Case Report

# Granular cell tumor: A rare tumor at rare location

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## ABSTRACT

Granular cell tumor (GCT) is a rare tumor affecting the middle-aged people with an incidence of 0.017–0.029%. In 1926, Abrikossoff was the first to describe this tumor as myoblastoma as it was arising from muscle in the tongue. Common sites are the tongue, skin, and subcutaneous tissue. Here in, a 29-year-old female presented with a swelling on the ring finger of the right hand. Excision biopsy was performed. Histological features were suggestive of GCT. This case is of particular interest due to the atypical location and clinical presentation.

**Key words:** Granular cell tumor, myoblastoma, ring finger

## INTRODUCTION

Granular cell tumors (GCTs) are rare mesenchymal soft tissue tumors believed to be of neural origin presenting as asymptomatic, slow-growing, benign, and solitary lesions.[1] A GCT generally follows a benign clinical course; however, 1–2% of these cases are malignant and can metastasize, particularly when they arise in deep to fascia or are over 4 cm in diameter.[2,3] The GCT of the hand is extremely rare with only 17 cases being reported in the literature till date.[4,5] The GCT mimics dermal adnexal tumor in subcutaneous tissue, soft tissue tumor, and inflammatory lesions; therefore, preoperative diagnosis is essential.

## CASE REPORT

A 29-year-old female presented with a slow-growing swelling for the last 2 years on her right ring finger [Figure 1]. She complained of dull aching pain over the swelling for 1 year. She had no history of undue trauma. Physical examination showed a very firm, slightly tender, oblong mass located on the ventral aspect of the right ring finger at proximal interphalangeal joint level. X-ray showed soft tissue enhancement without any bony changes. No other masses were found. Fine-needle aspiration cytology was performed, yielded hemorrhagic aspirate. Excisional biopsy was performed.

Grossly, the lesion was well defined, firm, yellowish-white color, and approximately 2 cm × 2 cm in size located in the dermis and subcutaneous area. Histopathological examination revealed the sheets of tumor cells having abundant granular eosinophilic cytoplasm, round to oval centrally or eccentrically located nuclei with mild variation in size, and occasional small nucleoli [Figures 2 and 3]. Few larger, brightly, eosinophilic ovoid bodies surrounded by a clear halo were also seen. On immunohistochemistry, the tumor cells show strong cytoplasmic positivity for S-100 protein and also positive for neuron-specific enolase (NSE) and vimentin [Figures 3 and 4]. The cells were negative for CD68, cytokeratin, desmin, alpha-smooth muscle actin, and HMB-45. The hand incision healed uneventfully, and the patient is well after 15 months without recurrence.

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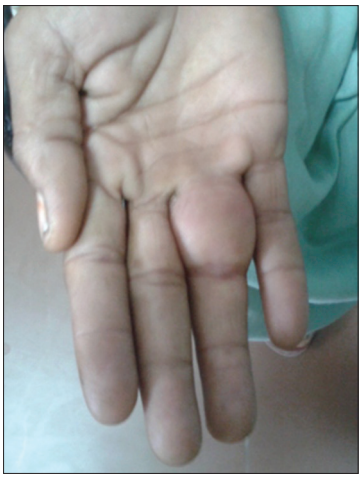

**Figure 1:** Photograph showing swelling on the ring finger of right hand

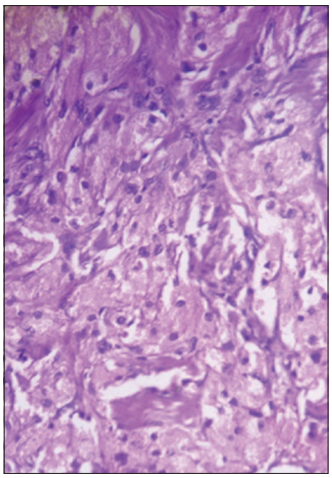

**Figure 2:** Photomicrograph showing sheets of tumor cells having abundant granular eosinophilic cytoplasm, round to oval centrally or eccentrically located nuclei (H and E, ×400)

