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Available online at: http://www.iajps.com *A Case Report*

ABDOMINAL WALL DESMOID TUMOR: TYPICAL PRESENTATION OF A RARE DISEASE.

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Abstract:
Desmoid tumors are rare slow growing benign but locally aggressive tumors with known local recurrence and no metastatic potentials. They account for 3% of all soft tissue tumors and 0.03% of all neoplasms. Definitive treatment of abdominal wall desmoid tumor is wide local excision with reconstruction of the defect. We report a case of a young female patient who presented with right lower abdominal wall mass. Preoperative evaluation included ultrasonography, CT scan and a diagnostic tissue biopsy. The patient was treated successfully by laparoscopic surgery with wide local excision.

Key Words: *Desmoid, tumors, abdominal wall.*

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INTRODUCTION:

Desmoid tumors (DT) also known as aggressive fibromatoses are very rare slow growing benign tumors. It originates from fascial or muscle aponeurotic structures that foster fibroblast proliferation. Desmoid tumor account for 3% of all soft tissue tumors and 0.03% of all neoplasms. Although aggressive fibromatosis does not have the ability to metastasize, it is characterized by locally aggressive growth with destructive infiltration of the surrounding tissues and high rates of local recurrence despite surgical resection, leading to significant functional impairments and morbidity. It has an estimated incidence of 2-4 cases per million people, they are usually diagnosed among fertile female patients between the ages 12-40 [2]. DT is rare during menopause, and it increase in size during pregnancy, which supports the estrogen-stimulated tumour growth hypothesis [3]. 37-50% of desmoid tumors are localized in the abdominal area, and in some cases, they are entailed in the areas of previous trauma, scars or irradiation [4]. 5-10 % of abdominal desmoid tumors is associated with familial adenomatous polyposis (FAP) and it is a major cause of morbidity and mortality especially in patients who undergone colectomy [5].

This report presents a case of abdominal wall desmoid tumor in a young female who had a history of caesarean delivery. This is the second case of abdominal wall desmoid tumour reported in Saudi Arabia. Informed consent was obtained from the patient prior to the study.

CASE PRESENTATION:

A 28-year-old female presented with right lower abdominal wall painful mass for two weeks. The pain is aggravated by movement and coughing and not radiating to other sites. The patient had caesarean delivery 2 years ago. On physical examination there was a tender mass in the right iliac fossa, measuring 6 cm in maximum dimension, with a smooth surface. The mass was oval, hard, non-pulsatile, non-compressible and non-reducible. The skin overlying the mass moves freely. The regional lymph nodes were not palpable. Superficial ultrasonography revealed subcutaneous irregular shaped ill-defined, homogenously hypoechoic lesion, measuring (60 x 30 mm). The lesion is vascular, showing regular arterial vascularity (Fig.1). CT scan of the abdomen and pelvis was done and revealed hypodense slightly enhancing soft tissue solid mass within the right rectus abdominus muscle at the level of iliac crest measuring (6 x 8 x 3.5 cm). The mass was localized at the muscle, and totally separable from underlying structure suggesting the diagnosis of abdominal wall desmoid tumour (Fig.2, 3,4). Ultrasound guided True-Cut biopsy was performed, and histological examination revealed proliferation of elongated slender spindle cells of uniform appearance arranged in sweeping bundles and set in a collagenous stroma containing blood vessels with mild perivascular edema. The cells have a small non-hyperchromatic staining nucleus with 1-3 minute nucleoli and atypia. The tumor cells show scattered mitotic figures. No necrosis is seen (Fig.5). Immunochemistry stains were performed which revealed that the tumour cells show variable nuclear and diffuse cytoplasmic staining for Beta-Catenin. The tumor cells are flat negative for C-KIT and desmin. Therefore, histopathology examination supported by immunohistochemistry staining confirmed the diagnosis of desmoid tumor.

