Case for diagnosis

Caso para diagnóstico

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CASE REPORT

A 23-year-old female patient with no comorbidity reports a lesion in the cubital fossa of the superior left arm which has been present for two years. Initially, the lesion appeared as a purulent hyperchromic papule that evolved into a nodular lesion with posterior ulceration and elimination of a liquid substance (Figures 1 and 2).

She reported previous use of topical neomicin with no healing. On dermatological examination, a 1.5 x 1.0 cm ulcerated tumor with fibrous consistency, a hyperchromic halo and clean background could be observed in the left cubital fossa. No lymphadenomegaly was found.

The following diagnostic hypotheses were posed: leishmaniasis, squamous cell carcinoma, lymphoma, sporotrichosis and dermatofibrosarcoma. An excisional biopsy was performed. Histopathological sections revealed a dermal lesion, formed by big cells, with an abundant granular cytoplasm and a small hyperchromatic nucleus without abnormalities. Malignant signs were absent and the borders were clear of tumor (Figure 3).

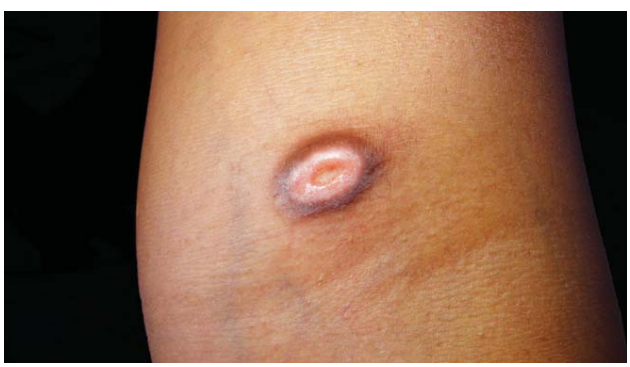

FIGURE 1: Nodular lesion with ulceration in the cubital fossa

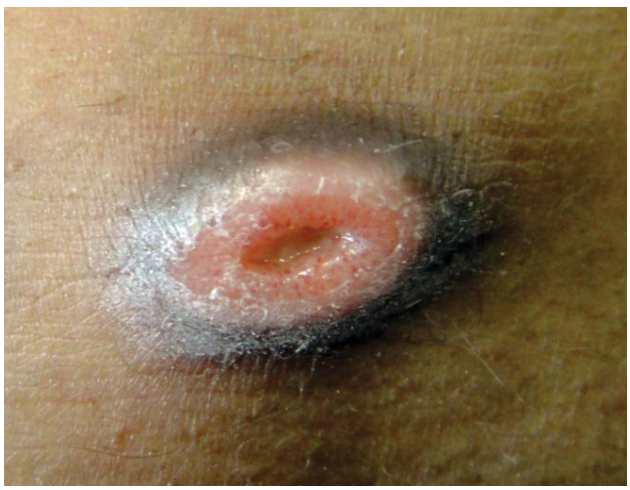

FIGURE 2: Details of the lesion

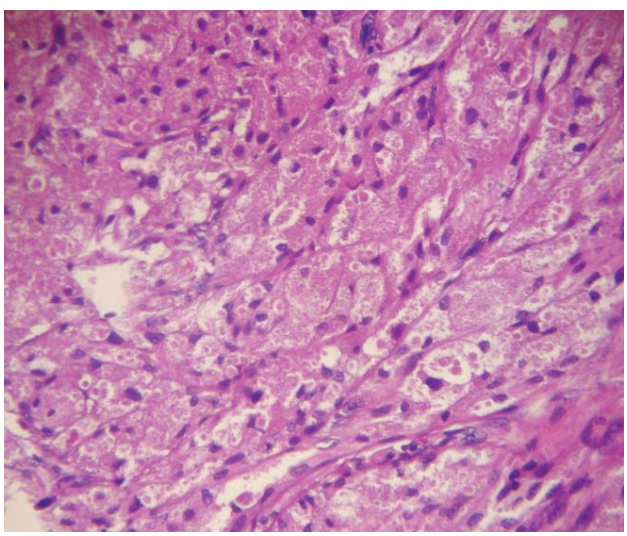

FIGURE 3: Dermal lesion with granular cytoplasm

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DISCUSSION

Granular cell tumor (Abrikossoff´s tumor) was first described in 1926 by Abrikossoff as a myoblastoma. That description was probably due to the high frequency of the tumor on the tongue, where it infiltrates between the striated muscle tissue sections and blends in, confusing itself with the muscle.[1] Its origin is believed to be neuroectodermal.

It is a rare tumor that preferentially occurs in the cervicofacial segment, especially on the tongue, but it can also affect the skin and other organs, like the esophagus, larynx, trachea, parotid gland, thyroid and lungs.[2,3] It has also been described in the vulva, bladder, thoracic cavity, perianal area, breasts, stomach and appendix.[4-5] The authors have not found reports of this tumor on the arms in the medical literature. Women are usually more affected than men, and the preferred age is between the second and sixth decades.[6]

The tumor manifests itself clinically as an isolated, firm and well-defined lesion. It can be sessile or pedunculated and ranges in size from 5 to 30mm. There can be multiple lesions (10 to 25%), which rarely reach bigger dimensions. Most of them are diagnosed as slowly growing papulo-nodular lesions smaller than 3cm whose color may vary from pinkish to grey and even brown.[6] The lesions can ulcerate and develop a hyperchromic halo very similar to the one on the patient described in this article.