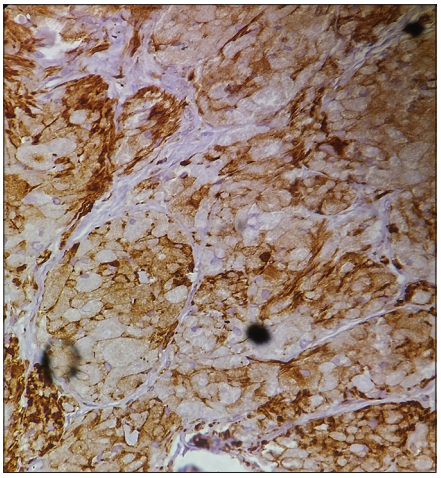

**Figure 3:** Photomicrograph showing tumor cells positive for S-100 (IHC, ×200)

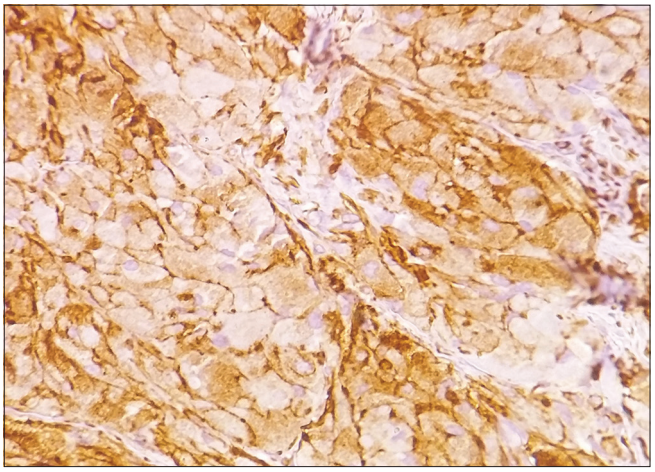

**Figure 4:** Photomicrograph showing tumor cells positive for vimentin (IHC, ×200)

## DISCUSSION

The GCT of hand is very rare with an incidence of <0.1%.[6] It commonly occurs in the third to fifth decades of life with two-third of cases reported in women and in the black population. The common sites are not only the tongue, skin, and subcutaneous tissue but also have been reported in soft tissue, nerve, breast, scalp, abdominal wall, head and neck, back, extremities, lymphnode, mediastinum, soft palate, orbit, salivary glands, respiratory tract, vulva, gastrointestinal tract, and brain.[6]

It commonly presents as an asymptomatic solitary, painless, slow growing nodule of size usually <3.0 cm and may undergo partial regression.[7] Multiple GCTs have been reported in <10% of cases, in association with neurofibromatosis, Watson's syndrome, Lentiginosis profusa, Noonan syndrome, facial and ocular alterations, cardiovascular abnormalities, muscle and bones malformations, and neurologic deficits and in association with malignancies, i.e., squamous cell carcinoma of esophagus, adenocarcinoma of prostate, and small cell lung cancer rarely.[8]

Microscopically, GCT is composed of sheets of large polygonal tumors cells having abundant eosinophilic granular cytoplasm, round to oval centrally or eccentrically located nuclei with mild variation in size, and occasional small nucleoli. The granularity of the cytoplasm is caused by the massive accumulation of lysosomes which gives a positive reaction to CD68 and periodic acid–Schiff stain.[9] However, CD68 reaction was negative in our case. In the present case, the tumor cells were positive for S-100 protein and NSE, which supports the Schwann cell differentiation.

Malignant GCTs account for <2% of all GCTs. Malignant GCTs are usually large and deep-seated.[1] Fanburg-Smith *et al.* classified GCT on the basis of six histological criteria as benign, atypical, and malignant. These criteria include necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity (>2 mitoses/10 high-power fields

at ×200 magnification), high nuclear-cytoplasmic ratio, and pleomorphism. Tumors that fulfill 3 or more criteria were classified as histologically malignant, one or two criteria were classified as atypical, and those that displayed only focal pleomorphism, but fulfilled none of the other criteria, were classified as benign.[1]

The origin of GCT is still unknown and ongoing.[2] The cell of origin is now accepted to be the Schwann cell. It shows strong S-100 protein expression on immunohistochemistry.[10]

GCT may give an impression of malignancy to the clinician, but a careful microscopic examination with attention to size can resolve the confusion. The common preoperative clinical diagnoses are dermatofibroma, fibromatosis, keloid, or lipoma. The differential diagnosis includes rhabdomyoma, rhabdomyosarcoma, alveolar soft part sarcoma, hibernoma, dermatofibroma, dermatofibrosarcoma protuberans, schwannoma, neurofibroma, paraganglioma, oncocytic neoplasms, leiomyosarcoma, melanoma, and metastatic renal cell carcinoma. Rhabdomyosarcoma reveals characteristic rhabdomyoblasts with positivity for desmin and myogenin. Alveolar soft part sarcoma shows marked nuclear pleomorphism and prominent nucleoli, with an alveolar arrangement. In hibernoma, the tumor cells have vacuolated cytoplasm. Paraganglioma does not occur in the extremities and shows moderate nuclear pleomorphism, with positivity for chromogranin and synaptophysin. Epithelioid sarcoma displays vesicular nuclei with large nucleoli in the background of necrosis and inflammatory cells and positivity for cytokeratin and epithelial membrane antigen. Metastatic renal cell carcinoma is negative for the S-100 protein.[2,9]

Surgical wide excision with safe margins is the main-stay of treatment. Follow-up is necessary to monitor the recurrence and malignant transformation. Lack *et al*. reported an 8% recurrence rate in surgically treated cases.[10] The malignant variant is aggressive and can metastasized to bones, lungs, liver, and regional lymph nodes. Wide *en bloc* excision is recommended for malignant lesions. Chemotherapy, alone or in association with radiotherapy, is useful in malignant GCTs.[1]

## CONCLUSION

The GCT of hand is rare. Complete resection with disease-free margins is usually curative for benign GCT. Follow-up is required to monitor the recurrence and malignant transformation.

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### Conflicts of interest
There are no conflicts of interest.

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