After preoperative workup, the patient was planned for laparoscopic excision of the tumor. Three trocars were used, the tumor borders were easily identified and stay sutures were used for easy manipulation. The mass was completely resected using Ligasure® device with 1 cm margin from surrounding muscles. The aponeurosis of the external oblique muscle was intact. The resulting defect was approximated by separated sutures using 2.0 synthetic absorbable thread and reinforced with a 15x10 cm Parietex™ composite mesh fixed in place using AbsorbaTack™ fixation device. The specimen was extracted using a laparoscopic retrieval bag after enlarging the camera port site (Fig.6).

The final histopathological report of the resection confirmed desmoid tumor with negative surgical margins. The postoperative course was uneventful and the patient was discharged on postoperative day two. She was followed in the out-patient department after 1 week and 2 weeks and will have a follow up CT scan after 6 months.

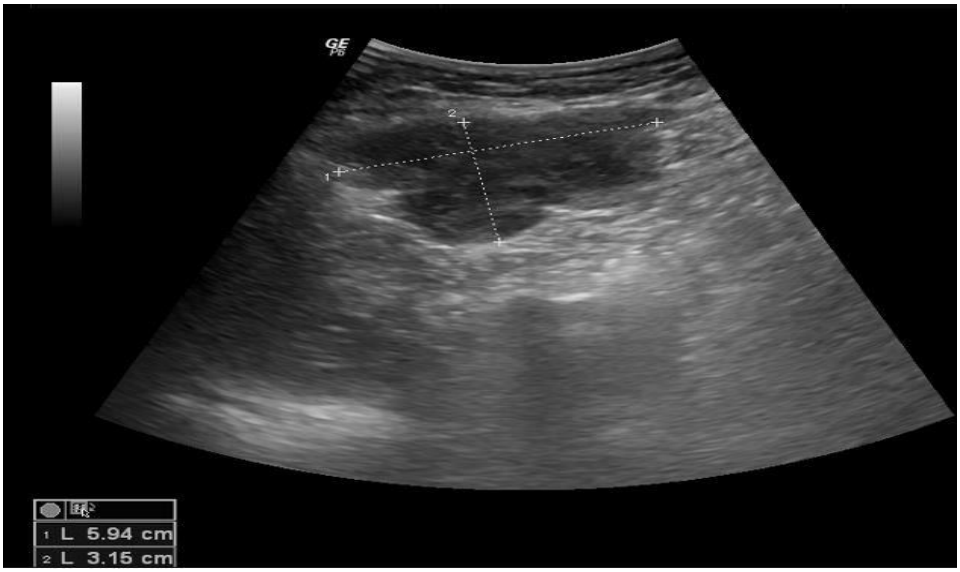

Figure 1: Superficial ultrasonography revealed an irregular shape ill-defined homogenously hypoechoic lesion, measuring (60 x 30 mm)

