Multicentric granular cell tumor: a rare presentation

Tumor de células granulares multicêntrico: uma apresentação rara

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ABSTRACT

The granular cell tumor, also known as Abrikossoff tumor, is a rare neoplasm with controversial etiology that usually arises as a solitary benign lesion. The authors describe a case of multicentric granular cell tumor in a 13-year old patient treated with surgical excision.

Keywords: granular cell tumor; neoplasms, soft tissue neoplasms; S100 proteins

RESUMO

O tumor de células granulares, também conhecido como tumor de Abrikossoff, é neoplasia rara e de etiologia controversa, que se apresenta geralmente como lesão benigna solitária. Relata-se um caso de tumor de células granulares multicêntrico em paciente de 13 anos tratado com exérese cirúrgica.

Palavras-chave: *tumor de células granulares; neoplasias de tecidos moles; proteínas S100*

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INTRODUCTION

The granular cell tumor (GCT) – or Abrikossoff's tumor – was first described in 1926 by the Russian pathologist Alexei Ivanovich Abrikossoff. Some authors have suggested that these tumors represent approximately 0.5% of all soft tissue tumors, with a prevalence of 0.019% to 0.03% of all neoplasias.[1] It usually arises as a solitary nodular lesion that is painless, slow growing and benign, although multicentric (10%-25%) and malignant (1%-2%) variants have also been reported.[2]

These tumors mainly involve the cervicofacial region, particularly the tongue and palate, however they can affect the skin and other organs such as the esophagus, larynx, trachea, thyroid, with few reports of it occurring in the extremities.[2,3]

The granular cell tumor is most commonly found in middle-aged dark skin women.[2, 4-7] Its histogenesis is controversial, with several authors implicating diverse cell types in its origin, including muscle cells, histiocytes, fibroblasts, nerve sheath cells and undifferentiated mesenchymal cells. The neuroectodermal origin is generally considered due to the GCT's reactivity for neural markers like S-100.[1, 3-5, 7, 8]

CASE REPORT

A 13 year-old mulatto female patient presented nodular lesions that had emerged on the body approximately three years before. The lesions affected the scalp (Figure 1), right flank, right thigh, left palmar and right plantar regions (Figure 2), the latter being painful on deambulation.

The dermatological examination revealed nodular lesions, the largest located on the medial side of the right thigh, measuring 1cm and with hyperpigmentation in the overlying skin. The other lesions were normochromic. The examination of the oral cavity evidenced two yellowish nodular lesions on the left side of the tongue, (Figure 3) painless on palpation and measuring 1cm and 0.5 cm.

The surgical excisions of the lesions on the right flank, left palm and right thigh were carried out, all with granular cell tumor histology. The histological section of the skin allowed the observation of an intense infiltration of the dermis by polygonal cells with granular eosinophilic cytoplasm, with small dark nuclei (Figure 4). The PAS staining color was positive for the intracytoplasmic granules.

The patient is currently under clinical follow up for the other lesions, with absence of recurrence of the excised lesions.

Figure 1: Normochromic nodular lesion with firm consistency on the scalp

Figure 3: Two yellowish nodular lesions on the left side of the tongue

Figure 2: Normochromic nodular lesion in the plantar region of the right foot

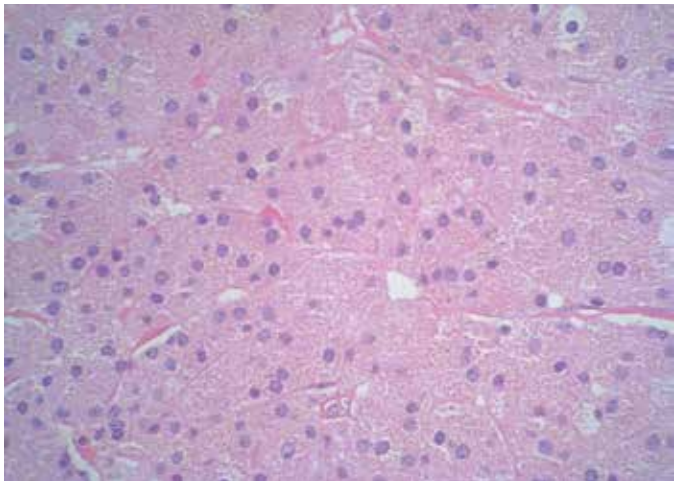

Figure 4: Histology of the lesion in the right flank. It is possible to observe clusters of polygonal cells with granular eosinophilic cytoplasm and rounded basophilic nuclei (HE x 200)

DISCUSSION

GCT is a rare neoplasm of uncertain etiology and histogenesis, occurring as a solitary, asymptomatic, slow growing, normochromic or brownish lesion. This tumor usually measures less than 3cm in its longest diameter, nevertheless it can be considerably larger than that. 3 Its preferred locations are the head and neck regions (50%), the tongue (35%) and the vulva (5.3%).[9] Although in most cases the lesion is solitary, nearly 25% of patients may have multiple lesions, as observed in the present case.[2,5]

The clinical diagnosis of GCT is difficult since the lesions are often unspecific and its identification usually occurs only through histological examination.[3] The clinical differential diagnosis includes dermatofibroma, adnexal tumors, compound melanocytic nevi, nodular prurigo, dermoid cyst and seborrheic keratoses.[7]

Histologically, the dermis contains a poorly circumscribed nodule constituted by polygonal cells pale in color, which can infiltrate the adjacent dermis. The cells have abundant cytoplasm, with PAS-positive granulation, slightly eosinophilic, with round dark nuclei.[3,7] Mature GCT (the classic form), with presumed Schwannian origin, are described as positive for S-100 protein and neurospecific enolase.[3]

Malignant granular cell tumors are perhaps the rarest of all soft tissue tumors, representing a percentage that varies from 1% to 2% of cases, with reports of occurrence in diverse locations.[2,3] The clinical features suggestive of malignancy include large size (> 4cm), necrotic and hemorrhagic areas, fast growth and invasion of adjacent organs.[9] The metastasization of this tumor to lymph nodes and lungs is common and highly aggressive, unresponsive to treatment and eventually lethal.[3]

A third GCT type has benign pathological characteristics, however presents malignant clinical behavior. Malignant GCT with benign histologic appearance are usually identified only after the metastatic and lymph node spread. Notwithstanding its histologic appearance, the GCT's biological potential is uncertain.[3] The treatment of choice is the complete surgical excision.[1-9]

If incompletely removed, this tumor has a high rate of local recurrence.[2-5,7,9] The use of radiotherapy and chemotherapy is recommended in the treatment of malignant forms of this tumor.[4,6] The present case report describes a clinical picture in a young patient, outside the most affected age group and with multiple lesions – some of them in the distal ends – constituting an unusual presentation of this rare tumor.

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