

## Case Report:

### Granular cell tumor: a report of two cases

<sup>1</sup>Dr. R. Sujitha , <sup>2</sup>Dr. S. Sowmya

<sup>1</sup>Post-graduate Student and Junior Resident, Department of Pathology, S.M.V.M.C.H., Puducherry-605107, India

<sup>2</sup>Professor & Head of Department , Department of Pathology , S.M.V.M.C.H., Puducherry-605107, India

Corresponding author: Dr. R. Sujitha

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#### Abstract

Granular cell tumor (GCT) is a rare neoplasm arising from skin, soft tissue and internal organs. It is hypothesized to be originating from the Schwann cells. It is usually benign but rarely malignant. The tumor is diagnosed by microscopic examination and can simulate any adnexal or soft tissue neoplasm. We report our findings in two cases, presenting with GCT of soft tissue. Nervous tissue elements were seen in both. The diagnosis was based on the characteristic tumor cells on Hematoxylin and Eosin stain (H & E) and confirmation of the neural origin required S-100 and NSE immuno-histochemistry.

**Key words:** Granular cell tumor, S-100, NSE

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#### Introduction

Granular cell tumor (GCT) was first reported by Abrikossoff in 1926.<sup>1</sup> It is a rare neoplasm of nervous tissue origin, arising from skin or soft tissue. They can arise at virtually any body part, but head and neck, chest wall and arms are the three most commonly affected sites. The other sites less frequently affected are tongue, upper respiratory tract, gastrointestinal tract, breast, thyroid, eye, reproductive organs and abdominal wall.<sup>2</sup> We report our findings in two cases, presenting with GCT of soft tissue, diagnosed based on microscopic appearance.

#### Case Report

##### Case 1

50 year old female, presented with swelling in the back of 6 months duration. On examination it was firm, non-tender and measured 4 x 2 x 1 cms.

##### Case 2

38 year old female, presented with swelling in the neck of 1 year duration. On examination it was

firm, non-tender and measured 1.5 x 0.8 x 0.2 cms.

Excision biopsy was done in both the patients and the tissue was sent for histopathological examination. The histological sections were stained with hematoxylin and eosin (H & E) and Periodic Acid Schiff (PAS) stains, and immunohistochemistry (IHC) was performed with S-100 and neuron specific enolase (NSE).

Microscopically the tumor was composed of nests and lobules of large round to polygonal cells (**Figure 1**) with abundant eosinophilic, coarse granular cytoplasm (**Figure 2**) and vesicular centrally placed nuclei. The cytoplasmic granules were strongly PAS positive. The cell nests were interspersed by fibrous tissue with thin-walled blood vessels. Case 1 had overlying skin which was free of the tumor, however deep resected margin and nerve sheath showed tumor cells. In Case 2, the margins were free. IHC for S-100 (**Figure 3**) and NSE (**Figure 4**) showed strong intracytoplasmic

granular positivity. These findings observed in both the cases were consistent with GCT.

#### **Discussion**

GCT was initially postulated by Abrikossoff to be originating from striated muscle cells and termed as 'Myoblastoma'.<sup>1</sup> However, in subsequent studies the tumor was found to exhibit ultrastructural similarities to Schwann cells and expressed positivity for neural S-100 protein. Hence, presently the most accepted theory of origin of GCT is from Schwann cells of the nervous system.<sup>3</sup> GCT, more frequently occurs between age group of 40-60 years. The ratio of incidence in males to females is 2:3. It is usually benign but rarely, in 1 to 2 % of cases can turn malignant and cause local invasion or distant metastasis.<sup>4</sup>

The usual clinical presentation of GCT is a solitary asymptomatic swelling. Rarely multiple GCT have been described. It presents as nodular, pinkish-yellow well defined lesion, rarely exceeding 3 cm in diameter. It usually involves sub-cutaneous or sub-mucosal tissue. The skin surrounding the tumor may be normal or it may be thickened or ulcerated. Rarely a tuft of hair is seen arising from the tumor site.<sup>4</sup>

