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CASE REPORT

Abrikossoff tumor: rare dermatosis in a case with epidemiological and therapeutic specificities

Tumor de Abrikossoff: dermatose rara em caso com especificidades epidemiológicas e terapêuticas

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Abstract

Granular cell tumor is a rare benign neoplasm, which preferentially affects the oral cavity, more common in adults and rare in children. This report is about a 9-year-old patient with a painful periungual nodule of the fifth finger of the right hand, with two years of evolution. The anatomopathological examination revealed an Abrikossoff tumor, without bone involvement on X-ray and positive markers on immunohistochemistry. Due to the location of the lesion, Mohs micrographic surgery was chosen in order to have greater tissue preservation and local functionality. There was no recurrence of the tumor, with resolutive surgery and satisfactory aesthetic results.

Keywords: Mohs surgery. Granular cell tumor. S100 proteins. Abrikossoff tumor.

Resumo

O tumor de células granulares é uma neoplasia benigna rara, que afeta preferencialmente a cavidade oral, mais comum em adultos e rara em crianças. Este relato trata de paciente de 9 anos com história de nodulação periungueal dolorosa do quinto dedo da mão direita, com dois anos de evolução. O exame anatomopatológico revelou tumor de Abrikossoff, sem envolvimento ósseo na radiografia e marcadores positivos na imunohistoquímica. Devido à localização da lesão, optou-se pela cirurgia micrográfica de Mohs, para maior preservação tecidual e funcionalidade local. Não houve recidiva do tumor, com cirurgia resolutiva e resultado estético satisfatório.

Palavras-chave: Cirurgia de mohs. Tumor de células granulares. Proteína s100. Tumor de abrikossoff.

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Introduction

Granular cell tumor (GCT) or granulosa cell myoblastoma, described by Abrikossoff in 1926, is a rare benign neoplasm that probably originates from Schwann cells[1]. It is believed to occur due to the altered metabolism of these cells, a theory reinforced by the constant presence of the S-100 protein in immunohistochemistry (IHC). It affects predominantly individuals between the third and sixth decades of life (30-60 years), predominantly females (1.8-2.9:1)[2], with phototypes V to VI, being a rare diagnosis in the pediatric population. It represents only 0.5% of all soft tissue tumors[3].

In the general population, GCT most frequently affects the tongue and oral cavity (30–50%)[4] and can also affect other locations. In children, it most commonly affects the oral mucosa and extremities. The lesions normally appear as solitary, slow-growing, asymptomatic, or painful nodules with a firm consistency. The clinical differential diagnoses are fibroma, hidradenoma, and lipoma[4]. Multiple nodules in the same person have been reported (5-16%), associated with congenital syndromes.

Around 1-2% of cases progress to malignancy with a poor prognosis. There are reports of regional or distant metastases with multiple organs involved[4]. The clinical parameters for malignancy include size greater than 4 mm, invasion of adjacent tissues, and rapid progression[4]. Malignant tumors are clinically larger and often located in subcutaneous tissue.

The histological examination demonstrates pseudoepitheliomatous hyperplasia. In the dermis, there are large polygonal or round cells with small central vesicular nuclei and abundant eosinophilic granular cytoplasm. These granules are lysosomes or components of the Golgi complex. Positive Periodic Acid-Schiff (PAS) granules and resistant diastasis indicate the presence of myelin. The cells may present ovoid-pustular Milian bodies, specific to the disease, but found in few cases, normally in older lesions (they represent the accumulation of granules inside the lysosomes in cells that have already lost mitochondria and the endoplasmic reticulum). Perineural and perivascular involvement are common and are not markers of a worse prognosis. The criteria for malignancy include the presence of necrosis, elongated and thin cells, large vesicular nuclei, increased mitotic activity, nuclear pleomorphism, and increased nucleus-cytoplasm ratio[4]. Tumors that present three or more of these histological characteristics are considered malignant[2]. Finally, immunohistochemical findings are not specific, with positivity for S100 protein, CD8, vimentin, myelin basic protein, and neuron-specific enolase being more common.

Complete excision of the tumor, with safety margins, is recommended due to local recurrence[4]. For cases with atypical histological characteristics, lymph node dissection is recommended[4]. Mohs micrographic surgery (MMS) can be an effective tool in invasive tumors or tumors that occur in areas of functional or aesthetic impairment (where maximum tissue preservation is required)[4]. During MMS, special markers may be required, such as S100, as perineural tumor invasion is difficult to detect by conventional hematoxylin-eosin staining.

The prognosis of GCT is good. Cases of recurrence are generally related to incomplete excision of the tumor (15% of cases). Progression to malignancy is rare (less than 2% of the cases)[4]. This dermatosis must be followed due to recurrence or development of greater aggression.

