Case Report

Desmoid Fibromatosis of the Lower Abdominal Wall in Irrua Nigeria

Oluwafemi Olasupo Awe[1,2], Sylvester Eluehike[3]

Departments of [1]Surgery and [3]Radiology, Irrua Specialist Teaching Hospital, [2]Department of Surgery, Faculty of Clinical Sciences, Ambrose Alli University, Ekpoma, Edo State, Nigeria

Abstract

Desmoid fibromatosis (desmoid tumors) is rare tumors. It can occur as intra-abdominal, extraabdominal, or abdominal wall tumor depending on the site. The abdominal wall type is usually sporadic, but few have been associated with familial adenomatous polyposis. They are commonly seen in young females who are pregnant with a history of the previous cesarean section scar or within the 1st year of the last childbirth. There is an association between this tumor, presence of estrogen receptors, and abdominal trauma. We present a 29-year-old Nigerian woman with fungating lower abdominal wall tumor. This tumor is rare, a high index of suspicion will be very important in making the diagnosis.

Keywords: *Abdominal wall, desmoid fibromatosis, Nigerian woman, young*

Introduction

Desmoid tumor, also referred to desmoid fibromatosis, is an uncommon neoplasm.[1] The abdominal wall fibromatosis is a subtype of the desmoid fibromatosis, which constitutes about 3% of all soft-tissue neoplasm. Desmoid tumors are two main types: the familial and the sporadic types. Each of these is subclassified based on the location as intra-abdominal, extraabdominal, and abdominal wall desmoid tumors. The abdominal wall desmoid tumors are typically seen in young females who are pregnant or had the last childbirth within the last 1 year.[2] These tumors are often a temporal association with a history of abdominal trauma or previous cesarean section. These associations are correlated with the detection of increase estrogen receptors in the substance of the tumor.[3-5] The typical presentation is that of a painless enlarging mass which sometimes is ulcerated if the patient presented late. Despite their benign histopathological features, they are diffusely infiltrative. The treatment is usually complete resection with tumor-free margin.[6] The local recurrence even with tumor-free margin still approaches 40% in some series. Some researchers have documented the genetic predispositions to recurrence such as mRNA (messenger Ribonucleic acid), APC (adenomatous polyposis coli) mutation 3' of codon 1444, especially in patients with Familial adenomatous polyposis (FAP).[7] The use of chemotherapy, radiotherapy, and hormone therapy is controversial and limited to the unresectable tumors.[8]

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Case Report

A 29-year-old female patient who presented with 8 months rapidly growing and initially painless tumor on the lower abdomen. It started as a small swelling at the edge of the previous cesarean section scar 6 months after the surgery. It became ulcerated 4 months before the presentation after applying the herbal medications. There was associated contact bleeding, especially during dressing [Figure 1]. There was associated history of weight loss but no jaundice, loss of appetite, change in bowel habit, or symptoms suggestive of tuberculosis. There was no history of similar tumor, bleeding per rectum, or cancer of the bowel in first-degree relatives.

On examination, the patient was a healthy looking young woman, apprehensive with ulcerated, nontender and firm mass on the lower anterior abdominal wall. The mass was extending from the suprapubic area just above the pubic bone to few centimeters below the umbilicus, measuring 19 cm by 12 cm with bleeding ulcer in the central aspect. The ulcer had raised edge and unhealthy granulation tissue and necrotic floor. It measures about 15 cm by 8 cm [Figure 1]. There were no ascites and organomegaly. Bowel sounds were normoactive. The surrounding skin was normal with prominent veins.

Address for correspondence: Dr. Oluwafemi Olasupo Awe, Irrua Specialist Teaching Hospital, Km 86 Benin-Abuja Expressway, PMB 008, Irrua, Edo State, Nigeria.
E-mail: olasupoawe06@yahoo.com

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She had chest X-ray and abdominopelvic ultrasound scan done which shows no features of chest and abdominal metastases, and a mass on the anterior abdominal wall without any intraperitoneal extension [Figure 2]. The initial clinical diagnosis was ulcerated soft-tissue sarcoma (dermatofibrosarcoma).