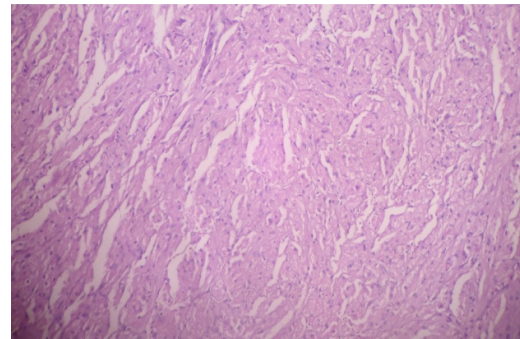
Histopathological examination of GCT classically reveals large cells with eosinophilic granular cytoplasm and PAS positivity, as was demonstrated in our cases.<sup>5</sup> This condition should be differentiated from Hibernoma, which can have granular to vacuolated cytoplasm. Positive IHC staining for S-100 and NSE, helps to differentiate GCT from hibernoma.

Malignant GCT may demonstrate increased mitotic activity, nuclear atypia and necrosis of tissue. Differential diagnosis of malignant GCT includes alveolar soft tissue sarcoma, which shows cytoplasmic granular PAS positivity, but negative for S-100 and NSE immunostaining.

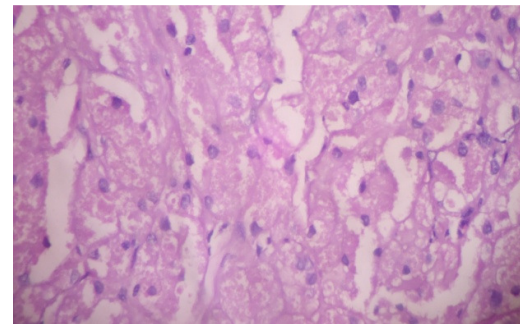
Surgical excision is the treatment of choice for GCT. Radiotherapy may be required in case of malignant invasion of local tissue.

#### **Conclusion**

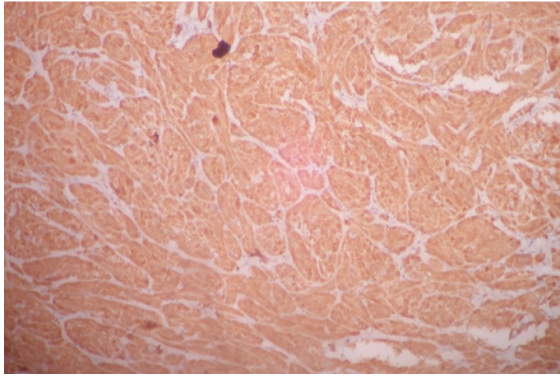
GCT is a very rare and usually benign neoplasm of nervous tissue origin. It usually arises from skin and subcutaneous tissue. Histopathological examination is necessary for diagnosing GCT and IHC showing positivity for S-100 and NSE is confirmatory.



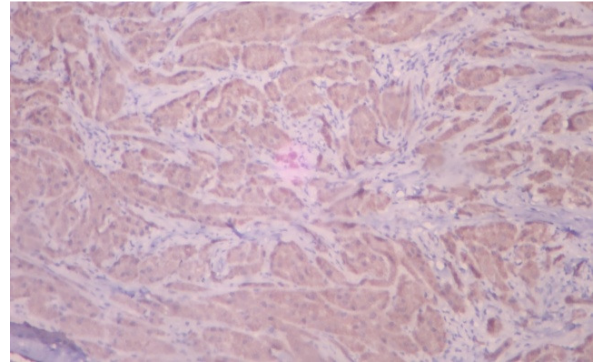
**Figure 1- Nests of cells with granular cytoplasm (H&E - 20x)**



**Figure 2- Intracytoplasmic eosinophilic granules (H&E - 40x)**



**Figure 3- Intense intracytoplasmic granular positivity (S-100 - 20x)**



**Figure 4- Intense intracytoplasmic granular positivity (NSE - 20x)**

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