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ATYPICAL GRANULAR CELL TUMOR OF THE ABDOMINAL WALL: A RARE TYPE

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INTRODUCTION

Granular cell tumors are rare mesenchymal soft tissue tumors which may occur anywhere in the body; However, they are rarely located in the abdominal wall. These tumors are mainly benign, but 1-2% of these lesions may exhibit malignant behavior. On the other hand, atypical GCTs are much rarer. They are neither benign nor malignant. Hence appropriate treatment with subsequent close monitoring of malignant behavior is required.

CASE REPORT

We report a case of 44-year-old lady who presented with right upper abdominal mass for 3 months. On examination, a 5x3cm mass with irregular margin over right hypochondrium was palpated. Radiographic appearances were consistent with possible rectus sheath hematoma or desmoid tumor. She underwent a wide local excision of the mass. Intra-operatively, an elliptical incision was made and dissection was carried down to pre-peritoneal space. The lesion was confined at the muscular layer, with multiple large feeding vessels. Abdominal wall defect was repaired with mesh. Histopathological examination showed poorly circumscribed tumor composing of neoplastic cells infiltration arranged in nest and cord. It also composed of large polygonal cells with vesicular nuclei and prominent nucleoli. Otherwise, mitosis and spindling area were hardly seen, and no necrosis observed. Immunohistochemistry revealed the granular cytoplasm stained strongly for S100 and weakly for CD68. They were negative for CKAE1/AE3. Post-operatively patient recovered well and was discharged on day 3 after surgery. Subsequent follow-up showed no evidence of recurrence.

DISCUSSION

Granular cell tumor (GCT) is diagnosed through histology with their significant immunoreactivity towards S100 and CD68. GCTs occurring at abdominal wall are extremely rare, regardless of benign or malignant [1]. Up till year 2020, only 30 cases of abdominal wall GCTs were reported, with only 1 out of the 30 cases was reported as atypical [2][3]. Histopathological diagnosis of malignant GCTs is based on the six features of the Fanburg-Smith criteria. In our case, only two criteria were met, making it an atypical GCT. Interestingly, lymphovascular invasion was also demonstrated, though it is not a criterion of the Fanburg-Smith criteria. Mainstay of treatment is complete excision to negative margins with close clinical follow-up with ultrasound assessment. In our case, the large abdominal wall defect is a surgical challenge which a composite mesh (Symbotex™) was used for the repair of the defect.

CONCLUSION

Granular cell tumors are extremely rare in abdominal wall. It is often misdiagnosed as desmoid tumor which is relatively more common as compared to GCTs. Surgical excision with negative margins is the mainstay treatment. In this case, abdominal wall repair was done with mesh cover in view of large abdominal wall defect. Ultrasonography follow-up is recommended to identify tumor recurrence, particularly in large GCTs (more than 4cm), as well as atypical and malignant tumors. Chemotherapy or radiotherapy effectiveness remains unproven in GCTs.

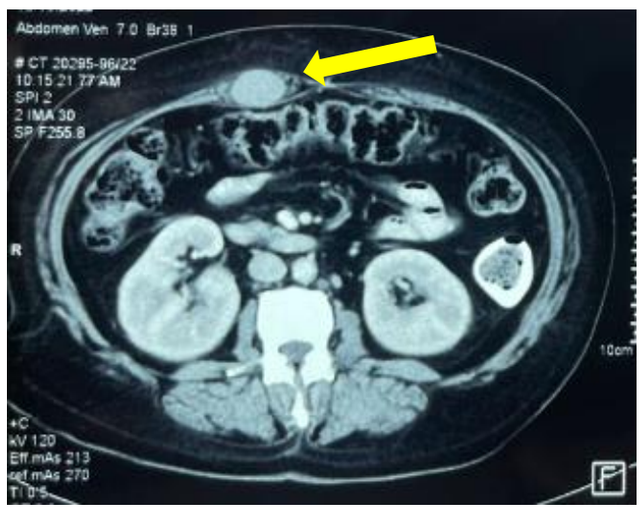

Fig. 1: CECT abdomen revealed right rectus abdominis intramuscular lesion measuring up to 55mm. There is no intraabdominal extension, no internal calcification or fat component.

