Case Report

Diagnostic dilemma in a case of abdominal wall tumour: a case report

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ABSTRACT

Desmoid tumour or aggressive fibromatosis is a rare abdominal wall mass often confused with other conditions. Though imaging modalities are important for diagnosis of an abdominal wall mass, histopathology gives the definitive diagnosis. Here, we present a case history of a 28-year-old lady who presented with a painful swelling over the previous Caesarean section scar. Local examination was suggestive of a lipoma or scar endometriosis but a pelvic ultrasonography suggested a mass in the right adnexal region. So, with varied differentials in mind, laparotomy was done and ultimately it was diagnosed as a case of abdominal wall desmoid tumour on histopathology.

Keywords: *abdominal wall tumour, desmoid tumour, fibromatosis.*

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INTRODUCTION

Desmoid tumour (DT) is a monoclonal fibroblastic neoplasm originating from musculoaponeurotic structures. It is an infiltrative tumour and has a tendency toward local recurrence, especially when surgical resection is not complete but has no metastatic potential. They can occur anywhere throughout the body and are classified according to the site of origin; as extra-abdominal (in the chest wall, limb girdles, thigh and back), in the abdominal wall (in its aponeurotic structures) or intra-abdominal (involving the mesentery, pelvis or bowel wall). Almost 50% of the desmoid tumours occur in the anterior abdominal wall.[1] Clinical and radiographic features of abdominal wall desmoid tumours are often similar to other abdominal wall masses and have different compositions making the diagnosis difficult. A meticulous clinical examination along with ultrasound imaging can be very useful for distinguishing these masses. But lesions may have similar and overlapping features on imaging. So, histopathogy findings are necessary for a definitive diagnosis.[2] Here, we present a case of this rare medical entity mimicking an intra-abdominal adnexal mass on imaging.

CASE REPORT

A 28-year-old lady, mother of 2 children, both delivered by Cesarean section, presented with a painful swelling for 3 months over the previous Caesarean section scar. The patient noticed a swelling above the scar mark of previous Caesarean section done 3 years back, which was gradually increasing in size and became painful. The pain was localized and was not related to menstruation. She had no history of dysmenorrhoea. She is a known case of diabetes mellitus for 2 months.

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On examination, a lump was visible at right lower abdomen just above the previous Caesarean scar. Cough impulse was negative. On palpation, a parietal mass of about 7 cm x 6 cm size was felt in right lower abdomen which was firm, tender with nodular surface, well defined margin and it was free from the overlying skin but fixed to underlying structures. Per vaginal examination revealed no abnormality. So, our differential diagnoses were lipoma or scar endometriosis. A pelvic ultrasonography (USG) showed a fairly large (7.7 cm x 4.8 cm) mixed echogenic area in the right adnexal region (Figure 1). Tumour marker CA-125 was done which was 28 IU/L (normal < 35 IU/L).

Though clinically it seemed to be a parietal mass, the pelvic USG finding was conflicting. So, with the suspicion of adnexal mass, laparotomy was planned after counselling the patient and her family. A Pfannenstiel incision on the overlying skin revealed a bulging of rectus sheath on the right side where it was thinned out. There was a mass beneath the rectus sheath which was fixed to underlying rectus muscle (Figure 2 A, B). The mass infiltrated the rectus muscle at places. It was taken out with meticulous dissection. The mass was extraperitoneal and the lower border was free and not attached to any structure. Peritoneal cavity was opened and uterus, both the ovaries and adnexae were found healthy. There was no other tumour or cyst. Rectus muscle, sheath and overlying fascia were repaired with vicryl 1 suture. The excised tumour specimen measured 8.5 cm x 6.0 cm x 4.0 cm (Figure 3 A, B). Cut section looked grey white and whorled.

