

Case Report

A rare case of cutaneous granular cell tumour

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ABSTRACT

Granular cell tumors are uncommon tumors of uncertain etiology. It accounts for approximately 0.5% of all soft tissue tumors. However, the involvement of skin is rare. Only few cases of cutaneous granular cell tumor is reported till date. Here, we present a case of cutaneous granular cell tumor in a 48 years female patient.

Keywords: Eosinophilic, Granular cytoplasm

INTRODUCTION

Granular Cell Tumor (GCT) is a soft tissue tumor with neuroectodermal differentiation. Granular cell tumor is rare and accounts for approximately 0.5% of all soft tissue tumors.¹ Manifestation in the skin is in the form of a dermal or subcutaneous tumor with abundant granular cytoplasm and showing schwannian differentiation. Epidermis may exhibit pseudoepitheliomatous hyperplasia. Common sites affected are peripheral soft tissues (38%), trunk (36%), upper limbs, head and neck. Tongue is the most common location in the head and neck region. Pathogenesis of conventional granular cell tumors is unknown and a subset of non-neural granular cell tumors harbouring ALK gene fusions are rare lesions reported in the skin. Immunohistochemical staining for S100, SOX10, and CD 68 are known to be 100% positive.

CASE REPORT

A 48 years old female, presented with a swelling over lower back, lateral to midline, which was present for the past five years. There was a clinical diagnosis of calcinosis cutis with a differential diagnosis of dermatofibrosarcoma protuberans. On examination, a well localized 7×5 cms soft tissue mass was located in the back lateral to midline on the left side. The skin

underlying the lesion was remarkable. The patient was otherwise well with no other medical conditions. There was no family history of any malignancy or cutaneous lesions. The lesion was excised and sent to pathology for histopathological evaluation. No dissection of lymph nodes and adjuvant treatment was performed.

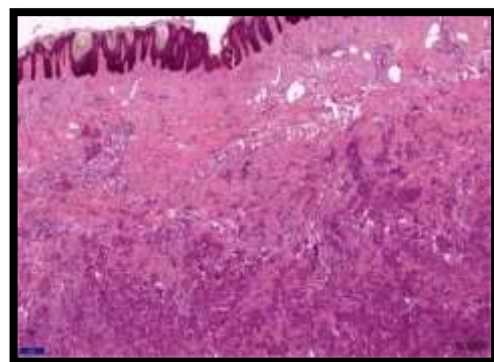


Figure 1: Overlying acanthotic epidermis with dermis showing irregular infiltrate of polygonal cells with abundant granular eosinophilic cytoplasm (H and Ex100).

On gross examination, the lesion was firm, well circumscribed, skin covered weighing 25 gm and

measuring 7 cm in greatest dimension. External surface of the lesion was unremarkable. The cut surface of the lesion was grey white in color and firm in consistency with infiltrative borders. Deep resected margins were also unremarkable.

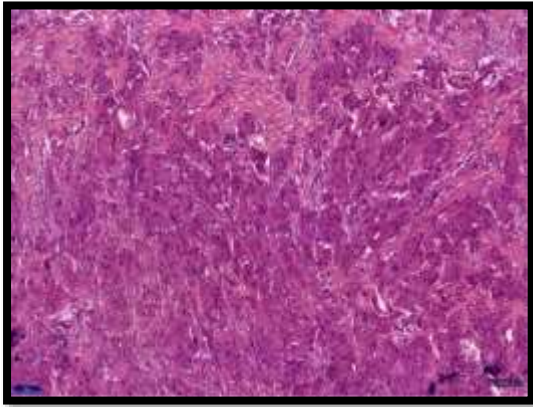


Figure 2: Dermis showing polygonal cells with abundant granular eosinophilic cytoplasm (H and Ex200).