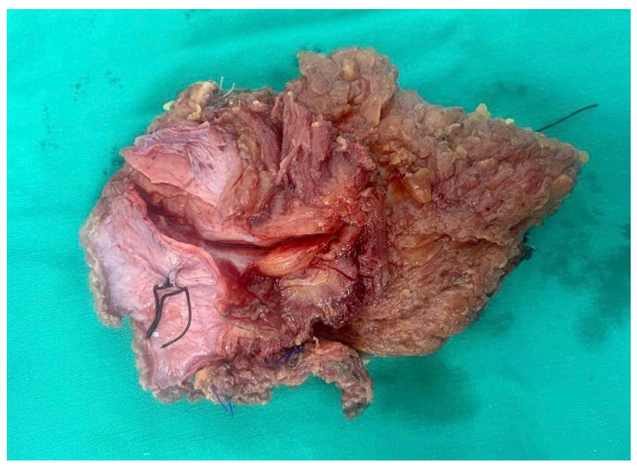

Fig. 2: Cut surface of the underside of operative specimen revealed homogenous whitish lesion

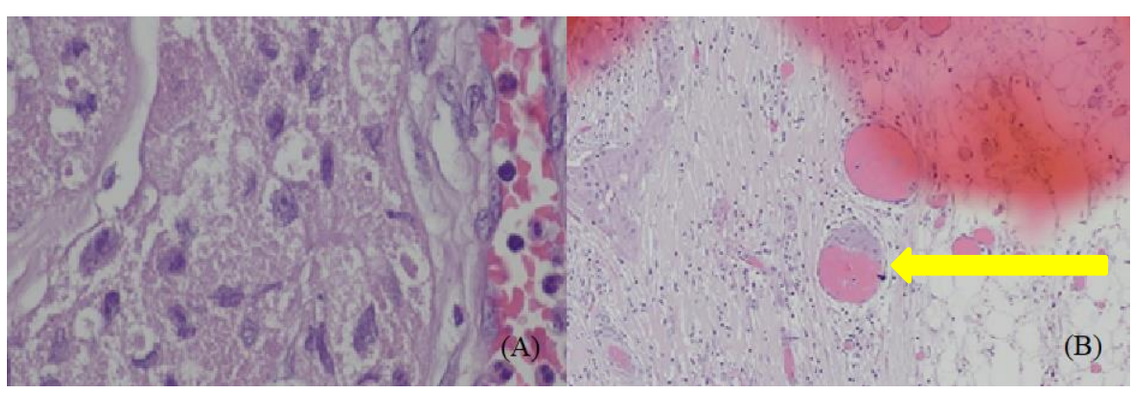

Fig. 3: Photomicrograph images of the GCT: (A) Tumor cells with vesicular nuclei and prominent nucleoli (x60); (B) Lymphovascular invasion is present.

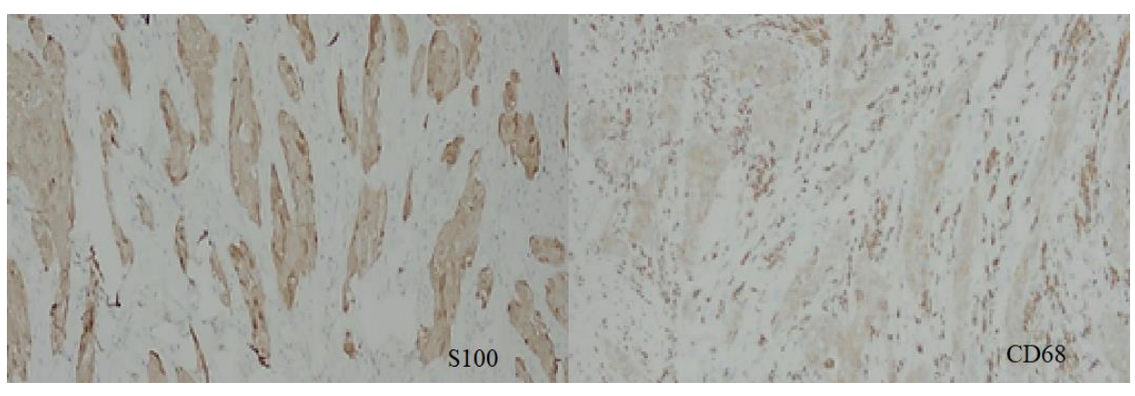

Fig. 4: Immunohistochemical staining, the granular cytoplasm is reactive to S100 (strong) and CD68 (weak); negative for CKAE1/AE3.

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