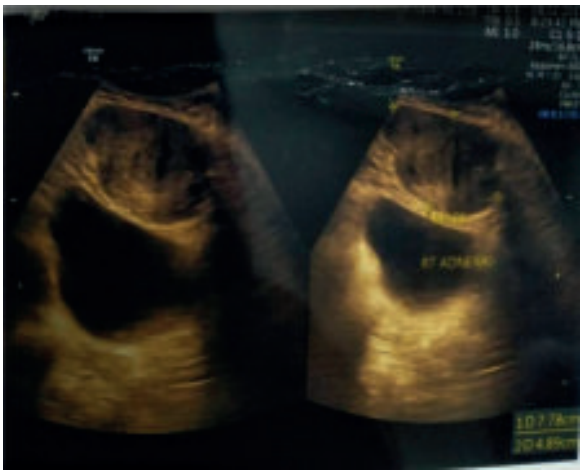


Figure 1. Pelvic ultrasonography findings were suggesting right adnexal mass

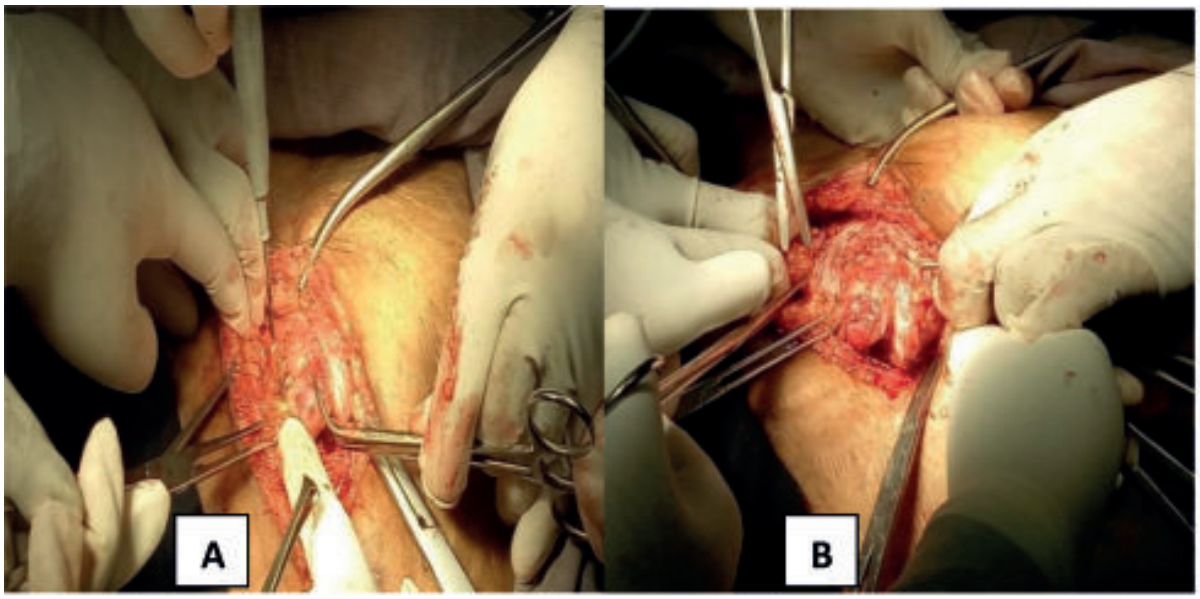


Figure 2. A. Parietal mass beneath the rectus sheath, **B.** The mass being taken out

