International Journal of Advances in Medicine
Khan S et al. Int J Adv Med. 2017 Feb;4(1):293-295
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: http://dx.doi.org/10.18203/2349-3933.ijam20170130

Case Report

Infrascapular granular cell tumor: an unusual entity

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Received: 07 December 2016
Accepted: 01 January 2017

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ABSTRACT

Granular cell tumor (GCT) is an uncommon soft tissue tumor of schwannian origin frequent among women and blacks between the second and sixth decades of life. The common location of GCT is the oral cavity, but it can also occur at other sites. Cutaneous lesions constitute about 30% of cases and are characterized by a gradually developing nodular lesion. Due to their subtle clinical appearance and symptomatology, GCTs are often misdiagnosed. We report a case of subcutaneous GCT in the infrascapular region in a 40 year old female which mimicked granular cell dermatofibroma on histopathology. Although a rare entity, Granular cell tumor should be considered in the differential diagnosis of the subcutaneous soft tissue tumours and require histopathological examination along with immunohistochemistry to confirm the diagnosis and differentiate them from other benign and malignant tumors showing granular cell change.

Keywords: Granular cell tumor, Histopathology, Subcutaneous

INTRODUCTION

Granular cell tumors, first described by Abrikossoff in 1926 as myoblastomas, are tumors of Schwannian cell origin which represent 0.5% of all soft tissue tumors.[1] The tumor occurs frequently among women and blacks, between the second and sixth decades of life.[2] GCT occurs in almost any part of the body.

The common sites are the tongue, skin, and subcutaneous tissue. Its malignancy potential is 1-3%.[3] Due to their rarity and usually a subtle clinical presentation, they are often misdiagnosed, with histological examination along with immunohistochemistry helping in the correct diagnosis. Here we report a case of granular cell tumor in a 40 year old female which was diagnosed as benign mesenchymal lesion on cytology and mimicked granular cell dermatofibroma on histopathology.

CASE REPORT

A 40 year old female presented in the surgical OPD with a swelling in left infrascapular region since 3 years which was gradually increasing in size. Apart from an increase in size, it remained asymptomatic. There was mild pain since few days for which she took medical advice in our hospital. There was nothing significant in personal or family history.

On clinical examination, a palpable swelling was present in the left infrascapular region measuring 4x3 cm, firm to hard, non-tender with limited mobility. Overlying skin was inflamed. No other similar swelling was present in the body. Routine investigations, including complete blood counts (CBC) and biochemical analysis were unremarkable. Patient was advised fine needle aspiration cytology (FNAC) which revealed richly cellular smears consisting of singly lying cells and small cohesive aggregates. Fair number of these cells showed abundant granular eosinophilic cytoplasm (Figure 1A). Few small aggregates showed plump spindle shaped nuclei with bland chromatin and regular margins. Collagen was also seen interspersed in between cells (Figure 1B). Based on these cytological findings a diagnosis of benign mesenchymal lesion was given. The patient was then advised excision of the subcutaneous mass.

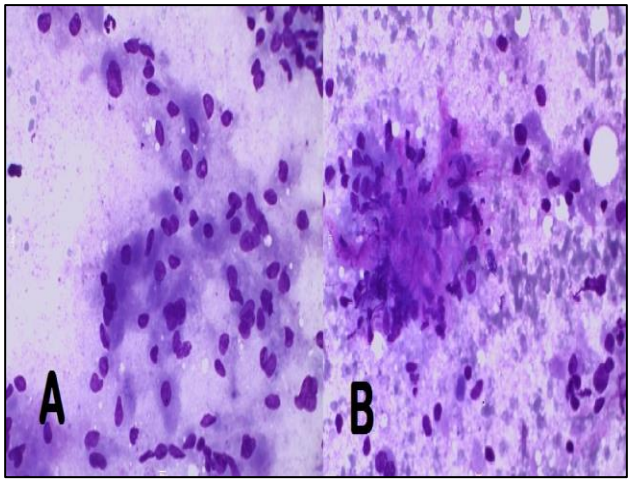

Figure 1: a) FNAC smears showing few clusters of cells having abundant granular eosinophilic cytoplasm (MGG stain, 40x). b) Few small aggregates showing plump spindle shaped cells with collagen interspersed in the background (MGG stain, 40x).

