Subcutaneous Granular Cell Tumour of the Lumbar Region

Granular cell tumour (GCT), also known as Abrikossoff tumour, is an uncommon neoplasm, probably of neural origin derived from Schwann cells. It usually presents as a subcutaneous solitary asymptomatic nodule. It has been the subject of much debate in the literature concerning the tumour origin and the association with other malignancies. We report a case of subcutaneous GCT in the lumbar region in a 31-year-old Caucasian male. Although they are a rare entity, GCTs should be considered in the differential diagnosis of the subcutaneous soft tissue tumours. Surgical removal with wide margins is the treatment of choice as malignant changes have been reported after long-term follow-up.

KEYWORDS: Abrikossoff, benign, granular cell tumour, malignant, non-neural granular cell tumour, subcutaneous

INTRODUCTION

Although they consist a rare entity, Granular cell tumours have been the subject of much debate in the literature. Due to their usually subtle presentation, they are often misdiagnosed, with histological examination setting the correct diagnosis subsequently. Moreover, they can be found in any kind of tissue. In the case we present, the tumour was located subcutaneously in the lumbar region, and exhibited unusual immunohistochemistry.

CASE REPORT

An otherwise healthy 31-year-old Caucasian male presented with a slowly growing soft tissue mass of the right lumbar region. The patient first palpated it approximately 14 years ago. Apart from an increase in size, it remained asymptomatic ever since, painless and without any changes of the overlying skin. The family or medical history of the patient contributed nothing relevant.

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On clinical examination, the mass was a palpable lump in the subcutaneous tissue with relatively clear margins and poor mobility. Preoperative investigations, including complete blood counts (CBC), biochemical analysis, and chest X-ray were unremarkable. A computed tomography (CT) scan of the abdomen demonstrated a 5 × 3 × 3 cm subcutaneous lesion located in the right lumbar region laterally to L3, with solid, smooth, and well-defined margins [Figure 1]. The initial differential diagnosis included subcutaneous fibroma and lipoma, although malignancies, such as soft tissue sarcomas could not be ruled out.

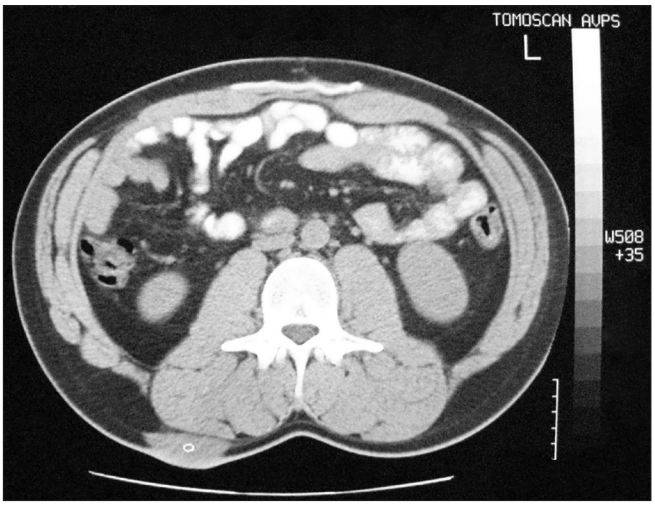

Figure 1: CT scan demonstrating a 5 × 3 × 3 cm subcutaneous lesion located in the right lumbar region laterally to L3

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The patient was scheduled for surgical removal of the subcutaneous mass. Intra-operatively, the tumour presented with adhesions to the lumbar fascia, which was dissected en bloc with the tumour. The specimen was excised with macroscopically clear margins of normal tissue. The wound was closed primarily and the patient was discharged in the same afternoon after an uneventful post-operative course.

Histological examination of the specimen demonstrated a macroscopically pink-yellow lesion of elastic texture, with dimensions of 4.5 × 3 × 2 cm. Microscopically on hematoxylin–eosin stain, the lesion included neoplastic cells, containing plenty to abundant granular eosinophilic cytoplasm and small dense nuclei in the cutaneous and subcutaneous fatty tissue. The cells formed nests or strands circumscribed by fibrous septae and strands of collagen [Figure 2]. The immunohistochemic assay of the tumour was negative for neurone-specific enolase (NSE), weakly positive for CD68, and moderately positive for S100 and Vimentin [Figure 3]. The microscopic and immunohistochemical features were suggestive of granular cell tumour. The follow-up of the patient 16 months after surgery revealed no signs of local recurrence or metastases.

