Case Report

Giant anterior abdominal wall desmoid tumor successfully managed with abdominal wall reconstruction

Dhivakar S[1], Farhanul Huda[1], Sudhir K. Singh[1], Arvind Kumar[2], Asish Das[1], Preeti Acharya[1]

[1]Department of General Surgery, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India, [2]Department of Pathology and Laboratory Medicine, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India

Abstract

Anterior abdominal wall fibromatosis is a benign soft tissue tumor that is rare, but fast-growing with minimal chances of malignant change. We report a young female with a large abdominal swelling which on evaluation was provisionally diagnosed as anterior abdominal wall fibromatosis on imaging and confirmed by histopathology. She was successfully managed with resection of the tumor with a challenging abdominal wall reconstruction with bilateral inferiorly based external oblique muscle flap followed by a mesh repair. Though rare, these tumors are difficult to miss. The importance of this case report is that it describes the methods of multimodal management of a patient with surgery, reconstruction, and adjuvant therapy leading to better patient outcomes.

Keywords: Desmoid tumor, oncology, plastic surgery, reconstruction, soft tissue tumor

Introduction

Abdominal wall desmoid tumors (ADT) are rare but locally aggressive benign tumors that arise from fascial and musculoaponeurotic tissue that invades the local tissue and neurovascular structures.[1] The incidence is approximated at 0.03% of all neoplasms and 3% of soft tissue neoplasms.[2] ADT is more common in females and more aggressive especially in women of childbearing age or those with a previous history of pregnancy as the tumor is encountered in increased estrogen states.[3,4] A core needle biopsy or an incisional biopsy often gives a pre-operative idea, which would be quite useful in planning surgery. These tumors can occur anywhere in the body. Hence, primary care physicians need to have a clear understanding of this disease, which would help in early diagnosis and prompt referral and better outcomes.

Case Presentation

A multiparous female in her 20s presented to our OPD with complaints of swelling in the lower abdomen, which was initially small and gradually progressed in size over a duration of one year. General physical examination was unremarkable. On abdominal examination, a 15 cm × 14 cm swelling was present in the lower abdomen over a lower midline incision, which was done for a cesarean section a few years back. The swelling was nontender, hard in consistency with well-defined margins and irregular surface. She underwent a core needle biopsy at a local facility and presented to us with a provisional diagnosis of anterior abdominal wall fibromatosis. Computed tomography (CT) of the abdomen showed a well-defined heterogeneously enhancing lobulated mass of 9.5 × 15 × 14.5 cm in the bilateral rectus abdominis in the anterior abdominal wall. Due to the coronavirus pandemic and nonfunctional operating rooms, she was given a

Address for correspondence: Dr. Dhivakar S, Department of General Surgery, All India Institute of Medical Sciences, Rishikesh - 249 203, Uttarakhand, India.
E-mail: kishankis94@gmail.com

Received: 25-02-2023 **Revised:** 14-06-2023
Accepted: 22-06-2023 **Published:** 29-08-2023

Access this article online

Quick Response Code:

Website: http://journals.lww.com/JFMPC

DOI: 10.4103/jfmpc.jfmpc_379_23

How to cite this article: Dhivakar S, Huda F, Singh SK, Kumar A, Das A, Acharya P. Giant anterior abdominal wall desmoid tumor successfully managed with abdominal wall reconstruction. J Family Med Prim Care 2023;12:1716-9.

trial of medical therapy with tamoxifen 20 mg once a day dosing for 3 months, but there was no reduction in size or symptoms.

Technique

After adequate preoperative workup, she was planned for resection of the desmoid tumor with abdominal wall reconstruction under general anesthesia with epidural analgesia. The tumor was superiorly extended up to the umbilicus and inferiorly to the public bone and abutting onto the fundus of the uterus. The tumor was resected completely [Figure 1]. After a satisfactory resection, a large abdominal wall defect of 25 cm × 12 cm was noted. The rest of the abdominal viscera were grossly normal.

