as an isolated therapy, it leads to a high rate of recurrence.

For the prophylaxis of keloids in genetically predisposed individuals, procedures should be avoided and patients should be educated about the areas of greatest risk. When necessary ex-

ereses are performed in these patients, in addition to avoiding areas of risk, it is crucial that the incisions always agree with the skin's tension lines in order to cause as little trauma as possible to the area to be treated. Electrocoagulation and any elements that favor infection or inflammation, such as threads that cause an inflammatory reaction, should also be avoided. Suture should be carried out with the least possible traction.

Lipomas

Lipomas are well-defined slow growth nodules or tumors, usually painless, soft to palpation, mobile, skin-colored, or slightly bluish (angiolipomas). They can arise isolatedly or in multiple, measuring from 0.5 to 10cm or more. Despite being common, they are rare in the auricular region (Figure 5), occurring generally in the trunk, limbs and nape. Angiolipomas (with vascular component) can be painful. Lipomas are composed by mature adipose tissue lobes. Surgical excision is the treatment of choice. Although some surgeons approach lipomas with liposuction, there is a need for diagnostic certainty, when sarcomas, liposarcomas, myxosarcomas, and others must be excluded before implementing this technique.

ADNEXAL TUMORS

Tumors involving glands are rare, however they may arise in the ear due to the presence of a great amount of ceruminous glands – modified apocrine glands – which are found in the upper wall of the auditory canal. Their secretion combines with sebaceous secretion in the upper portion of the hair follicle forming a complex substance, the cerumen, which has antimicrobial action and forms a layer that lines the auditory canal, thus protecting it. The rate of migration of cerumen varies in each individual, sometimes leading to the obstruction of the auditory canal.

Cerumen can be removed via irrigation or with the assistance of instruments, the first method being simpler than the latter. Nevertheless, this procedure should only be performed after examination of the tympanic membrane. Irrigation can be carried out with syringes, after rectifying the canal by lifting and pulling the ear. Once direct vision of the canal is possible, the solution is carefully injected towards the wall, gently removing the cerumen with aid of the liquid flow. If irrigation is carried out hastily, the eardrum might be perforated, which would allow the solution to penetrate the middle ear, possibly causing otitis media. The liquid must be collected in a dome or kidney-shape dish. If the masses are large and compacted, in order to prevent trauma it is first necessary to soften them with topical compounds, which should be applied carefully due to the fact they cause irritation and external otitis. There are appropriate surgical instruments such as handles, curettes and forceps. They should be used carefully and, regarding children, general anesthesia is recommended due to the fact any sudden movement can lead to trauma.

Tumors involving the glands are extremely rare, nonetheless the fact that the ear presents a large number of ceruminous glands, makes it susceptible to the onset of benign and malignant tumors.[11] Among the benign ones are the ceruminous adenomas, apocrine cystadenoma, the Pringle-type sebaceous adenoma, trichoepitheliomas, and eccrine spiradenoma (Figure 6). Benign adenomas usually present obstructive symptoms of slow development and, being treated with local excision.

The other tumors may emerge in the face, neck and scalp regions, thus possibly located in the auricular and periauricular regions. These tumors are less frequent, however according to our experience they are not extremely rare. All of them may arise as papules (most often the chondroid syringoma, papillary syringocystadenoma and trichilemmoma), single cysts (chon-

Figure 5: Lipoma

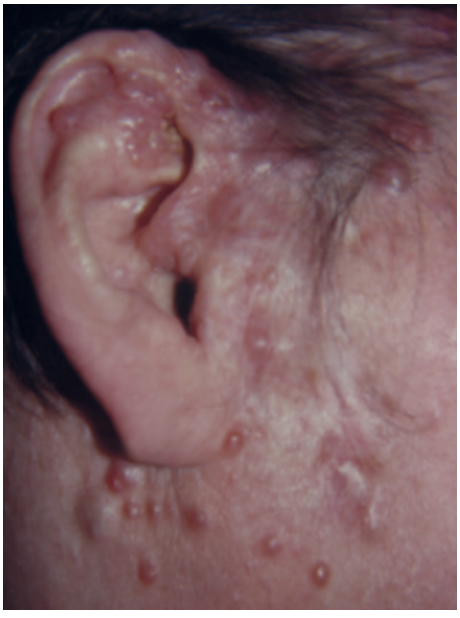

Figure 6: Eccrine spiradenoma

droid syringoma, hidradenoma papilliferum, apocrine hidrocystoma, calcifying epithelioma of Malherbe, trichofolliculoma), multiple cysts or in turban in the scalp (cylindroma), light and bluish coloration (hidradenoma and hidrocystoma), hardened or calcified (pilomatricoma). All of which have their diagnosis confirmed after anatomopathological evaluation, with surgical excision as the basic treatment.

Cylindroma

Also called Spiegler's tumor or turban tumor, it consists of a benign protruding, bright, normochromic, or erythematous lesions located in the head and neck, and might reach the ears (turban tumor). It has controversial, probably eccrine histogenesis, and is treated with exeresis.

OTHER SITUATIONS

Auricular appendices

These structures are not malignant, however they call for the investigation of some syndromes, such as those of Goldenhar and Wildervanck, vertebral or cardiac defects, anal atresia, tracheoesophageal fistula, and renal or limb anomalies. The excision should be performed according to the patient's wishes.

Petrified ear

The term *petrified ear* means calcification or ossification of the auricular pavilion, caused by traumatic processes, including thermal or physical damage, inflammation, and endocrine and metabolic disorders, such as hypercalcemia secondary to Addison's disease.[12] The ear becomes stiff and non-malleable. Diagnosis is carried out by imaging or histological examination, and underlying endocrine and metabolic disorders must be investigated for treatment.

CONCLUSIONS

Benign lesions located in the ear can be diagnosed clinically or through anatomopathological examination. Analyzing the probable diagnoses in advance, allows adequate surgical planning with a better prognosis for the patients. ●

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