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Keratoacanthoma centrifugum marginatum: report of a rare variant of keratoacanthoma

Queratoacantoma centrífugo marginado: relato de uma variante rara de queratoacantoma

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ABSTRACT

We report a case of a 72-year-old man who underwent surgical excision of a keratoacanthoma centrifugum marginatum (KCM) located on the left forearm. It is an uncommon variant of keratoacanthoma, which can assume large dimensions (up to 20 cm) and does not tend to spontaneous regression. The lesion is usually single, with raised borders, and has progressive centrifugal growth associated with central healing and atrophy. Due to its rarity and lack of pathognomonic histopathological features, KCM can be a diagnostic challenge for dermatologists and pathologists.

Keywords: Keratoacanthoma; Neoplasms; Skin neoplasms

RESUMO

Relatamos o caso de um homem, 72 anos, submetido à exérese cirúrgica de queratoacantoma centrífugo marginado (QCM), localizado no antebraço esquerdo. Trata-se de variante incomum do queratoacantoma, que pode assumir grandes dimensões (até 20cm) e não tende à regressão espontânea. A lesão geralmente é única, com bordas elevadas, e tem crescimento centrífugo progressivo, associado à cura central e atrofia. Devido a sua raridade e ausência de características histopatológicas patognomônicas, o QCM pode ser um desafio diagnóstico para dermatologistas e patologistas.

Palavras-chave: *Ceratoacantoma; Neoplasias; Neoplasias cutâneas*

Case report

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INTRODUCTION

Keratoacanthoma centrifugum marginatum (KCM) is a rare variant of keratoacanthoma, first described by Miedzinski and Kozakiewicz in 1962.[1] Typically, the lesion is a single plaque with raised edges, with peripheral centrifugal growth, associated with simultaneous central scarring.[2]

KCM is composed of keratinizing squamous cells originating from pilosebaceous follicles.[1,3] It grows rapidly and, unlike classic keratoacanthoma, does not tend to spontaneous regression, in addition to being able to assume large dimensions (up to 20cm).[3,4] Although locally destructive, KCM is a tumor with a low risk of metastasis, as are other keratoacanthoma lesions.[5]

CASE REPORT

A 72-year-old man, a smoker, without other comorbidities or use of medication, sought care due to the appearance, three months ago, of a lesion on the posterior surface of his right forearm, which was rapidly growing and painless. The patient denied a family history of similar injuries or local trauma. He denied occupational exposure to chemicals or prior treatment for the lesion. He reported intense photoexposure during adulthood without photoprotection measures.

Dermatological examination showed an erythematous plaque, not infiltrated on palpation, with an ulcerated and atrophic center, measuring 7 x 4 cm, with raised, well-defined, discretely hyperkeratotic edges (Figure 1A) on the posterior surface of the right forearm. There was no axillary lymph node enlargement on palpation.

The patient underwent surgical excision of the lesion with a safety margin (1 cm). The resulting surgical defect was closed through total skin grafting, using triangular redundancy of adjacent skin ("dog ear") – local graft (Figure 1B).

The histopathological examination showed an epidermis with irregular acanthosis and hyperkeratosis at the expense of ortho and parakeratosis. The squamous cells exhibited intense loss of polarity and cytoarchitectural orientation, with severe cytological atypia and cellular anaplasia, without infiltrating or surpassing the basement membrane. The dermis showed moderate basophilic collagen degeneration and diffuse mononuclear infiltrate. Given the clinical and histopathological findings, the definitive diagnosis was keratoacanthoma centrifugum marginatum with free resection margins (Figures 2A and B).