The external oblique muscle was separated from the anterior sheath and internal oblique bilaterally. It was mobilized superiorly up to the costal margin and iliac crest laterally on both sides. Marking and measurements were performed for the external oblique flap. Flap was raised based on the subcostal vessels, rotated medially, and sutured with Vicryl 3-0. Adequate hemostasis was achieved, and commercially available polypropylene mesh was placed between the flaps and the subcutaneous plane [Figure 2]. A subcutaneous mini-suction drain was placed, and wound closure was done in layers with satisfactory outcomes [Figure 3]. The patient was managed post-operatively with regular dressings and had an uneventful recovery. After discussion in a multidisciplinary tumor board, she received 60 gray/30 fractions over 6 weeks as adjuvant treatment. On monthly follow-up, till one year she is hale and hearty. CT abdomen done at 1-year postoperative period showed no recurrence. The patient is under our active surveillance.

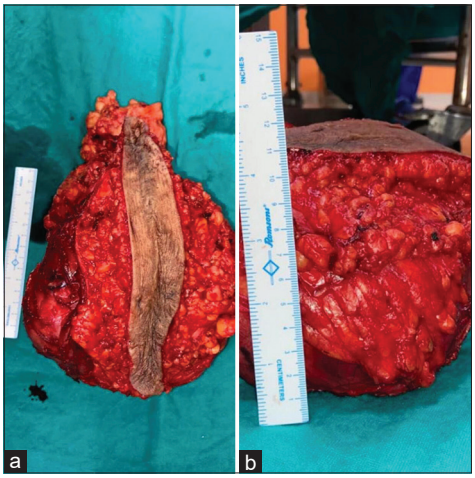

Figure 1: (a-b) Resected specimen

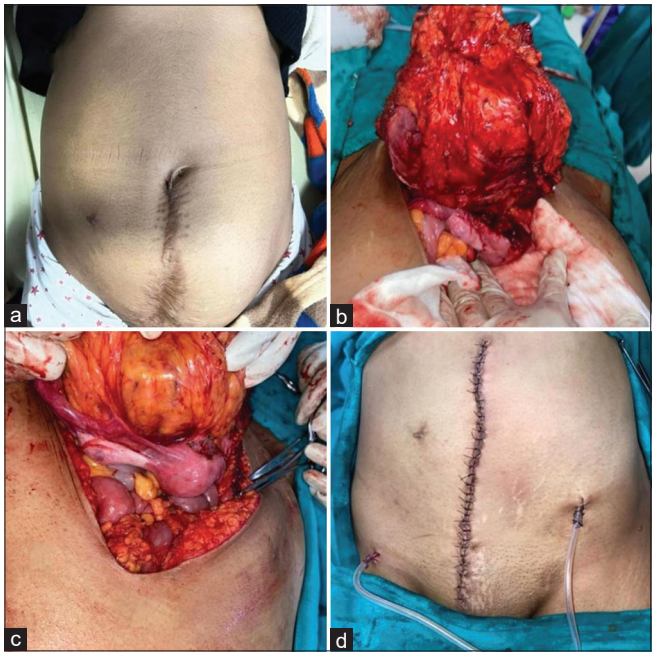

Figure 3: (a-d) Preoperative vs postoperative images

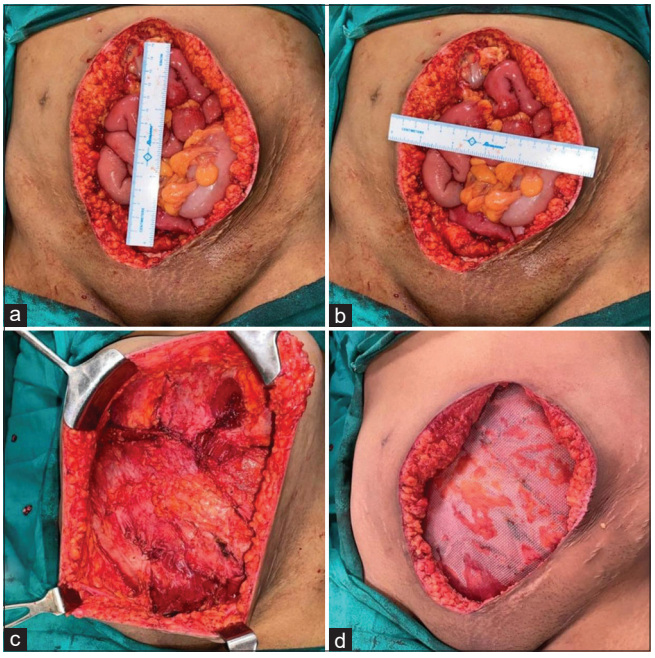

Figure 2: (a-d) Intra operative image of Resection and Reconstruction

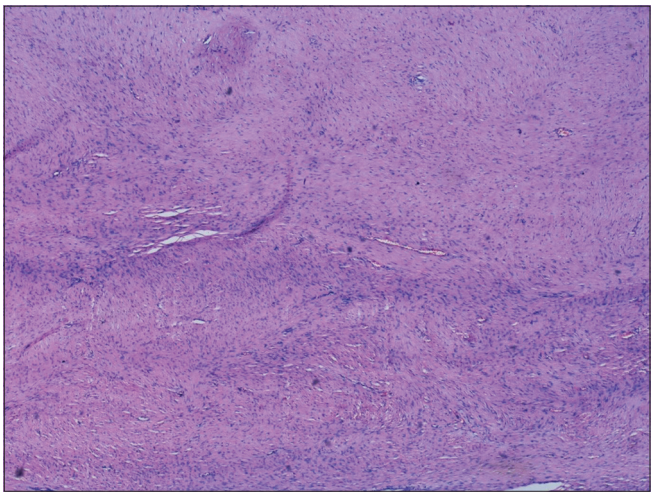

Figure 4: Shows fascicular pattern of monomorphic cells along with fibrocollagenous stroma. (H and E, 40X)

Discussion

Desmoid tumors (DT) are hormone-dependent tumors, the disease either develops spontaneously in women of reproductive age or is linked to inherited diseases like Gardner syndrome. Our patient had a history of surgical trauma in the form of a cesarean section, which is also thought to be one of the trigger variables, as supported by literature.