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Case Report

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Vulvar Abrikossoff's Tumour: Case Report and Review of the Literature

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Abstract

Background: Abrikossoff's or granular cell tumours (GCTs) are rare neoplasms, probably neurogenic in its origins. Vulvar involvement is uncommon and most are benign.

Case report: A 63 year-old woman presented with aprogressive growing vulvar tumour, localized in the left labium majus, noticed 4 years before. No concomitant lesions were found. Biopsy revealed a benign GCT and surgical wide excision was performed. Fresh frozen section showed disease-free surgical margins. To date, there was no evidence of recurrence.

Conclusions: Despite its nonaggressive behaviour, GCT should be considered in the differential diagnosis of vulvar masses, even though it is rare. The clinicians should be aware of the possible multi centricity of the lesions and its recurrence potential, particularly if surgical margins are involved. Thus, the importance of seeking for concomitant lesions and performing fresh frozen section, to promote an effective and safe surgical approach.

Keywords

Granular cell tumour; Abrikossoff's tumour; Vulva

Introduction

Abrikossoff's or granular cell tumours (GCT) are uncommon, usually benign, soft tissue tumours of neural origin, characterized by large granular-appearing eosinophilic cells. Vulvar presentation is uncommon, being reported in 5-16% of all cases [1].

Case Report

A 63-year-old white woman sought consultation complaining of vulvar pruritus and burning, associated with a gradually growing vulvar lump, sometimes painful, with erosions and bloody discharge, with 4-years evolution. She had been medicated with clotrimazol and estriol, without success. Of her medical records, only a hysterectomy for leiomyoma, complicated with iatrogenic ligation of the ureter and requiring left nephrectomy were noteworthy.

Gynaecological examination revealed a well-circumscribed 2 cm nodule, pinkish, firm, painless and apparently not infiltrating the surrounding tissues, localized in the upper thirdof the left *labium majus* (Figure 1). No inguinal enlarged lymph nodeswere detected. Physical examination was otherwise unremarkable. A biopsy of the lesion was performed, showingthe typical characteristics of benign granular cell tumour (GCT) (Figure 2), confirmed by histochemistry (periodic acid–Schiff positive and diastase resistant) and immunocytochemistry (reactive to S100; non-reactive to HMB45 and Mib1).

A chest X-ray was performed, excluding the presence of pulmonary lesions. She underwent wide local excision of the tumour, preserving the clitoris, and fresh frozen section was performed, confirming the absence of disease in the surgical margins. Histopathologically examination confirmed the previous diagnosis. Postoperative recovery was uneventful. At 12 months follow-up, there was no evidence of recurrence and the patient was counselled and discharged.

Discussion

Granular cell tumours were first described by Weber in 1854. Abrikossoff, in 1926, proposed that it originated in smooth muscle cells, therefore naming it as granular cell myoblastoma [2]; only in the late 20th century it was established that it probably derives from neural tissue, possibly from the Schwann cells of peripheral nerves, as supported by immunohistochemistry.

GCTs are most commonly found in the head and neck (45-60%), mostly in the tongue. Other frequent locations include the thorax and upper limbs. Internal organs can also be affected, especially the respiratory and gastrointestinal tract, namely the oesophagus [3]. Breast GCT represent 5-8% of reported cases [4]. The vulvar involvement is found in 5-16% of cases [1]. It can be rarely found in other genital locations: vagina, cervix, ovaries and Fallopian tubes.

The peak age incidence is in the 4th to 6th decades of life, but can be found at any age and even be congenital (mostly epulides). GTCs are more frequent in females and in individuals of African ethnicity [5]. Rare forms of hereditary GCTs have been described [6].