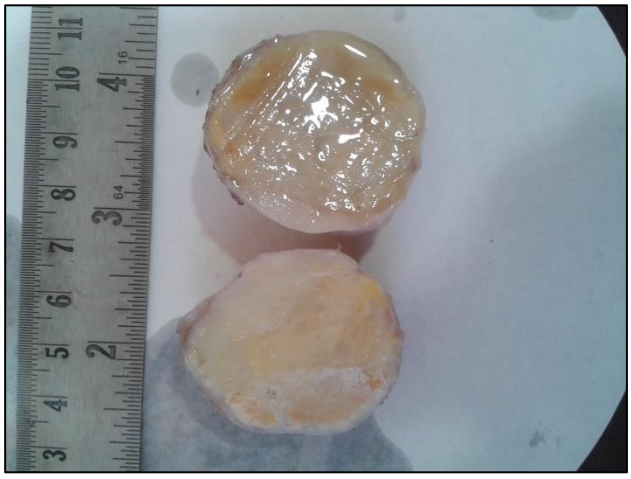

Figure 2: Gross examination of the specimen showed globular tissue, greyish brown in color. Cut surface was smooth and homogenous.

Gross examination of the specimen showed globular tissue, greyish brown in color measuring 3× 3 × 2 cm. Cut surface was smooth and homogenous (Figure 2). Microscopically, nests and strands of tumor cells were seen separated by fibrous septa. These cells showed vesicular nuclei with prominent nucleoli in some cells and abundant granular eosinophilic cytoplasm (Figure 3A). However, few spindle cells, collagen and some interspersed lymphohistiocytic infiltrate was also seen at the periphery of the lesion which suggested fibrohistiocytic origin (Figure 3B). Thus possibilities of granular cell tumor as well as granular cell dermatofibroma was suggested on histopathology. Immunohistochemistry was advised which showed diffuse positivity for both S 100 (Figure 4A) and CD 57 (NK1C3) (Figure 4B) thus clinching the diagnosis of Granular cell tumour. Patient is on regular follow up and did not show any signs of recurrence or metastasis.

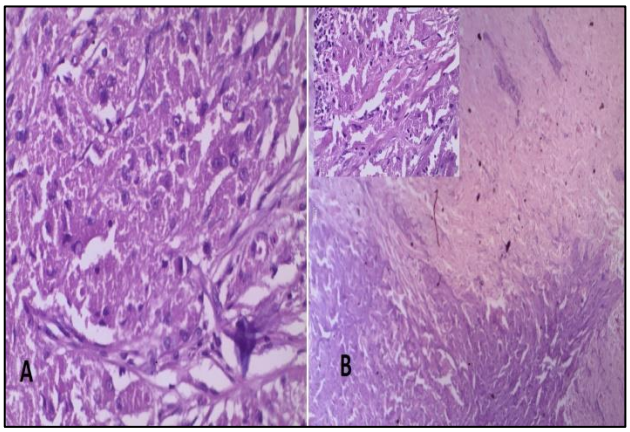

Figure 3: A) Nests of tumor cells having vesicular nuclei with prominent nucleoli in some cells and abundant granular eosinophilic cytoplasm (H&E stain, 40X); B) Abundant collagen at the periphery of the lesion. Inset shows aggregate of lymphocytes.