Figure 1: Fungating mass

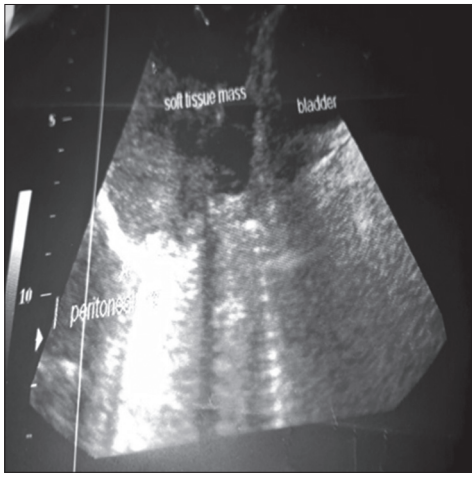

Figure 2: The abdominal ultrasound scan

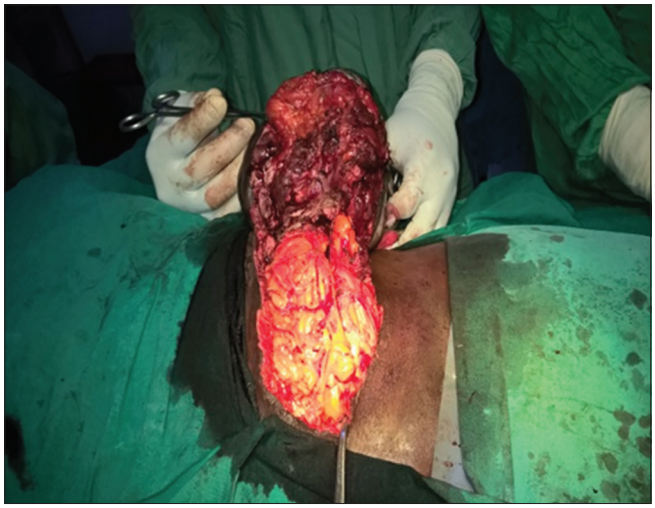

Figure 3: Excision of the mass

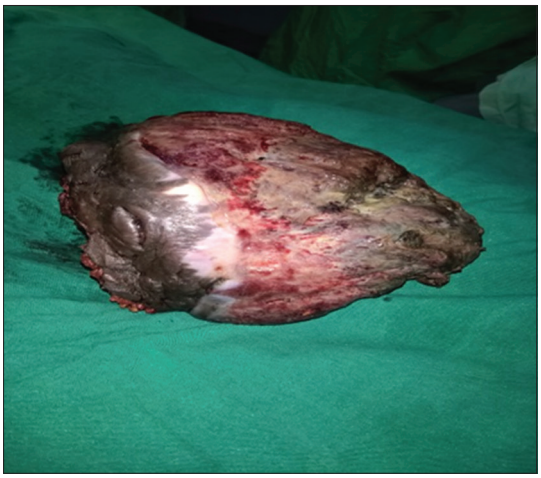

Figure 4: The excised mass (1.2 kg)