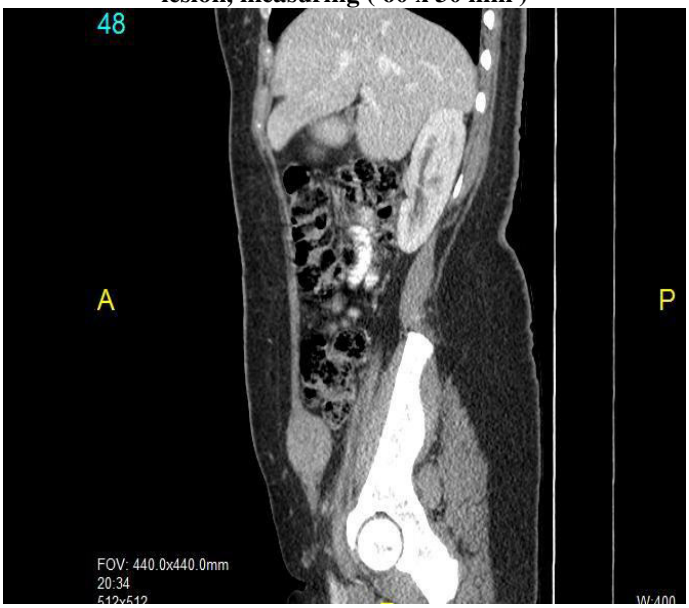

Figure 2 sagittal cut

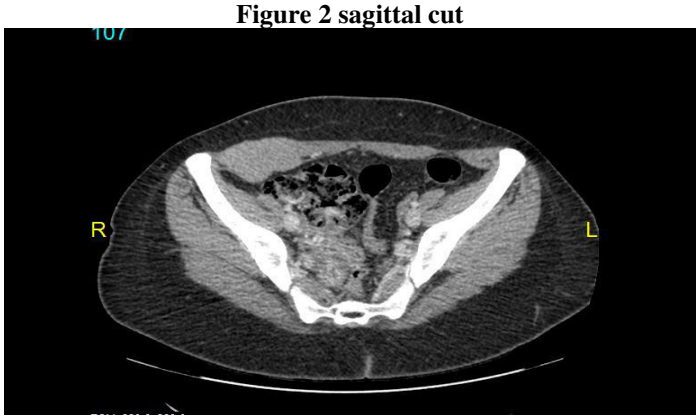

Figure 3 Axial cut

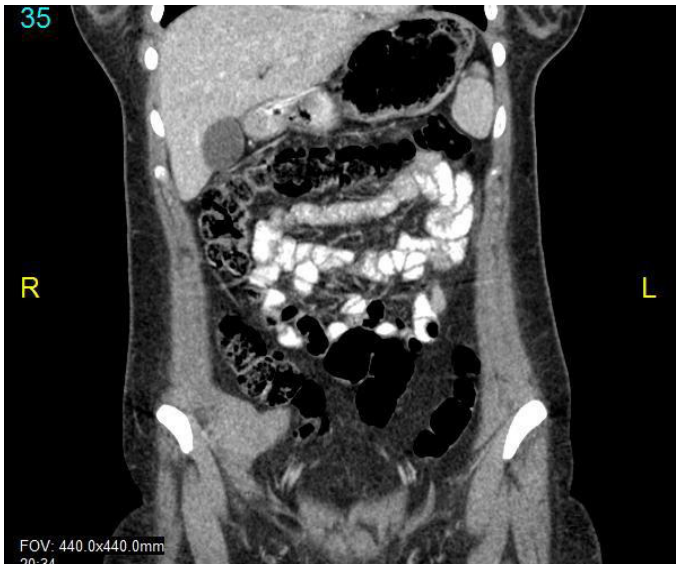

Figure 4 coronal cut

Figures 2,3,4: CT scan demonstrates hypodense slightly enhancing soft tissue solid mass seen at right rectus abdominus muscle at the level of iliac crest measuring (8 x 6 x 3.5 cm). The mass localized at the muscle and totally separable from underlying structure.

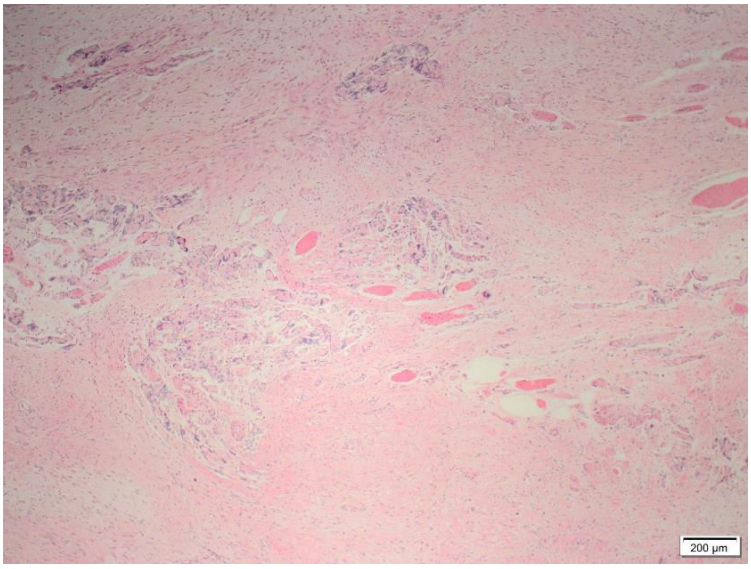

Figure 5 Microscopic view of the excised abdominal wall desmoid tumor demonstrating bland appearing hypocellular spindle cells infiltrating skeletal muscle fibres with atrophic and degenerative changes of the muscle; Hematoxylin and eosin staining).