Case report

Male patient, 9 years old, phototype IV, complaining of the periungual hardening nodule on the tip of the fifth finger of the right hand presenting slow growth, local pain, and occasional itching for 2 years.

Dermatological examination revealed a normochromic, hardened nodule on the distal phalanx of the fifth right finger with slight nail changes (Fig. 1).

A spindle biopsy was performed, which demonstrated an epidermis without atypia and, in the dermis, a proliferation of cells with a large, frankly acidophilic cytoplasm and vesicular, central nuclei, without atypia (Fig. 2). There were no signs of malignancy.

Immunohistochemical examination was positive for S-100 protein, CD68, inhibin, and SOX-10 and negative for the markers Ki67 and epithelial membrane antigen (EMA).

Given the lack of definition of the tumor margins, with the aim of preserving tissue in a noble area and the high degree of clinical infiltration, the Mohs technique was chosen (Fig. 3).

Resection was performed in two phases, with lateral and deep involvement in the first phase without signs of malignant transformation. There was no involvement of the nail matrix, only the cutaneous portion of the distal half of the distal phalanx of the affected finger (Fig. 4). Finally, the reconstruction of the wound was made with a skin graft from the contralateral arm. The 1-year clinical follow-up of the patient was carried out without signs of recurrence after surgery and a good aesthetic result (Fig. 5).

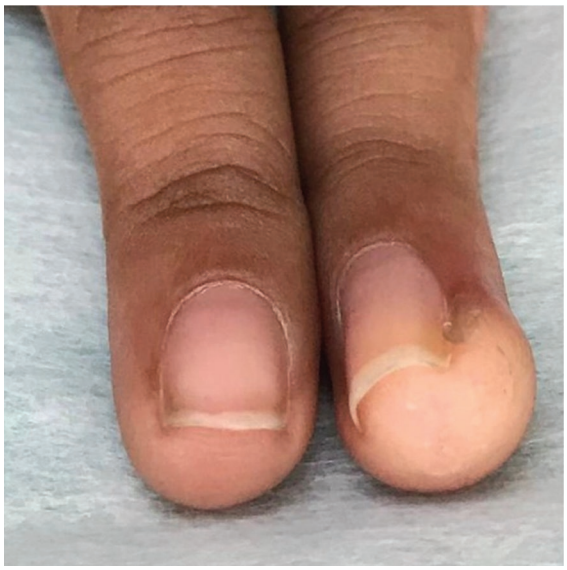

Figure 1. Clinical image showing a nodule on the tip of the fifth finger with minor deformity of the nail plate.

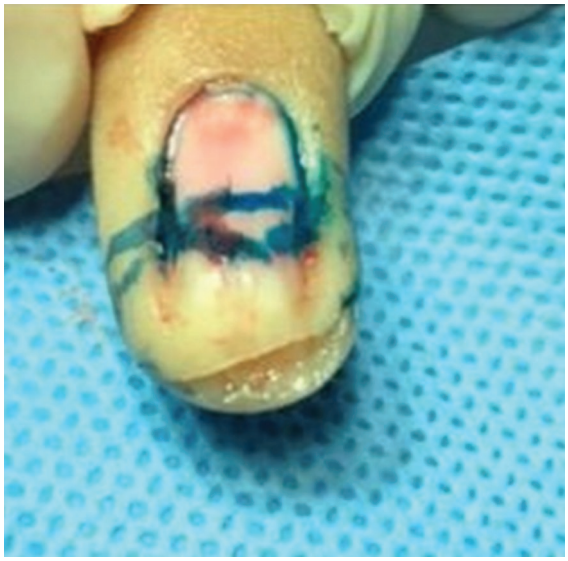

Figure 3. Marking of tumor and excision margins.

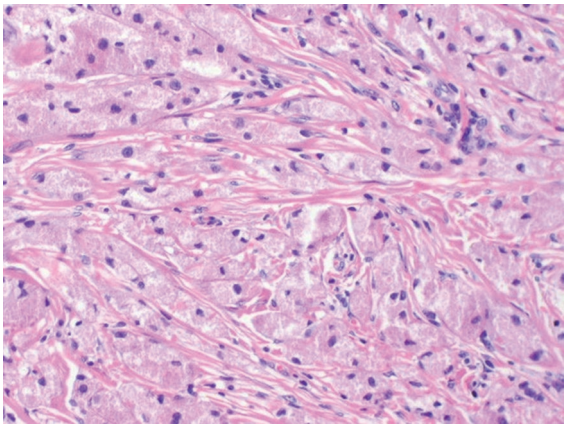

Figure 2. Histopathology showing large polygonal or round cells with small central vesicular nuclei and abundant eosinophilic granular cytoplasm (H&E).