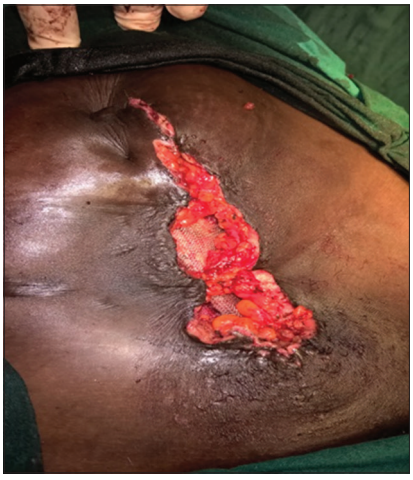

Figure 5: Mesh repair

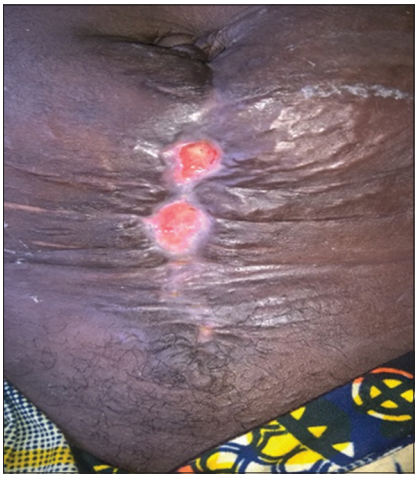

Figure 6: Postoperative day 20

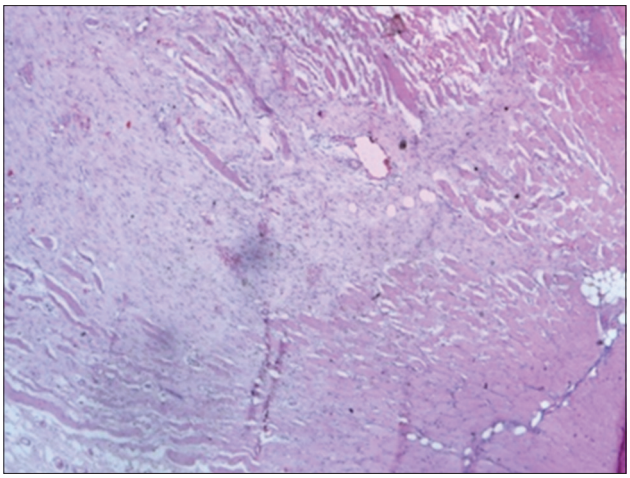

Figure 7: Photomicrograph ×40

The mass was adherent to the anterior rectus sheath down to the parietal peritoneum on the posterior surface on the central aspect. The mass was completely resected with 2 cm tumor-free margin macroscopically. The resultant defect was repaired with mesh, attaching it to the remnant rectus sheath. The excised tissue weighed 1.2 kg [Figures 3–5]. There was a postoperative wound infection which was managed with oral antibiotics and wound healed after 5 weeks. She was discharged home but lost to follow-up in the clinic [Figure 6].

The histopathology revealed a section of the anterior abdominal wall mass which shows a lesion composed of proliferating fibroblast and myofibroblast disposed within a myxocollagenous connective tissue stroma. Areas of congested thin to thick wall vessels, proliferating skeletal muscles, perivascular, and intercollagenous infiltrated with mainly lymphocytes with unremarkable overlying skin epithelium and dermis. The resection margin was free of infiltration. It was estrogen-receptor positive. There was no evidence of malignancy. The diagnosis of abdominal fibromatosis was made [Figure 7].

Discussion

Desmoid tumors are benign, deep fibromatosis originating from the fascia and muscle aponeurosis with an infiltrating growth. It could actually affect any part of the body.[3] It could be the aggressive or the slow-growing types. The aggressive type is usually associated with FAP. The slow-growing type is usually sporadic and not associated with FAP.[8] The slow-growing ones occur commonly in the shoulder girdle or the abdominal wall. The abdominal wall desmoid fibromatosis, like the one found in our patient, is more common in young females either during pregnancy or within the 1st year of childbirth.[4,5]

It is commonly associated with surgical trauma, usually a previous cesarean section. This tumor in the index patient was noticed about 6 months after cesarean section along the scar. Due to the late presentation and application of herbal medication, the mass became ulcerated. Most of the patient reported in the literature; the tumors were not ulcerated which contribute to initial misdiagnosis of ulcerated soft-tissue sarcoma or dermatofibrosarcoma.

The mass was fungating, so there was a need to eliminate the possibility of metastases, which will have required a computerized tomography scan or magnetic resonance imaging (MRI) scan of the abdomen and chest. This was not done due to financial constraints. We did a chest X-ray and abdominopelvic ultrasound scan instead. MRI could have assisted in demonstrating the homogenous and intense appearance of muscles on T1 weighted and heterogenous signal in intensity slightly lower than that of fat on T2-weighted imaging. Histopathology is the main stay of diagnosis.[9,10]

This type of tumor has been reported in a pregnant woman who had the previous cesarean section.[4,11,12]

Surgical resection of these tumors has been the main stay of the management. Complete resection with tumor-free margin of about 2 cm is still associated with local recurrence of 20%–40%.[3,13]

