Granular Cell Tumor: An Uncommon Benign Neoplasm

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Abstract
Granular cell tumor is a distinctly rare neoplasm of neural sheath origin. It mainly presents as a solitary asymptomatic swelling in the oral cavity, skin, and rarely internal organs in the middle age. Histopathology is characteristic, showing polyhedral cells containing numerous fine eosinophilic granules with indistinct cell margins. We present a case of granular cell tumor on the back of a 48-year-old woman which was painful, mimicking an adnexal tumor.

Key Words: Back, granular cell tumor, painful

Introduction
Granular cell tumor (GCT) is an uncommon benign tumor, affecting the skin and internal organs. The condition was first described by Abrikossoff, and the name ‘granular cell myoblastoma’ was suggested by Horn and Stout.[1] The name was given on the erroneous notion regarding its origin from muscle. These neoplasms are now considered to be of neural origin. The tumor is usually asymptomatic although some patients may experience mild pruritus or pain. A case of a large solitary painful GCT is reported here for its rarity.

Case Report
A 48-year-old housewife presented to us with a painful solid mass over her upper back present for the preceding 2 years. Starting as a pea-size swelling, it increased gradually in size and became more painful. There was no history of prior trauma or any discharge from the lesion [Figure 1].

On examination, there was a well-defined, very tender, yellowish, hard lesion of about 3 cm diameter, having a smooth irregular surface located over the midline in the upper back. It was not fixed to underlying tissue. There was no regional lymphadenopathy and no other mucocutaneous findings. Systemic review was non-contributory.

Fine-needle aspiration cytology (FNAC) showed large round to oval cells with vesicular nuclei. Few binucleated cells were embedded in abundant proteinaceous material. Histology showed sheets of large polygonal cells infiltrating the dermis. The cells were characterized by abundant, coarsely granular and amphophilic cytoplasm; with eccentrically located small vesiculated nuclei and prominent nucleoli. There was neither any mitotic figure nor spindling of the lesional cells [Figures 2 and 3].

Routine laboratory tests, blood biochemistry panel and imaging studies were within normal limits. Immunohistochemistry was not done due to local unavailability and financial constraints. Based on the clinical and histological findings, a diagnosis of GCT was made.

Figure 1: Well-defined, yellowish hard nodular lesion with smooth irregular surface over midline in the upper back
**Discussion**

GCT, also known as Abrikosoff’s tumor, is an uncommon benign neoplasm. It was long debated whether the tumor originates from muscle, fibroblast, histiocyte, neural crest or nerve sheath. However, it is now accepted to be of neural sheath origin, indicated by S‑100 protein, peripheral nerve myelin protein, such as P2 and P0 protein positivity.[2,3]

Although common during the third and fourth decades of life, GCT can occur at any age. Two thirds of the cases had been reported in women.[2,4] Around 40% of cases are found in the tongue while skin and subcutaneous tissue share one third of the cases. Other sites such as esophagus, stomach, larynx, bronchus, uvea, muscle and pituitary stalk may be involved. GCT had been reported to occur in vaccination scar.[5] In 10% cases multiple tumors may be found at a time.[6] GCT usually presents with a well‑circumscribed, solitary, raised, firm nodule of 0.5 to 3.0 cm diameter with a smooth, rough or verrucous surface. It usually remains asymptomatic, but sometimes may be associated with mild pruritus or tenderness. Our case was painful and very tender, an uncommon finding.

Histopathological examination is necessary for a conclusive diagnosis. FNAC may sometimes be helpful in this regard. The characteristic cytologic findings are large, pale oval, round, bipolar or polygonal cells with abundant fine or coarsely granular, eosinophilic cytoplasm, inconspicuous cell border, eccentrically located nuclei, fine nuclear chromatin and indistinct nucleoli.[7] Histopathology shows the same cellular architecture, pseudoepitheliomatous hyperplasia and cords of cells infiltrating into the dermis. Electron microscopy indicates that the characteristic granularity is owing to the accumulation of lysosomes.[8] On immunohistochemistry, the tumor cells are positively stained with vimentin, neuron‑specific enolase, S‑100, myelin protein, p75 nerve growth factor, calretinin, NKI/C3 and PGP9.5.[9]

Malignant GCT is found in only 1 to 2% of cases, indicated by large size, rapid growth, lymph node involvement, necrosis and cytologic features of malignancy. As mitoses are normally found in GCT, they are not considered to be the marker for malignant transformation.[10] An important feature of malignancy in a granular cell tumor is spindling of lesional cells.

Complete excision is the best mode of intervention. Rarely the tumor can recur, usually following inadequate removal.

Our case presented with a well‑defined very painful nodular lesion with a smooth, but bosselated surface. We considered different painful dermal adnexal tumors as differential diagnoses. However, typical FNAC and histopathological findings indicated a diagnosis of GCT. We referred the patient to plastic and reconstructive surgeon for complete excision and follow up.

**What is new?**

Our case presented with an extremely painful and very tender swelling, an uncommon finding.

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