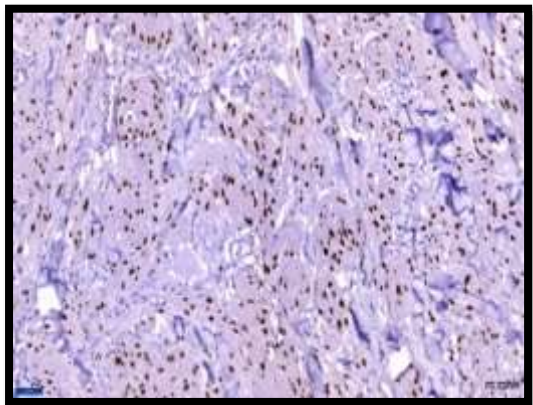


Figure 3: Tumor cells with SOX 10 nuclear positivity (IHCx200).

Histopathology showed skin with normal epidermis, with the dermis showing an infiltrate of large polygonal cells with abundant eosinophilic granular cytoplasm and small central nuclei, vesicular nuclear chromatin and mild nuclear atypia (Figure 1, 2).

The cytoplasm stained for Periodic acid Schiff stain (PAS) and Periodic acid Schiff stain–diastase (PAS-D) and immunohistochemistry showed a nuclear positivity for SRY-Box transcription factor 10 (SOX-10) (Figure 3)

DISCUSSION

Granular cell tumor was originally named granular cell myoblastoma, but currently, it is considered to be neural in origin according to immunohistochemical studies. Granular cell tumor accounts for 0.5% of all soft tissue

tumors.² Granular cell tumor is a multifocal tumor usually presenting in the fourth to sixth decades of life with a slight preponderance in adult females. The tumor can be localized on the skin or submucosa of various locations.

In 30-45% of cases, it can affect the skin, followed by head and neck, where the most common location is the tongue and oral cavity.³ Other locations are the breast, the gastrointestinal tract, the respiratory tract, the thyroid gland, urinary bladder and genitalia.⁴

Granular cell tumor usually presents as a painless mass with overlying normal skin, occurring mostly in the 30-50 years age group.⁵ Women are more commonly affected with a female to male ratio of 1.8-2.9:1. The best imaging modality for the characterization of this tumor is magnetic resonance imaging (MRI). Histopathology shows the presence of characteristic large polygonal cells with abundant eosinophilic cytoplasm. There are six histological parameters to distinguish benign from malignant Granular cell tumors. This includes presence of necrosis, spindling of the nuclei, vesicular nuclei with large nucleoli, increased mitotic activity (>2 mitosis/10 high power field), high nuclear to cytoplasmic ratio and nuclear pleomorphism.^{6,7} Tumors with one or two criteria can be classified as atypical. Also, GCTs of the skin may show locally invasive features. Battistella et al⁸ reported that the tumor infiltrated arrector pili muscles in 23% of 119 cases and had a perineural spread in 66%. The diagnosis usually can be confirmed by the histomorphology and immunohistochemical profile.

Neoplasms that meet three or more of these criteria are classified as histologically malignant, which may result in death in 40% of cases because of the high chance of local recurrence and metastasis; and those that display only focal pleomorphism but fulfilled none of the other criteria are classified as benign with no metastasis or local recurrence after adequate resection. Poor prognostic factors associated with Malignant granular cell tumor (MGCT) include large tumor size, older patient age, increased mitotic activity and Ki-67 labelling index greater than 10%.⁹

Surgical excision with a clean margin is the best treatment for this tumor and patient recovers and responds well to the surgery. Our patient also had surgical clearance and there is no evidence of any recurrence.

Local recurrence and metastases are common in malignant GCTs, presenting a year or two after the initial diagnosis, and distant metastases often occur in the lung, liver and bone. Thus, a sentinel lymph node biopsy during the initial surgical resection should be necessary.

CONCLUSION

Biopsy confirmation is important in diagnostically challenging cases which may rarely metastasise, if not

excised adequately. Hence knowledge of this entity, as a close differential is very important.

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