

INTRODUCTION

Granular cell tumors (GCTs) (Abrikossoff tumors) are rare, usually **benign soft tissue neoplasms** from **Schwann cells**. Typically solitary and smaller than 3cm located in dermis, some may be locally aggressive and <2% are malignant. More common in women with **peak incidence in 4th to 6th** decades of life. Here we present a case of childhood granular cell tumor.

CASE HISTORY

A 7yrs old girl came with complaints of painful swelling over both upper limbs for 6 months. Examination revealed skin coloured well circumscribed firm tender nodules of size 1*1cm over volar aspect of both forearms

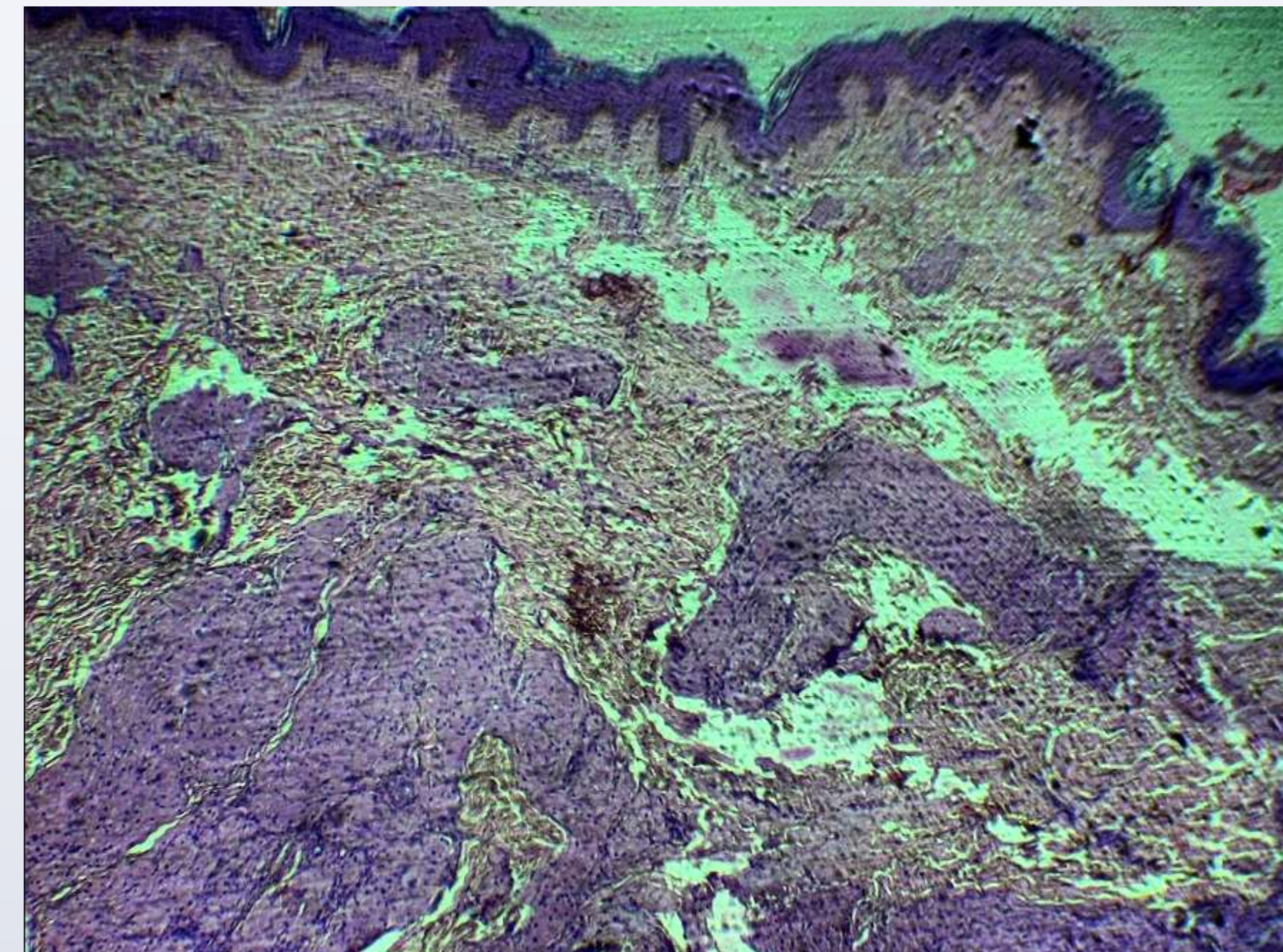


Two well circumscribed firm tender nodules of size 1*1 cm over the dorsal surface index finger of left hand

