

Vulvar granular cell tumor (Abrikossoff's tumor) – Case report

Tumor de células granulosas (tumor de Abrikossoff) vulvar – Relato de caso

ABSTRACT

Introduction: The granular cell tumor was first described by Abrikossoff in 1926. It is a benign, uncommon neoplasia, most frequently found on the tongue. It occurs in the vulva in 5 to 6% of cases. Its histogenesis is not known for sure, but is probably linked to Schwann cells. It is treated surgically, with a good prognosis. The tumors may recur, and some may become malignant according to the literature. The authors describe a case of a vulvar nodule with a histopathologic diagnosis of granular cell tumor, which was successfully treated with surgery.

Keywords: vulva; vulvar diseases; vulvar neoplasms, granular cell tumor.

RESUMO

Introdução: O tumor de células granulosas foi descrito por Abrikossoff em 1926. Trata-se de neoplasia benigna, incomum, observada mais frequentemente na língua e, em percentual de cinco a 6% dos casos, na vulva. Sua histogênese é incerta, provavelmente ligada às células de Schwann. O tratamento é cirúrgico, com bom prognóstico. Podem ocorrer recidivas, e existem descrições na literatura de malignidade. Os autores relatam caso de paciente com nódulo na vulva com diagnóstico histopatológico de tumor de células granulosas, tratado cirurgicamente com sucesso.

Palavras-chave: vulva; doenças da vulva; neoplasias vulvares, tumor de células granulares.

INTRODUCTION

A granular cell tumor was first described by Abrikossoff in 1926, in a patient with a lesion on the tongue.¹ It is a neoplasia of uncertain histogenesis, probably linked to Schwann cells, with a typical histological appearance, presenting polygonal cells with the characteristic granular cytoplasm.^{2,3} The most frequent site is the tongue, but they can occur in the oral mucous membrane, gastrointestinal tract, biliary tract, musculoskeletal system, salivary glands, breasts, prostate, pituitary gland and, with lower frequency, in the vulvar 4 and clitoridian 2 region. Granular cell tumors affect more blacks, prevailing in women aged 20 to 50.^{3,4}

They usually appear as a single, asymptomatic or painful nodule, with a color that varies from brownish to red, or covered by normal skin. Cases with multiple lesions have been reported.⁴

The recommended treatment is the surgical exeresis of the lesion. The prognosis is usually good, with cases of recurrence probably correlated to the incomplete removal of the tumor. These tumors rarely become malignant.² The authors report a case of Abrikossoff tumor (granular cell tumor) with uncommon features in the vulva, which was treated surgically without

Case Report

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reincidence after 36 months.

CASE REPORT

A 48-year-old black female patient, originally from Santo André, São Paulo, Brazil was referred to the Gynaecology Service presenting a brownish, painless nodule approximately 2 cm in diameter. The nodule was movable in relation to deeper planes, as it was located in the left labium majus, close to the clitoris (Figure 1). A biopsy was carried out using a 3 mm dermatologic punch, with a histopathologic diagnosis of granular cell tumor (Abrikossoff tumor).

The excision and suture of the lesion was carried out under infiltrative local anesthesia (lidocaine chlorhydrate with norepinephrine 1:50,000). The macroscopic analysis of the excised tissue showed a whitish proliferation of poorly defined margins (Figure 2). The histopathologic examination, carried out using hematoxylin-eosin (HE) staining, revealed an epidermis with pseudoepitheliomatous hyperplasia, both in the superficial and deep dermis, and cellular proliferation with the appearance of small blocks, permeated by thin bands of dense conjunctive tissue (Figure 3). The proliferated cells present wide cytoplasm containing thin, PAS-positive granulation. The nuclei were centered and without atypias. The immunohistochemistry was positive for S-100 and enolase, both with a cytoplasmic pattern. Results were negative for CEA and HMB-45. There was no recurrence in the follow-up period of 36 months after surgery (Figure 4).

DISCUSSION

Granular cell tumors, also known as Abrikossoff tumors or granular cell myoblastomas, are uncommon and are found in several sites, most frequently on the tongue. They can occur on the vulva, mainly in the labia majora.⁵ Only 5-6% of the cases reviewed in the medical literature occurred in the vulvar region.⁶