Histology reveals a nodule composed of polyhedral cells that organize themselves in stripes and infiltrate the dermal collagen and subcutaneous fat. The cytoplasm is pale and contains numerous small acidophilic structures, which are lysosomes.[7] The nucleus is small, oval and centered. Mitosis is uncommon.

The epithelium that covers the tumor can show pseudoepitheliomatous hyperplasia, which can be mistaken for squamous-cell carcinoma in shallow biopsies.[8] Those hyperplasias are oblong stripes of squamous cells that extend down into the tumor tissue, forming columns. Occasionally, the tumor can invade the epidermis, in which case differentiation from melanoma can be difficult. Both tumors are strongly positive for S-100, but HMB-45 and melan-A are negative in the case of granular cell tumor.[7-8] The differential diagnosis also includes leiomyoma and angiosarcoma, which can also have super populations of cells with a granular cytoplasm. Cases of malignant (1%) granular cell tumors have been reported. Suspicion should arise when bigger tumors, located deep in the subcutaneous tissue and showing cellular augmentation, atypias, mitosis figures and necrosis, are frequently found on histopathology. Nevertheless, metastasis is the only definitive proof.[4-6]

Excision is the treatment of choice and, when incomplete, can lead to local relapse.[9] ❑

Abstract: Granular cell tumor (Abrikossoff´s tumor) is a rare benign disease that preferentially affects the cervicofacial segment. It is usually a solitary nodule that may ulcerate and present pearly infiltration on the borders, while keeping a clean background and a hyperchromic halo. This paper describes the case of an ulcerated granular cell tumor on an unusual location, which reinforces the necessity of including this tumor in the differential diagnosis of nodular-ulcerative skin lesions.
Keywords: Granular cell tumor; Neoplasms; Schwann cells

Resumo: O tumor de células granulares (tumor de Abrikossoff) é uma doença rara, benigna, que acomete preferencialmente o segmento cérvico facial. Geralmente é uma lesão única, nodular, que pode ulcerar e apresentar bordas infiltradas e peroláceas, com fundo limpo e halo hipercrômico. Esse trabalho descreve um caso de tumor de células granulares ulcerado e de localização pouco usual reforçando a necessidade de se incluir esse tumor no diagnóstico diferencial das lesões nódulo-ulceradas da pele.
Palavras-chave: Células de Schwann; Neoplasias; Tumor de células granulares

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