HISTOPATHOLOGY

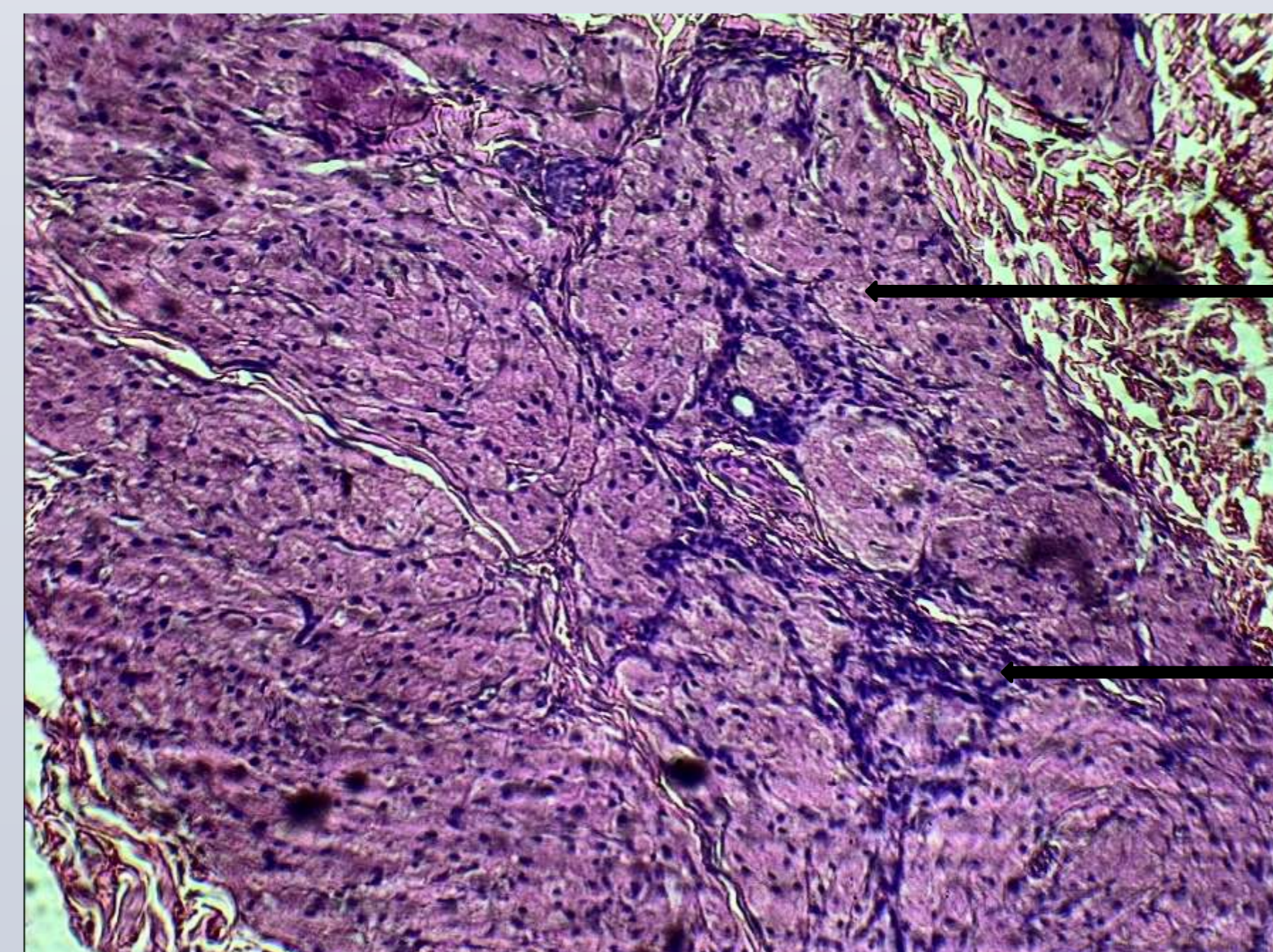
The differential diagnosis of xanthoma and leomyoma were considered clinically.

SCANNER VIEW



Poorly defined mass composed of sheets and nests of cells separated by collagenous bands

HIGH POWER VIEW



Round to polygonal cells arranged in syncytial pattern

Abundant eosinophilic cytoplasm with coarse granules (representing phagolysosome aggregates)

DISCUSSION

GCT s are usually single but when multiple, are associated with Noonan syndrome, LEOPARD syndromes. The most common sites are trunk, head and neck, but may also occur in GIT.

Loss of function mutation in **ATP6AP1** or **ATP6AP2** has been identified in these patients.

Gross examination shows nodular firm mass on cut surface shows yellow with finely granular texture.

Histopathological examination shows round to polygonal spindle shaped cells in a syncytial pattern with abundant eosinophilic cytoplasm containing coarse granules

Apart from typical H&E findings, the tumor cells stain positively for **S100**, **Neuron specific enolase**, **NK1-C**.

Wide surgical resection is the treatment of choice. Malignant lesion are aggressive with local recurrences and metastases.

CONCLUSION

GCT is a rare condition, often missed clinically. The HPE invariably clinches the diagnosis. This case is presented for its rarity, unusual childhood onset and to emphasize the role of HPE in the diagnosis.