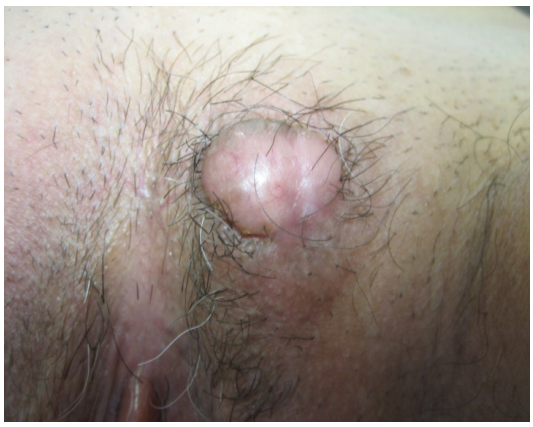

Figure 1: Tumor localized in the left *labium majus*

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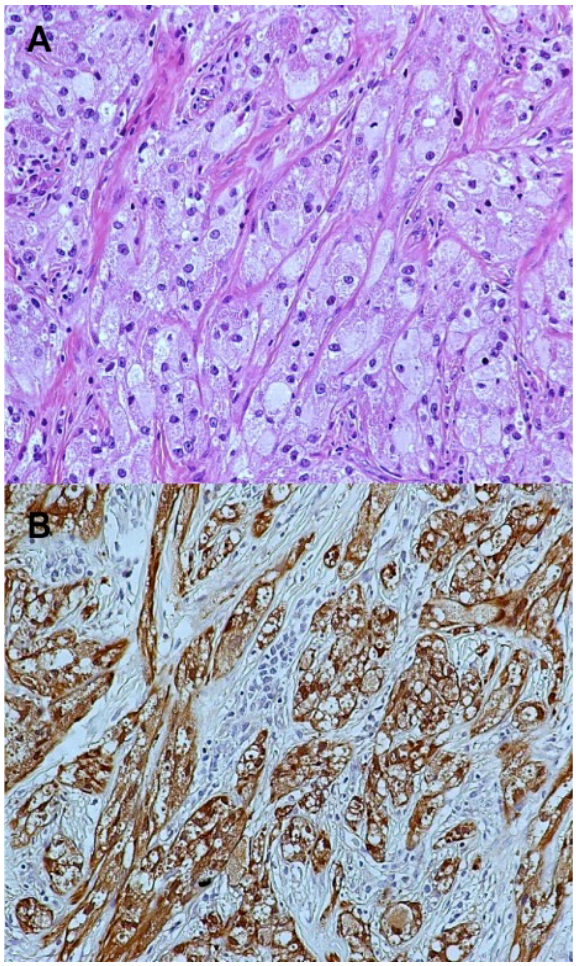

Figure 2: Histology (x200): A - Cells with granular eosinophilic cytoplasm; B - Immunorreactivity to S-100

GCTs usually present as firm nodules, but can also be found as cysts, papules or warty lesions, characteristically pale, poorly circumscribed with irregular margins, usually less than 4 cm and sometimes encapsulated. Its growth can be nodular and limited or, in the opposite, have an infiltrating behaviour, with a greater tendency to recur. The lesions can be single or multiple (3-20%) [7], appearing synchronously or metachronously over time. Multiple GTCs have been reported in LEOPARD syndrome associated with mutations in PTPN11.

Recurrence rates stands at 2-8% after a negative surgical margin and 20% if positive [8]. Most of them occur in the first two years of follow-up and may be local or distant. Only 1-2% of GCTs are malignant [1], but its diagnosis can be a challenge. Many times, it is only achieved when the tumour metastasizes or when it has an unusually locally aggressive behaviour [9].

Clinical factors that raise the suspicion of malignancy are: a tumour larger than 4 cm, fast growing, invasion of the adjacent tissues, recurrence and increased age [7].

The diagnosis is made by histology, and corroborated by immunohistochemistry. Characteristically, large pale round to polyhedral cell with abundant fine and coarsely granular eosinophilic cytoplasm. Nuclei are usually ranging from small and dark to large with vesicular chromatin. The main feature is the granular cytoplasm, due to the accumulation of lysosomes. Immuno reactivity to S-100, neuron-specific enolase, laminin and various myelin proteins, positivity to periodic acid-Schiff and resistance to diastase are typically found [1].

Wide local excision is the treatment of choice for the benign forms, which is curative in most cases. One study reported 17 cases, of which 7 had involved margins, with 2 cases progressing and requiring reintervention (14 and 8 years later) [10].The authors sustain that fresh frozen section can reduce the need for re-interventions due to recurrence, but it is still a controversial issue.

In the presence of a malignant GCT, radical surgery, including lymphadenectomy, is the primary treatment. Staging includes: hepatic ultrasound, chest X-ray, CT scan (thorax, abdomen and pelvic) and scintigraphy [7]. Adjuvant radiotherapy can play a role in some cases [11].

Conclusions

GCTs are rare soft tissue tumours, usually benign, probably arising from the Schwann cells. It is more common in females, in the 4-6th decades of life and black individuals.

Up to 5-16% are found in the vulva, presenting as a slow-growing, non-tender, firm and pale lumps, rarely larger than 4 cm.

The diagnosis is confirmed by histology, complemented by immunohistochemistry (protein S100) and histochemistry (PAS and PAS diastase). Pulmonary lesions must be excluded by X-ray. If malignancy is suspected, poor prognosis factors are present or the patient is symptomatic, further imaging study must be carried out.

In the benign forms, the treatment is wide local excision. The authors recommend fresh frozen section to ensure disease-free margins, reducing the risk of recurrence. Despite the favourable prognosis, particularly after complete excision, follow-up must be kept, especially in the first two years.

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