



## Malignant Granular Cell Tumor of Chest Wall: Case Report and Literature Review

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### Abstract

Granular cell tumor (GCT) is a rare tumor which arise from Schwann cells in the skin, soft tissue and internal organs. Granular cell tumor are usually benign although malignant and multifocal forms are known to occur. Malignant GCT represent less than 2% of all granular cell tumors. These tumors can be found in any site in the body including tongue, skin, subcutaneous tissue, breast, gastrointestinal and urogenital systems. However, these tumors are rarely described in chest wall. Here we present a case report of 50-year-old lady with malignant granular cell tumor.

**Keywords:** NIL

### INTRODUCTION

A 50-year-old lady presented to our hospital with 4 month history of swelling and pain in right side of chest wall. She had no significant history of previous illness. Physical examination revealed a 3×4cm solid lesion on right chest wall near scapula. Mass was movable and slightly tender. Overlying skin was normal. Ultrasonography showed a 2.8×3.6×5.8cm solid lesion in inter-muscular plane in right chest wall near scapula. Fine needle aspiration showed atypical cells with probable diagnosis of cartilaginous tumor /adnexal tumor.

Excisional biopsy showed tumor with epithelial morphology and abundant granular cytoplasm favouring diagnosis of granular cell tumor (Fig-1). IHC custom panel and histologic features were suggestive of granular cell tumor with immune-reactivity for S-100 (fig-2). Patient was re-assessed in the department of surgery and radiotherapy at PGIMER and was kept on follow up due to benign nature of disease.

Patient presented with complaint of swelling in right axilla in the department of surgery at IGMC Shimla. Computed tomography showed enhancing lymph nodes in right axilla of size 22×18mm and heterogeneous mass in right lateral chest wall at inferior scapular angle with loss of fat planes with adjacent muscles (fig- 3). She underwent excision of axillary and scapular mass. Histopathology examination revealed malignant granular cell tumor of chest wall with metastatic tumor deposits in axillary lymph nodes (fig-4).

After surgery patient was reviewed in department of radiotherapy. PET- scan showed residual lesion involving right serratus anterior muscle below the inferior angle of scapula with bilateral lung metastasis. Computed tomography also confirmed lung metastasis.

Patient was started on combination chemotherapy based on paclitaxel (175mg/m<sup>2</sup>) and carboplatin AUC 6. Drugs were delivered on same day and repeated after every 21 days. Total six were delivered at our

institute. She also received single course radiotherapy @ 600 cGy. Response assessment done after 6 cycles of chemotherapy showed decrease in size of primary lesion along with reduction of size of lung metastasis.

#### DISCUSSION:

Granular cell tumor (GCT) is usually a benign soft tissue tumor, but malignant course is encountered in 2% of cases. It may arise in any part of body but is commonly affects skin or sub-cutaneous tissue of chest, upper extremities, tongue, breast and female genital regions 1.

Granular cell tumors have uncertainty about its origin. According to Abrikossoff GCT originate from degenerating striated muscle cells but later changed his view in favour of origin of GCT from embryonic muscle cells 2. He termed this lesion as myoblastic myoma. Recently Aparicio and Lumsden<sup>3</sup> and Sobol *et al* 4 proposed that GCT originated from undifferentiated mesenchymal cells. Due to this uncertainty in the origin of these tumors' terminology of GCT instead of myoblastoma is suggested for these tumors.

Differentiation between benign and malignant GCT is usually difficult, sometimes presence of metastasis establishes the malignant tumor. Histologic criteria that are considered to be important to establish the diagnosis include spindling of tumor cells, presence of vesicular nuclei with large nucleoli, increased mitotic rate (>2 mitosis / high power magnetic field at 200× magnification), a high nuclear to cytoplasmic ratio, pleomorphism and necrosis 5. Histological diagnosis of GCT is made when three or more criteria are fulfilled. Histological diagnosis is also supported by PAS and IHC staining. PAS positive cytoplasmic granularity is characteristic feature of granular cell tumor. GCT are immunoreactive for S-100, NSE, NK1-C3, viamentin, CD57, CD68 6. Clinical characteristics like rapid growth, multicentricity, size larger than 5cm are also predictable of malignant growth.

According to Sercan and Aksoy *et al*, metastatic granular cell tumors more commonly seen in women with median age at diagnosis being 43 years. Patients were usually treated surgically at time of diagnosis. Most common sites of metastasis are lung liver and bones. Lymph nodes were other commonly affected site. Local recurrences are common in metastatic

GCT. Median overall survival from diagnosis was 84 months and after detection of metastasis is 44 months 7.

Management of GCT is not clearly defined due to rarity of the malignant nature of disease and lack of adequate follow up. But wide local excision with lymph node excision forms the mainstay of treatment 8. The role of adjuvant radiotherapy and chemotherapy is still uncertain, most of the reports describes poor response to these therapeutic modalities. Some observations have reported radiotherapy efficacy in case of local recurrence or inoperable metastasis. GCT are thought to be an aggressive chemo refractory disease there is only one reported case of good response of malignant GCT to chemotherapy (gemcitabine and paclitaxel) were agents used 9. Recently Sachi Morita, *et al* reported a case of metastatic GCT which showed response to pazopanib monotherapy<sup>10</sup>.

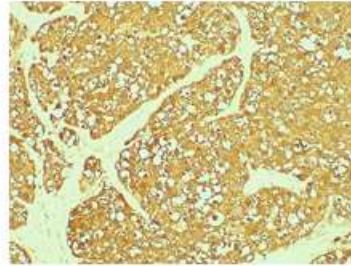
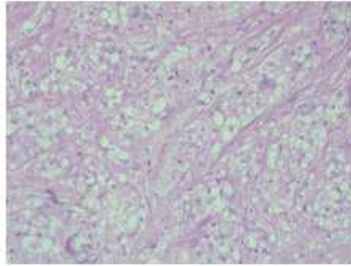
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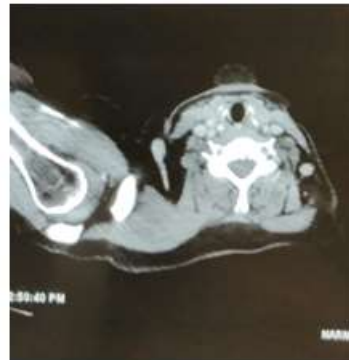
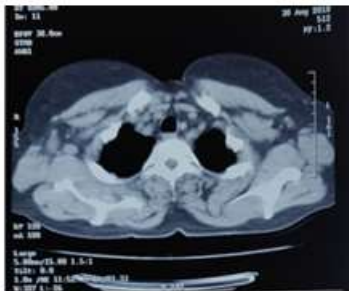
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**Fig-1 HPE image of granular cell tumor (cells with abundant granular cytoplasm)**

**Fig-2 IHC image – immunoreactivity for S-100 stain**



**Fig 3 CECT image showing heterogeneous mass in right lateral chest wall**

**Fig-4 CECT image showing axillary lymph node**