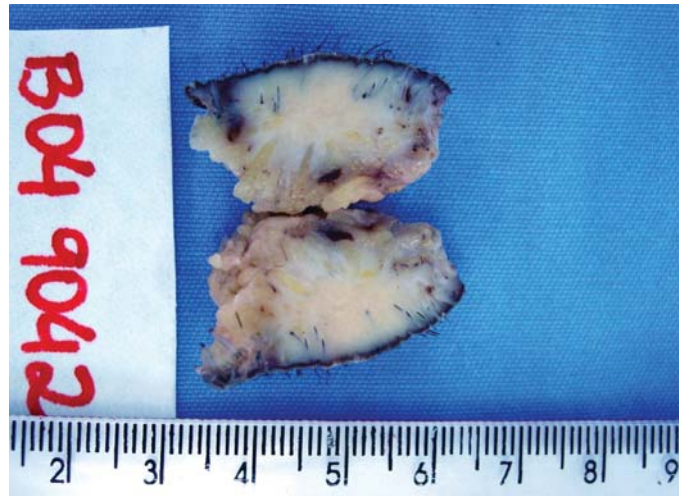


Figure 2 - Macroscopic examination showing a whitish tumor with a poorly defined margin

The histogenesis of granular cell tumors is uncertain. It is believed that the tumors are caused by alterations in the cellular metabolism of Schwann cells – a hypothesis that is reinforced by the constant presence of the S-100 protein in the immunohistochemistry, which appears with cytoplasmic pattern positivity, 3 which has also occurred in the present case.

Microscopically, polygonal cells with small nuclei and cytoplasm are observed, which contain abundant eosinophilic granular substance. Other neoplasias, such as angiosarcoma, leiomyoma, dermatofibrosarcoma protuberans and basal cell carcinoma, can also present granular cytoplasm, however they present other histological and immunohistochemical characteristics that distinguish them from Abrikossoff tumors.²

Granular cell tumors clinically present as an asymptomatic nodule, painful or slightly pruriginous, normochromic, brownish or erythematous hyperchromic, varying from 0.5-3.0 cm in



Figure 1 - Left labium majus: presence of brownish nodule approximately 2 cm in diameter

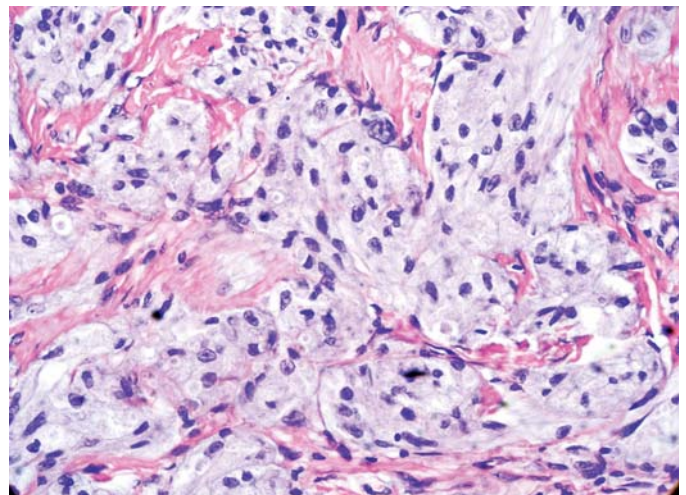


Figure 3 - Hematoxylin-eosin staining: cell proliferation, with granular cytoplasm arranged in blocks, permeated by collagen bars (200x)

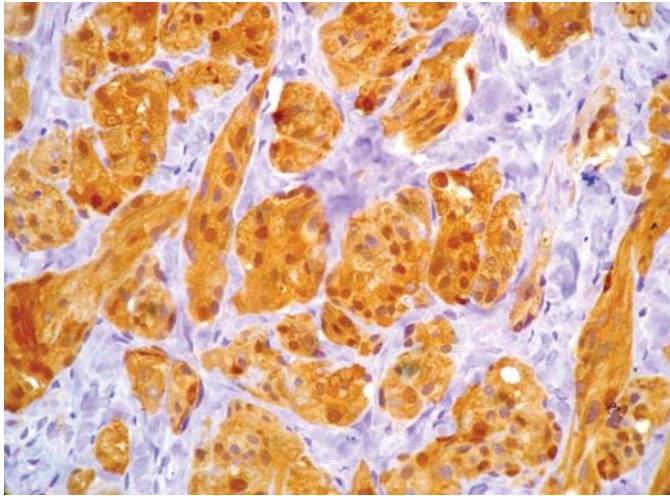


Figure 4 - S-100 immunoperoxidase. Positive reaction (brown), cytoplasmic pattern

diameter.⁷ The differential diagnosis in the vulvar case includes Bartholin gland cysts, lipoma, papilloma, hidradenoma and fibroma.⁸ Malignant transformation can occur in 1–2% of cases.⁹

Clinical diagnosis is very difficult, especially in the vulvar region, due to the rarity of this lesion and its capacity to mimic other pathologies that are more frequently found in that location; in general, it requires a histological diagnosis.

The recommended treatment is the surgical exeresis of the lesion. Local recurrence of the lesion can occur in 15% of cases if the excision is incomplete.¹⁰ In the present case, the lesion was completely removed, without recurrence after 36 months of follow-up. ●

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