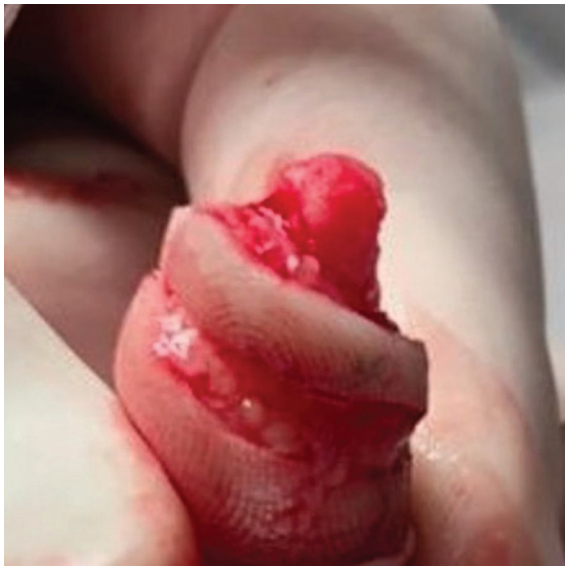

Figure 4. Clinical image during the surgery showing the finger after the second phase of MMS.

Discussion

The presented case of a GCT brings particular aspects regarding age, gender, and location[2], parameters that are not within the typical epidemiological profile of this disease.

There are 14 case reports of GCT in children, aged between 5 and 12 years. Locations included extremities, palms, soles, back, and anterior thorax[3]. In two cases, the Mohs technique was performed without signs of recurrence, even though in one of them a small margin was revealed.

GCT with nail involvement is very rare and only seven cases have been described in the literature (one in the index finger and six in the hallux). Nail injuries, which are often painful, can occur depending on the structure affected. If proximal, a longitudinal groove may appear

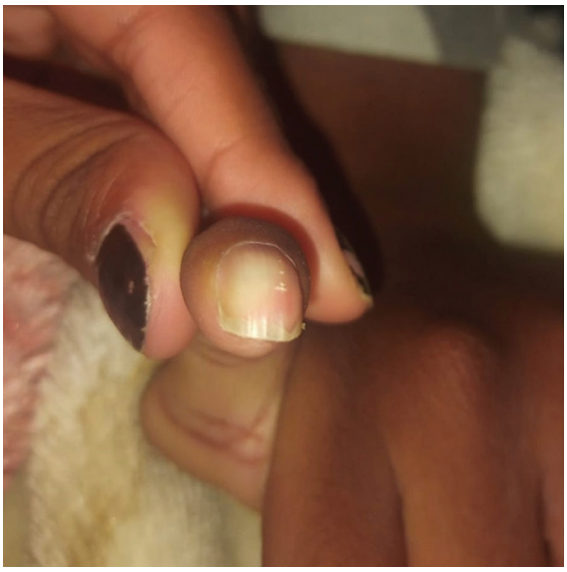

Figure 5. Good final clinical and aesthetic result 1 year after surgery.

on the nail plate. If distal, it generally presents as a warty appearance[1]. Dupin et al. described a case with distal subungual hyperkeratosis in the hallux that recurred after the first surgical approach, requiring expansion in a second approach. Furthermore, there is a report of MMS performed in a patient with extensive involvement of the labia majora of the vagina, with the aim of sparing tissue[4].

Due to the success provided by this technique to this child, it is possible to use this technique for other tumors with a high recurrence rate, malignant potential, or involvement of noble and deep structures. The two main objectives of MMS are resolution of the injury and tissue preservation.

Finally, MMS was chosen in the case reported due to its location, difficult delimitation, and risk of malignancy and recurrence. As it was a child, the preference for the graft was due to its practicality in a less time-consuming surgery. It was believed that there would be a need for some repair afterward, but it was not necessary, and the patient evolved with practically no nail dystrophy, just a slight increase in the convexity of the nail plate, with no impact on his quality of life.

Abrikossoff's tumor, despite its rarity, should be considered as a differential diagnosis for both the dermatologist and pathologist. Even though it has some specific histological characteristics, IHC is necessary for a diagnostic conclusion. Its treatment must be assertive, as it presents uncertain behavior regarding potential malignancy.

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Conflicts of interest

None.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

References

1. Dupin N, Carlotti A, Baran R, Moulonguet I. Abrikossof granular cell tumor at the nail unit: an exceptional location. Ann Dermatol Venereol. 2021 Sep;148(3):195-7.
2. Stemm M, Suster D, Wakely PE Jr, Suster S. Typical and atypical granular cell tumors of soft tissue: a clinicopathologic study of 50 patients. Am J Clin Pathol. 2017 Aug 1;148(2):161-6.
3. Olayiwola O, Hook K, Miller D, Maguiness S. Cutaneous granular cell tumors in children: case series and review of the literature. Pediatr Dermatol. 2017 Jul;34(4):e187-90.
4. Kim HJ, Lee MG. Granular cell tumors on unusual anatomic locations. Yonsei Med J. 2015;56(6):1731.