[5] Whatever the cause, DT is an unpredictable lesion as the growth potential or tumor biology is poorly understood. Diagnosing DT is not a challenge as it is confirmed on a histopathological examination in most patients. Long fascicles of fibroblastic cells [Figures 4 and 5] with ill-defined cytoplasmic boundaries and weakly colored nucleoli are the microscopic characteristics indicative of DT.[6]

Several treatment options have been postulated including surgical resection, chemotherapy, hormonal therapy, and radiotherapy with variable success rates. Indomethacin, sulindac, and tamoxifen are a few chemotherapeutic agents tried, but their role is controversial and limited only to recurrent and unresectable cases.[7] In our patient, tamoxifen therapy was tried, but there was no significant reduction in the tumor. Improved patient outcomes may be linked to increased use of neoadjuvant therapies.

Here, the most challenging part was the achievement of R0 resection and abdominal wall reconstruction. The DTs of the anterior abdominal wall are special in that they do not involve the skin; hence, skin closure was easier, but the peritoneal defect needed to be closed under tension in our patient. A good reconstruction prevents any hernia at the operative site which otherwise would be troublesome to reoperate as surgery would be difficult in such cases. There have been numerous reports on the use of prosthetic mesh as a fascial substitute or reinforcement when soft tissue coverage over the wound is inadequate.[8] We did an external oblique-based flap repair, which was further reinforced by a polypropylene mesh was performed, but the choice of mesh reinforcement is surgeon specific.

