

Recurrent Abdominal Wall Desmoid: A Case Report

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Case Report

Abstract: Desmoid tumors or desmoid fibromatoses are benign neoplasms arising from muscle and aponeuroses all over the body. The most common site for desmoids is the anterior abdominal wall. They are usually seen in women of reproductive age group. Recurrent desmoids make surgical excision and reconstruction a challenge due to the huge defect in the anterior abdominal wall. We report a 25year old female case of recurrent desmoid with previous surgical history of excision of an abdominal wall desmoid. Pre-operative USG (Ultrasound) Abdomen reports indicated a homogenous pelvic mass. Wide surgical excision of the tumor was done with safe margins. Subsequent histology revealed a desmoid tumor. Radical surgery is still the main stay of treatment of recurrent desmoids. Radiotherapy, chemotherapy and anti-estrogens are still questionable. Further research into their etiology is required combined with multicentric clinical trials of new treatments in order to improve management of this disease. This case report provides general knowledge of desmoid tumors, and may be used as guidance for diagnosis and treatment.

Keywords: abdominal wall; desmoid tumors; fibromatoses; mesh reconstruction; recurrent tumors

Case Report

A 25 year old woman presented to us with a painless lower abdominal mass gradually increasing in size for the past 1 year. She had a past history of surgery for an abdominal wall desmoid tumor 2 years back, for which a complete excision and polypropylene mesh repair was done at our hospital. The hospital lost follow up with the patient since then. She had no other relevant medical history or family history suggesting FAP (Familial Adenomatous Polyposis). She had full term normal vaginal deliveries for her two kids. On examination a diffuse lower abdominal mass was found involving the hypogastrium, the umbilical and both iliac regions. It was globular, fixed, non tender. USG revealed a large homogenous mass with smooth margins in the pelvis area extending into the pelvis measuring 18x16x15 mm. CT (Computed Tomography) scan was refused by the patient for financial reasons. She was scheduled for an Elective Laparotomy for a suspected recurrent desmoid tumor of the anterior abdominal wall. The surgery was done with a transverse lower abdominal incision at her previous incision site. On reaching the plane of the rectus sheath the tumor was identified to be originating from the lower

half of the rectus abdominus muscle involving the anterior abdominal wall muscles and fasciae and the mesh kept during the previous surgery (Fig 1). The tumor extended upto the pubic symphysis on its lower aspect. On its upper aspect the tumor extended intra-abdominally and was densely adherent to a portion of the small bowel and the omentum. The incision was converted to into an inverted 'T' shaped incision for better vision. The tumor was separated from the bowel by sharp dissection. Minimal serosal damage on the bowel wall was sutured. The tumor was completely excised with wide surgical margins (Fig 2). The rectus sheath was closed. The remaining muscle defect was repaired by primary suturing without tension. Mesh was not kept. Excess skin and subcutaneous tissue were excised and the abdomen was closed. The post-operative period was uneventful and the patient was discharged on the 7th day. Subsequent histopathology revealed the tumor to be a desmoid and confirmed negative margins.



Figure 1: Desmoid tumor in the anterior abdominal wall

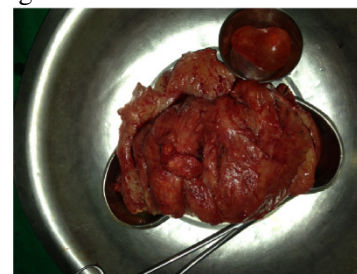


Figure 2: Excised desmoid tumor

Discussion

Desmoid tumors are uncommon benign neoplasms constituting 3% of all soft tissue tumors and 0.03% of all neoplasms [1, 9]. They are usually seen in multiparous females in the reproductive age group [1,2,4]. They maybe extra- abdominal, seen in sporadic cases involving proximal extremities or shoulder girdle, etc [8] or intra- abdominal (involving rectus abdominus muscle or the internal oblique) which maybe familial (FAP, Gardner's Syndrome). They mostly involve the anterior abdominal wall. They do not metastasize or

dedifferentiate but are locally infiltrative and recurrent tumors. They have been described as rare entities in our country but they prove to be very daunting when encountered especially with mutilating surgeries being the mainstay of treatment. Although the diagnosis of a desmoid can be suspected from the history, CT or MRI, a definitive diagnosis can be made only from histology. In most cases the diagnosis is made intra-operatively based on its origin and local infiltration. A pre-operative MRI followed by histopathology is the way to go forward with a case. An MRI usually indicates poor margination, low intensity on T1 weighted images, heterogeneity on T2 weighted images and variable contrast enhancement [1, 3]. Histology usually indicates spindle cells interspersed with partially intact muscle fibres interrupted by fibrotic sections. Immunohistochemical response for actin can be partially positive delimits desmoid tumors from fibrosarcomas [3, 4]. Their propensity to recur has been attributed to factors like positive surgical margins, or the tumor size or the choice of therapy. Some researchers believe that recurrences occur irrespective of surgical margins or whether radiotherapy was given adjuvantly [2, 8]. Tumor size of greater than 5cm has been an important predictor in some cases [2]. Lack of adequate knowledge of the etiology of the tumor has made the management choices to be individualised and patient centric. Complete resection of tumor with safe margins and reconstruction of the abdominal wall defect remains the mainstay of treatment for primary or recurrent abdominal wall desmoids.[1-4,8,9]. Restoring abdominal wall integrity after mutilating surgeries has proven to be quite a challenge especially with massive abdominal wall desmoids[3,4]. It can be restored by direct suturing [5], or the more widely practised method of reconstruction with synthetic polypropylene mesh [1,4,5]. Recent literature data recommend using free muscle flaps for greater abdominal wall defect coverage which may not be accessible to local flaps [4,5]. Radiotherapy in the management of recurrent desmoids has been extensively studied but its definitive role in treatment is yet to be established [2]. It is at present indicated for cases of inoperable tumors, or incomplete excision or positive margins [6]. Its use as definitive therapy or adjuvant therapy for cases with positive margins did not appear to influence the local recurrence rate [2]. Alternate modalities of therapy have also been suggested. Patients with multiple locoregional recurrences despite adequate local therapy are considered for systemic therapy[7]. Systemic therapy may also be considered with intra-abdominal desmoids in FAP and Gardner's syndrome. Chemotherapy with doxorubicin and dacarbazine have proven to be clinically useful[6, 7]. Anti-inflammatory drugs like Indomethacin, sulindac and anti-estrogens like

Tamoxifen have also been suggested[1,7]. But these lesser tried modalities are to be used when the conventional methods have been tried and proven to be unsuccessful. Recurrent desmoids usually surface in a minimum of six months. Stringent follow up usually improves our chances of better outcomes with conventional therapy [1, 2,9].

Conclusion

A diagnosis of an abdominal wall desmoid although a rare entity should be a strong primary diagnostic consideration in a woman in the reproductive age group irrespective of the imaging studies which may be misleading and thus prove a surprise during laparotomy. A complete resection with negative margins and reconstruction of the defect is the mainstay of treatment of both primary and recurrent abdominal wall desmoids.

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Ethical Statement: The manuscript does not contain clinical studies or any revealing patient data.

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