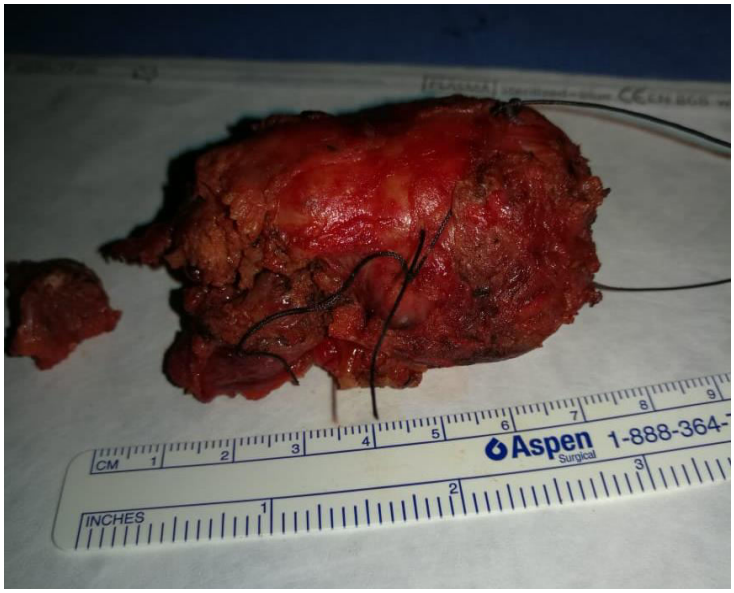

Figure 6: Macroscopic view of the excised abdominal wall specimen.

DISCUSSION:

Desmoid tumors are benign fibrous neoplasm originating from fascia and muscle aponeurosis with an infiltrative growth. They can be classified into intra-abdominal, extra-abdominal, multiple, familial or as part of FAP syndrome [6]. There is an association with female gender of child-bearing age, familial adenomatous polyposis, surgical trauma and estrogen hormone that acts as tumor growth factor, which makes the prevalence of desmoid tumor to be higher in women who experienced pregnancy and rare in males. Recurrence rate of DT depends on the tumour size, therapy and negative surgical margin but overall recurrence occurs in up to 45% [7]. Our patient was a 28 years old female in her child-bearing age, she had a history of pregnancy and surgical trauma which supports the literature. Several imaging methods are used for the diagnosis of desmoid tumours including ultrasonography, CT scan and MRI. On ultrasonography, DT appear as a well-defined smooth lesion with variable echogenicity. CT scan can localize the tumor and excludes metastasis and it demonstrates ill- or well-defined lesion with high attenuation relative to the muscles. On MRI, the findings that support the diagnosis of DT include poor margination, low-signal intensity on T1-weighted images and heterogeneity on T2-weighted images, and variable contrast enhancement. Beside imaging methods, a definitive diagnosis must be established with histopathological examination. Characteristic histopathological features of desmoid tumor include infiltrative spindle cell proliferation of uniform appearance set in a collagenous stroma and infiltrating skeletal muscle fibres. Immunohistochemistry staining can provide a valuable tool in supporting the diagnosis. DT cells can be partially positive for actin however they are negative for desmin [8]. Most helpful is staining for Beta-Catenin which is highly supportive for DT. Definitive treatment of abdominal wall desmoid tumor is wide local excision with reconstruction of the defect and any surrounding tissue involved by the tumor must be resected as well. Local recurrence rate is 20% to 77% in the presence of positive surgical margins or incomplete tumour resection. Other treatment options such as radiotherapy, chemotherapy and endocrine therapy must be considered in patients with inoperable tumour, local recurrence or incompletely excised lesions. Metastasis has not been reported in any patients with desmoid tumor [9].

CONCLUSION:

DT should be considered in female patients of child-bearing age who present with abdominal wall mass.
Laparoscopic approach is safe and can be done in selected cases.
Patients need long follow up plan to early discover any potential recurrence.

Declaration:
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Conflict of interest:
The authors declare that there is no conflict of interest regarding the publication of this paper.

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