DISCUSSION

Granular cell tumour (GCT) or Abrikossoff's tumour is a rare neoplasia considered to be of neural origin derived from Schwann cells.[1] The tumour can be found in almost every kind of tissue. It may be congenital or non-infantile occurring between 20 and 60 years of age with a peak around the age of 50 years. There is a female preponderance (8/1) regarding congenital and (3/1) for the non-infantile GCTs, and it is most common in blacks.[2] In 25% of cases the tumour is multicentric, and reports of familial cases with multifocal tumours raise the suspicion of genetic compound.[3]

The disease in 30%–45% of cases affects the skin followed by the area of head and neck where the most frequent location is intraoral in the tongue and the soft and hard palate.[4] Other locations affected are the breast, the gastrointestinal tract—mainly the lower third of the oesophagus—the respiratory tract, the thyroid gland, the urinary bladder, the central nervous system, and the female genitalia. Regarding the latter, the vulva is the predominant site affected in 5%–16% of these cases, but the disease can also be found in the cervix, the uterus and the ovaries.[5] While the GCT typically affects the skin and subcutis, location in the lumbar region as in our case, has not been reported except for intradural[6] or multiple GCTs.[7]

Cutaneous and subcutaneous disease is usually detected as a solitary, small, non-tender, slowly growing mass, sometimes with pruritus of the overlying skin and less frequently with pain. In some cases, pseudo-epitheliomatous hyperplasia of the overlying skin may be obvious, which is attributed to the chronic irritant effect of the microscopic infiltration of the tumour. This presentation can be mistaken for well-differentiated squamous cell carcinoma (SCC), although association of benign GCT with other malignancies of the skin exists. The clinical differential diagnosis also includes dermatofibroma, hidradenoma, dermoid cyst, prurigo nodularis, fibroadenoma, and fibrosarcoma.[2]

Microscopically, the cells are polygonal or oval, and less frequently spindle-shaped, with abundant, eosinophilic, granular, coarse cytoplasm. The nuclei are small and dense and bands of fibrous tissue separate the cells. In cases of malignancy, there is a peripheral infiltrative

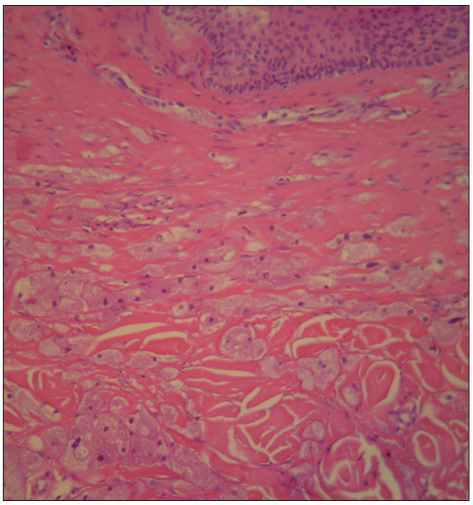

Figure 2: Tumour composed of large polyhedral cells with an abundant granular eosinophilic cytoplasm and centrally located nuclei. (H and E, ×100)

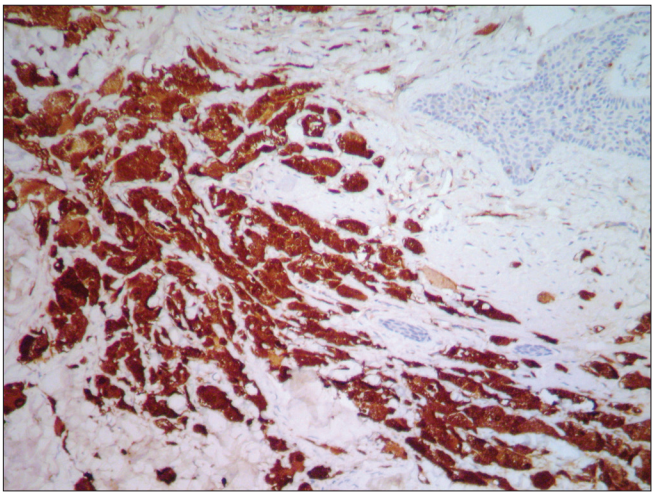

Figure 3: The tumour cells stain positively for S-100 protein. (×200)

pattern with satellite nodules in 10%–15% of the cases. In addition, the nuclei are large and vesicular containing a single or multiple nucleoli and demonstrate nuclear pleomorphism.

The granules are usually positive for Periodic acid–Schiff (PAS) and Sudan Black but negative for diastase. Although conventional GCTs usually show consistent NSE and S-100 positivity, atypical and non-consistent immunohistochemistry has been reported,[8] while there are non-neural GCTs that exhibit immunohistochemic diversity, probably due to a more mesenchymatic rather than a neural or Schwannian nature.[9] In our case, the tumour was NSE negative, with S-100 and Vimentin positivity.

The prognosis of the tumour mainly depends on whether it represents a benign or a malignant case. It is generally accepted that benign lesions can be safely managed with local excision on clear margins. Although most of the cases follow a benign course, 1%–2% of GCTs exhibit malignant behaviour and behave like high-grade sarcomas with a high rate of metastases and short survival.[10] In this case, it is supported that lymphadenectomy can improve the outcome. Moreover, despite that chemotherapy and radiotherapy are not shown to improve statistically significantly the outcome, their application in cases of metastatic GCT is accepted.[10]

Due to their subtle appearance and symptomatology as a typical subcutaneous lump, GCTs are often extemporarily misdiagnosed with histological examination setting the correct diagnosis subsequently. Nevertheless, although rare, the association of GCTs with malignancy renders a differential diagnosis including all possible benign tumours and malignancies with respect to the anatomic site of presentation being imperative.

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