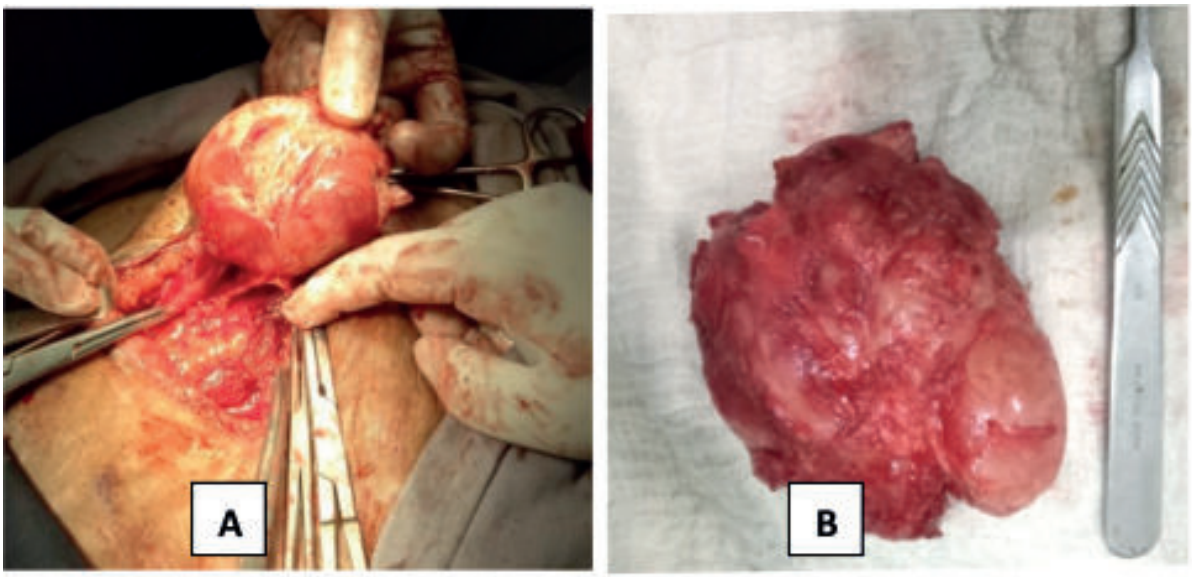
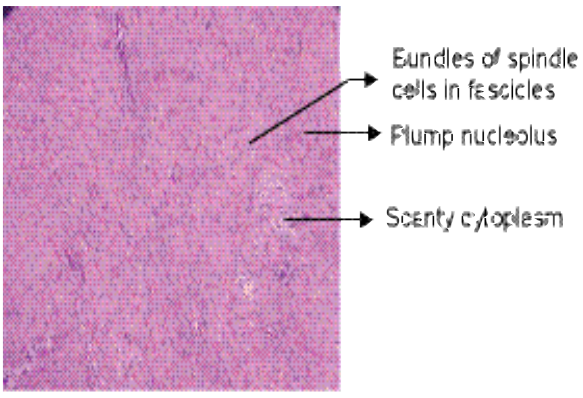


Figure 3 A, B. The resected mass

Microscopic examination revealed that the tumour tissue was composed of bundles of spindle cells arranged in fascicles, containing scanty cytoplasm and spindly slender to plump nucleoli (Figure 4). The tumour had an ill-defined border and had infiltrated into the skeletal muscle at places. Sections from the peripheral resection margin were free from tumour. Histopathological diagnosis was rectus muscle fibromatosis or desmoid tumour.



Bundles of spindle cells in fascicles

Plump nucleolus

Scanty cytoplasm

Figure 4. Microscopic image of the tumour (hematoxylin and eosin stained; x100 magnification)

The postoperative course was uneventful, and the patient was discharged on the third postoperative day. As there is chance of recurrence, the patient was counselled for further follow up and she remained well at subsequent follow-ups at one month, six month, and one year without any recurrence.

DISCUSSION

Desmoid tumour is an extremely rare entity with an incidence of 5-6 cases per 1 million of the population per year. They are more common among the age group of 30-40 years with a strong prevalence among women of reproductive age, during or within one year of pregnancy.[3,4] Several authors have reported that some of the risk factors for desmoid tumours are previous surgical interventions, excessive stretching of abdominal wall muscles and fascia during pregnancy and hormonal treatment with estrogen.[5] In a study by Guo et al.[6] 14 cases of abdominal wall desmoid tumourreported from 2021 to 2024 were reviewed where the age varied from the 20s to the 60s, with a significant predominance of female patients. History of surgery or trauma was present in most of the patients, including 4 cases of Caesarean sections.

In our case the patient was 28 years old and had a history of Caesarean section 3 years back which is one possible risk factor of the desmoid tumour. Regarding treatment and prognosis, most patients included in the study by

Guo et al.[6] receiving surgical treatment exhibited no recurrence during the short-term follow-up period, as in our case.