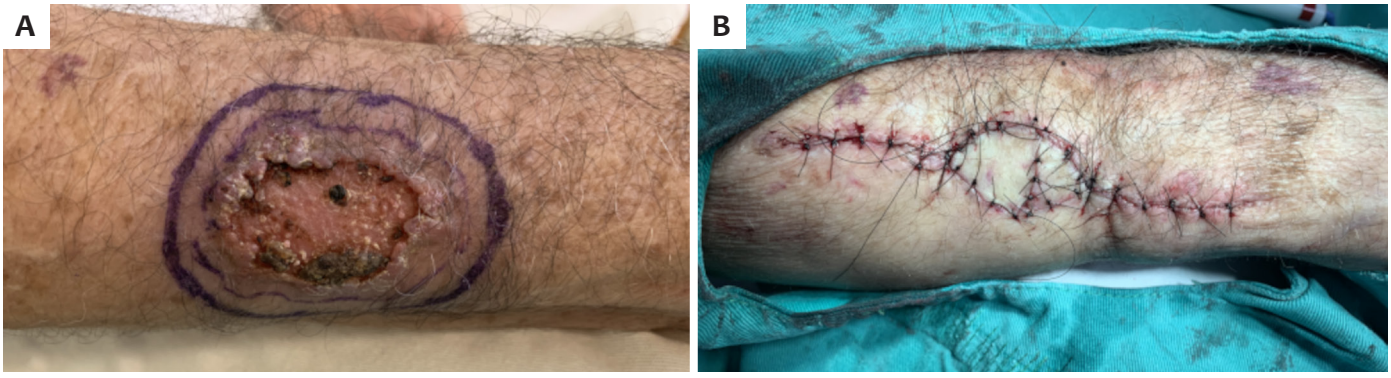

Figure 1: A - Erythematous plaque, with an ulcerated center, well-defined and infiltrated edges, with discrete hyperkeratosis. **B -** Surgical closure by skin grafting after excision of the lesion

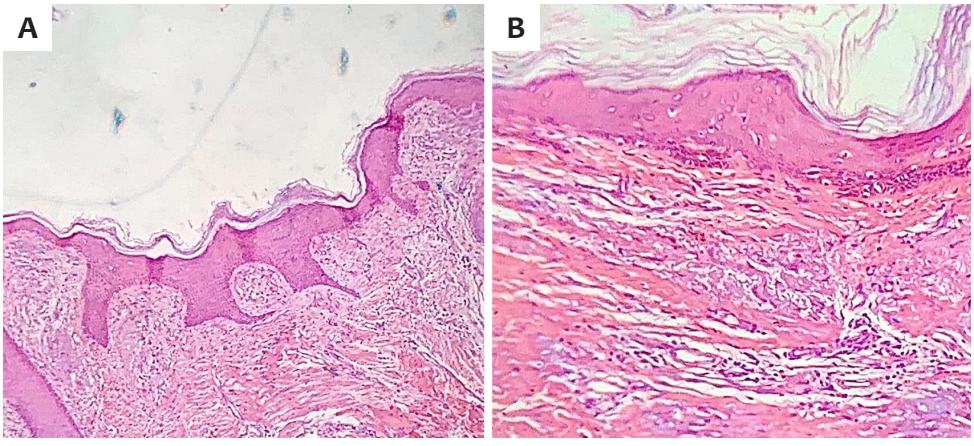

Figure 2: A - Histopathological examination of the edge of the lesion, with epidermis showing irregular acanthosis and hyperkeratosis, at the expense of ortho and parakeratosis (Hematoxylin & eosin, 40x magnification). **B -** Squamous cells with an intense degree of loss of polarity and cytoarchitectural orientation, intense cytological atypia and cellular anaplasia, without infiltrating or exceeding the basement membrane. The dermis showed moderate basophilic collagen degeneration and diffuse mononuclear infiltrate (Hematoxylin & eosin, 100x magnification)

On the 21st postoperative day, the patient presented deepithelialization in the central portion of the graft (Figure 3), but it progressed to spontaneous healing after two weeks. The patient continues to be under clinical follow-up at the institution's Dermatology Service for 36 months without recurrence (Figure 4).

DISCUSSION

Keratoacanthomas are fast-growing tumors composed of keratinocytes originating from the follicular infundibulum epithelium.5 KCM is a rare variant of keratoacanthoma, whose main characteristics are generally a single lesion, which can be large (up to 20 cm), with raised edges, and progressive centrifugal growth associated with central healing and atrophy. Unlike classic keratoacanthoma, it does not tend to regress spontaneously.[3,5] Its incidence is not known.[6]

The sites most frequently affected by KCM are the head and neck, and the upper and lower limbs of adults.4 The risk factors for its development are the same as those that lead to the emergence of classic keratoacanthomas or squamous cell carcinomas: exposure to ultraviolet radiation, smoking, and exposure to other chemical carcinogens.[3] There are five reports of cases of KCM that developed in a trauma area.7 The literature brings together around seven cases of individuals with multiple KCM injuries; all others are reports of isolated injuries.8 Although uncommon in this age group, there is a report of a five-year-old boy with multiple KCM injuries in the lower limbs.[8]