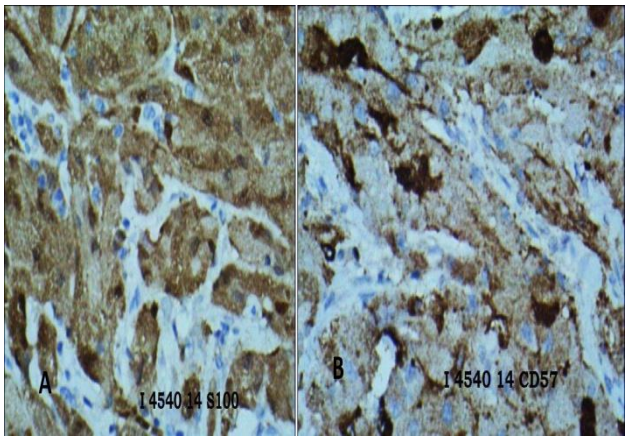

Figure 4: A) Immunohistochemistry shows tumor cells showing diffuse positivity for S 100; B) IHC shows positivity for CD57 (NK1C3).

DISCUSSION

Granular cell tumour is a rare tumor considered to be of neural origin derived from Schwann cells. Most of the published experience is based on sparsely presented case reports and few small series. It usually occurs between 20 and 60 years of age with a peak around the age of 50 years. There is a female preponderance and is most commonly seen in blacks. In our case also the patient was a forty year old female. In 25% of cases the tumour is multicentric, and reports of familial cases with multifocal tumours are also present.[4] The tumour can arise anywhere in the body and in almost every kind of tissue. The common location of GCT is the oral cavity, where the most frequent location is tongue followed by soft and hard palate.[3] Other sites affected are the breast, the gastrointestinal tract, respiratory tract, the thyroid gland,

the urinary bladder, the central nervous system, and the female genitalia.[5]

Cutaneous lesions constitute about 30% of the cases of Granular cell tumor. They often present as asymptomatic, slow-growing, solitary lesions but may be multifocal. The clinical presentation of cutaneous GCT is mostly nonspecific and hardly suspected. They behave in a benign fashion, but have a tendency to recur. Malignant granular cell tumors are exceedingly rare and represent 1-3% of all granular cell tumors. Fanburg-Smith et al described objective morphologic criteria to differentiate benign GCT from malignant GCT.[6] Tumors which met three out of six histopathologic criteria: necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity, high nuclear to cytoplasmic ratio, and pleomorphism were characterized as malignant. The present case was a benign GCT and did not exhibit any of the above features. The clinical differential diagnosis of GCT arising in a subcutaneous location includes dermatofibroma, hidradenoma, dermoid cyst, fibroadenoma and fibrosarcoma.[2] The final diagnosis of GCT is based on histological findings and confirmed by immunohistochemistry which usually shows positive staining for S-100 and NSE.[7]

The characteristic histological feature of GCT is the coarse eosinophilic cytoplasmic granules. Cellular granularity is the result of cytoplasmic accumulation of lysosomal structures and can be observed not only in conventional granular cell tumor but in a variety of benign and malignant cutaneous neoplasms.[8] Granular cell change has been reported in dermatofibroma, ameloblastoma, leiomyoma, leiomyosarcoma, angiosarcoma, MFH and melanoma. In our case histological findings resembled granular cell dermatofibroma. It is important to recognize dermatofibroma with granular cells because it may be confused with other soft tissue tumors containing similar granular cells that entail different significance or prognosis.[9] Cheng et al in 2001 described the histopathological findings in a case of dermatofibroma like granular cell tumor.[10] In such cases IHC is helpful as in contrast to classic Schwannian/neurogenic granular cell tumour, granular cell dermatofibroma is S100 negative and usually positive for NK1C3(CD57). In our case immunohistochemistry showed diffuse positivity for both S100 and CD57, thus favouring diagnosis of Granular cell tumor over granular cell dermatofibroma.

Although a rare entity, granular cell tumor should be considered in the differential diagnosis of the subcutaneous soft tissue tumours. Due to their subtle appearance and symptomatology, GCTs are often misdiagnosed and require histological examination for confirming the diagnosis. Although rare, the association of GCTs with malignancy further highlights the importance of detailed histopathological examination.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Khan S, Jetley S. Infrascapular granular cell tumor: an unusual entity. Int J Adv Med 2017;4:293-5.