Abdominal wall desmoids usually measure 5 to 15 cm in size and possibly arise from the aponeurosis of the anterior abdominal wall muscles embedded within either the rectus abdominis muscle, internal oblique muscle or the fascia. Occasionally, the mass can cross midline and may extend into the abdominal cavity or may even erode the adjacent bone.[5]

The clinical presentation of desmoid tumour also varies; it may be asymptomatic or may have severely restricted function due to chronic pain and/or deformity or may be even disabling and is related to location, tumour size and growth rate. Spontaneous regression may occur in 20% to 30% of patients who are followed for 2 to 3 years.[7] It is observed that regression is more common in abdominal wall desmoid tumour.[3] Due to its variable characteristics and unpredictable course, desmoid tumourmay be misdiagnosed in up to 30% to 40% of cases.[7]

Imaging modalities like ultrasonography, CT scan and MRI play key roles in the diagnosis, follow-up, and evaluation of recurrence of desmoids. Among these, ultrasonography is the most cost effective tool.[1,5] On USG, it appears as an infiltrative solid mass with variable echogenicity depending on fibroblast proliferation, fibrosis, collagen content and vascularity.[1,5] On color Doppler, there is no or minimal flow. But, marked vascularity may be seen in cases of aggressive fibromatosis.[8] Imaging is also crucial in the evaluation of the responses to non-surgical treatment options especially after systemic treatment.

Final diagnosis of desmoid tumours are confirmed by histological examination. In this case, final diagnosis could be reached only after histopathogical of the resected tumour. But at times asserting the histological diagnosis also becomes challenging. During microscopic examination special attention to the differential diagnoses of desmoid tumour like fibrosarcoma and benign fibrous lesions should be kept in mind. The presence of spindle cells with long fascicles and infiltrative borders mimic malignancy but characteristic features of malignancy like herring bone pattern, higher cellularity, nuclear hyperchromasia, pleomorphism and increased mitotic activity are characteristically lacking in fibromatosis. It may also be confused with reactive fibroblastic proliferations which can be distinguished by the presence of areas of variable growth pattern, haemorrhage or haemosiderin deposition.[9]

Abdominal wall desmoid tumour has an unpredictable clinical course and the treatment outcomes are also uncertain. So, there is there is lack of any single consensus on the optimal treatment approach. A “wait and see” approach is currently preferred in newly diagnosed and asymptomatic patients. Traditionally surgical operations have been preferred in the treatment of desmoids tumours. However, considering the general complications of surgery (such as infection, hemorrhage and thrombosis) and the local recurrence rate, active surveillance may be adopted as an initial treatment strategy for slowly growing, asymptomatic cases, involving monitoring with CT scan or MRI every 3-6 months. Surgical mass resection should be preferred as a treatment option in patients who refuse observation, have increased mass size, rapidly enlarging mass, present symptoms that impair quality of life and in whom malignancy cannot be excluded. During surgery care must be taken to remove the mass along with tumour free margin to reduce the risk of recurrence.[10]

The recurrence rate of desmoids tumours is about 20% to 77% depending on the location, extent and completeness of the initial resection. Abdominal wall desmoids tumours have a significantly lower recurrence rate. Their recurrence is 20% to 30% and usually becomes evident within six months after excision or may reappear with subsequent gestations or after deliveries.[9] In our reported case, there was no recurrence at subsequent follow-ups at one month, six month and one year.

Abdominal wall reconstruction may be done by direct repairing the gap with sutures, as done in our case or by using synthetic mesh or myocutaneous flaps when the defect is large.[10] Early detection of a desmoid tumour and the treatment of a smaller tumour can prevent large defects and avoid the mesh reconstruction.

Abdominal wall mass in women of reproductive age has given rise to misdiagnosis of desmoid tumour as endometriosis in several cases reported previously since the clinical signs are similar, and the imaging findings are nonspecific.[11] Though imaging modalities are

important for diagnosis of an abdominal wall mass, microscopic findings and immunohistochemistry are essential for a definitive diagnosis. Long-term follow-up of patients with detailed history taking, physical examination and appropriate imaging every 3-6 months for 2-3 years, and then annually for 2-3 years are recommended, considering the possibility of local recurrence.[6] It is true that significant progress has been made in research of desmoid tumour over the past decades, but the rarity of DT results in limited case reports and treatment experiences.

Authors' contribution: SS designed the study, prepared, reviewed, and drafted the manuscript. FTN, AN and NN helped in data collection, case management and manuscript review. All authors read and approved final manuscript to be submitted.

Consent: Informed written consent was taken from the patient regarding publication of this case report and the relevant images.

Conflicts of interest: Nothing to declare.

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