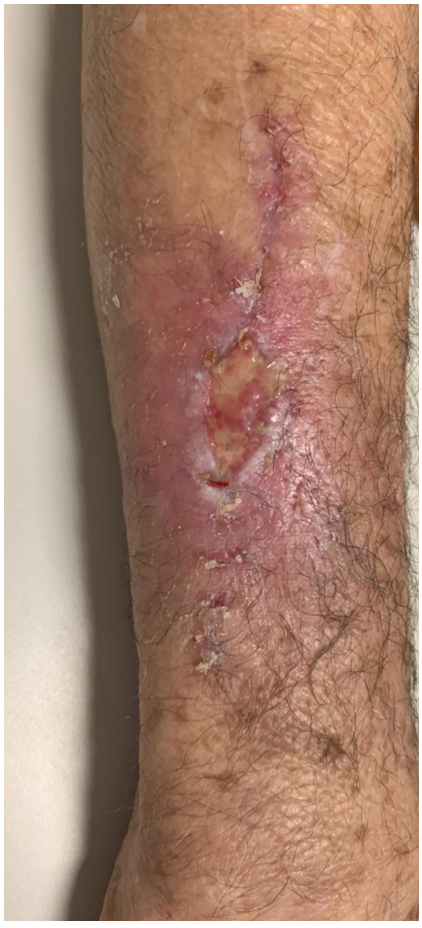

Figure 3: Surgical wound in the healing process (21st post-operative day)

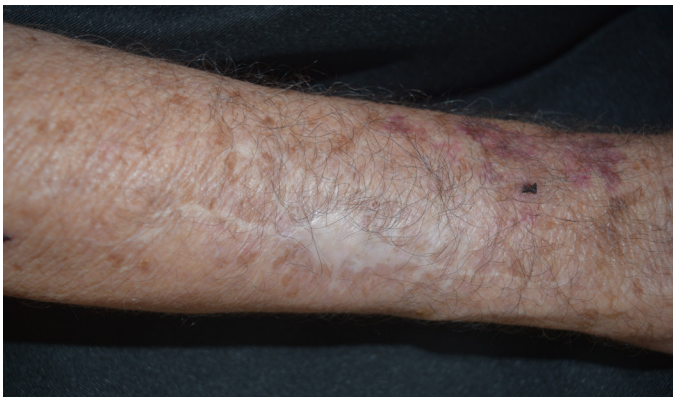

Figure 4: Late postoperative period (36 months after tumor excision and local reconstruction), without clinical signs of recurrence. Scar with good aspect

The main diagnostic differentials are fungal infection, atypical mycobacteriosis, botryomycosis, pyoderma gangrenosum, squamous cell carcinoma, and cutaneous lupus erythematosus.[4] The definitive diagnosis of KCM can be a challenge for the dermatologist, requiring a high level of clinical suspicion. An incisional biopsy may not be able to define the diagnosis, requiring serial biopsies or a definitive diagnosis after analysis of the complete surgical specimen.[9,10]

Histologically, the KCM margin tends to present the classic findings of keratoacanthoma, while the central portion has atrophic or healing tissue.[6] There is a central crater filled with keratin, with prominent edges and a clear delineation between the tumor nests and the stroma, in addition to the absence of stromal anaplasia and desmoplasia.[11]

The gold standard in the KCM treatment is surgical excision, generally by conventional surgery. Micrographic surgery tends to be restricted to cases of recurrent KCM, in lesions with poorly defined borders, of extremely rapid growth, or when preserving healthy skin is mandatory.6 In very large lesions, or when surgical approaches are not possible, oral retinoids, intralesional methotrexate, phototherapy, or bleomycin are therapeutic options.[9,12,13]

CONCLUSION

KCM is a rare variant of keratoacanthoma, and its diagnosis can be a challenge for the attending physician due to the absence of pathognomonic histopathological characteristics. Usually, it is a single, large lesion (up to 20 cm) with raised edges and progressive centrifugal growth associated with central healing and atrophy. It does not tend to regress spontaneously, and surgical excision is the therapy of choice whenever possible, given the size and location of the lesion. ●

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