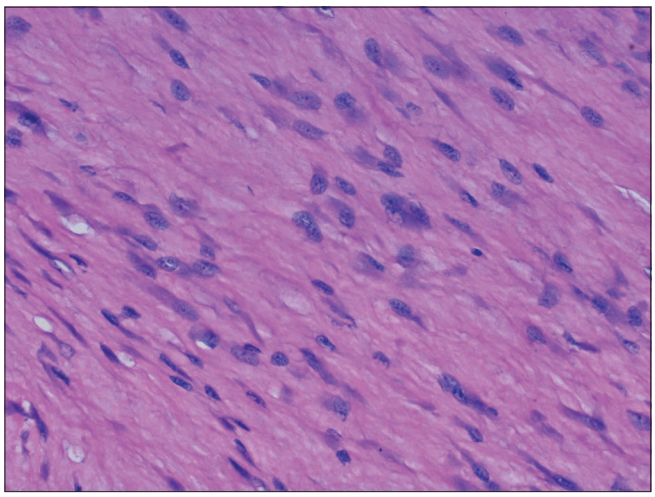

Figure 5: Tumor shows oval to spindle cells with bland vesicular nuclei, regular chromatin membrane, inconspicuous to conspicuous nucleoli, and indistinct cytoplasm. (H and E, 400X)

Radiotherapy is considered for unresectable tumors or those adjacent to nerves or major vessels. However, the best treatment modality described is the debulking surgery with postoperative radiotherapy in the operative field in case of positive margins. External beam Radiotherapy of 50 Gy has been reported with local reduction of the disease.[2] In some instances, RT is exclusively tried as a primary therapeutic choice in individuals who are unwilling to undergo surgery or who have unresectable fibromatosis.[9] Moreover, large-scale studies or evidence is not available to support this modality of treatment. We decided to give post-operative radiotherapy of 60 gray/30 fractions over 6 weeks as there were suspicious margins at the inferior border. Chemotherapy and surgery combined with a multidisciplinary approach are an efficient and function-preserving way to treat this disease.

Conclusion

Any woman belonging to the reproductive age group with a history of recent pregnancy or surgical trauma presenting with a fast-growing swelling of the anterior abdomen should point to the differential of fibromatosis (DT). The challenge in these cases is the reconstruction of the anterior abdominal wall, which is crucial.

Learning points/take-home messages

- Resectable DTs require a complete R0 resection with proper follow-up
- Treatment therapies are always to be discussed in an MDT so that the best outcome is achieved
- In unresectable cases, radiotherapy is a better treatment modality compared to other adjuvants as supported by the literature
- Resection has to be followed by the best possible reconstruction to avoid unnecessary surgical site hernias in future.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Goellner JR, Soule EH. Desmoid tumors. An ultrastructural study of eight cases. Hum Pathol 1980;11:43-50.

2. Nuyttens JJ, Rust PF, Thomas CR Jr, Turrisi AT 3rd. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: A comparative review of 22 articles. Cancer 2000;88:1517-23.
3. Mankin HJ, Hornicek FJ, Springfield DS. Extra-abdominal desmoid tumors: A report of 234 cases. J Surg Oncol 2010;102:380-4.
4. Lewis JJ, Boland PJ, Leung DH, Woodruff JM, Brennan MF. The enigma of desmoid tumors. Ann Surg 1999;229:866-72.
5. De Cian F, Delay E, Rudigoz RC, Ranchère D, Rivoire M. Desmoid tumor arising in a cesarean section scar during pregnancy: Monitoring and management. Gynecol Oncol 1999;75:145-8.
6. Teo HE, Peh WC, Shek TW. Case 84: Desmoid tumor of the abdominal wall. Radiology 2005;236:81-4.
7. Waddell WR, Gerner RE. Indomethacin and ascorbate inhibit desmoid tumors. J Surg Oncol 1980;15:85-90.
8. Shestak KC, Edington HJ, Johnson RR. The separation of anatomic components technique for the reconstruction of massive midline abdominal wall defects: Anatomy, surgical technique, applications, and limitations revisited. Plast Reconstr Surg 2000;105:731-8.
9. Choi SH, Yoon HI, Kim SH, Kim SK, Shin KH, Suh CO. Optimal radiotherapy strategy for primary or recurrent fibromatosis and long-term results. PLoS One 2018;13:e0198134.