It has been shown that the admission status, gender, tumor size, margin status, location, number, and genetic mutations, such as mRNA, APC gene, are predicting factors of local recurrence. The most important of these factors for recurrence was tumor size >5 cm[14] and genetic mutations especially in patients with FAP.[15]

The size of the tumor at surgery will determine the extent of the defect and subsequent need for the mesh reconstruction of the abdominal wall. The mesh reconstruction was done in our patient but was complicated with wound infection, which may be as a result of the presence of ulcer or the mesh repair.[15]

Many surgeons have tried other modalities depending on the clinical grading of the tumor. Small asymptomatic lesions have been managed by either watchful waiting, surgery, or hormonal therapy. Large resectable tumors have surgery with or without adjuvant radiotherapy or chemotherapy. Unresectable tumors have been managed with many modalities. These include chemotherapy, radiotherapy, and hormone therapy. Chemotherapy drugs used including doxorubicin, dactinomycin, and carboplatin. Hormone therapy such as tamoxifen has been used with partial response in some cases.[16,17]

Adjuvant radiotherapy has also been reported after the complete resection to reduce the recurrence. A recurrence free period of 6 years has also been shown with this treatment.[18]

In our case, adjuvant chemotherapy was considered after about 3 months postresection, but only complete resection has been done because she could not afford the radiotherapy. Complete regression has been noticed in one series with tamoxifen.[19]

Conclusion

The abdominal wall desmoid fibromatosis should be considered in a young female patient presenting with abdominal wall tumor, especially during pregnancy or within the 1st year of childbirth with the previous history of abdominal trauma or surgery (cesarean section) scar. The surgery has been the main stay of treatment, but hormone therapy has resulted in the tumor regression, only in estrogen-receptor positive tumors, which could avoid mutilating surgery. Adjuvant radiotherapy could also reduce the incidence of local recurrence after complete resection. Mesh reconstruction helps in maintaining the integrity of the abdominal wall following surgery in large tumor.

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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Park HC, Pyo HR, Shin KH, Suh CO. Radiation treatment for aggressive fibromatosis: Findings from observed patterns of local failure. Oncology 2003;64:346-52.
2. Singer S. Soft tissue sarcoma. In: Townsend C, Beauchamp RD, Evers BM, Mattox K, editors. Sabiston Textbook of Surgery: The Biological Basis of Modern Surgical Practice. 19th ed. Philadelphia, USA: Elsevier Saunders; 2015. p. 768-82.
3. Jain D, Lubane P, Pancholi M. Large desmoid tumour of the anterior abdominal wall: A case report of a 4.6kg desmoid tumour. Internet J Surg 2006;10:1-5.
4. Michopoulou A, Germanos S, Kanakopoulos D, Milonas A, Orfanos N, Spyratou C, *et al.* Management of a large abdominal wall desmoid tumor during pregnancy. Case report. Ann Ital Chir 2010;81:153-6.
5. Galeotti F, Facci E, Bianchini E. Desmoid tumour involving the abdominal rectus muscle: Report of a case. Hernia 2006;10:278-81.
6. Teo HE, Peh WC, Shek TW. Case 84: Desmoid tumor of the abdominal wall. Radiology 2005;236:81-4.
7. Bright-Thomas RM, Agrawal A, Hargest R. Preclinical studies of gene transfer for the treatment of desmoid disease in familial adenomatous polyposis. Br J Surg 2002;89:1563-9.
8. Kanyamuhunga N, McCall N, Tuyisenge L, Mumena C, Stefan DC. Aggressive desmoid fibromatosis: A first case in Rwandan child. S Afr J Child Health 2013;7:117-8.
9. Casillas J, Sais GJ, Greve JL, Iparraguirre MC, Morillo G. Imaging of intra-and extra-abdominal desmoid tumors. Radiographics 1991;11:959-68.
10. Pratap A, Tiwari A, Agarwal B, Pandey SR, Paudel G, Kumar A, *et al.* Inflammatory myofibroblastic tumor of the abdominal wall simulating rhabdomyosarcoma: Report of a case. Surg Today 2007;37:352-5.
11. Viriyaroj V, Yingsakmongkol N, Pasukdee P, Rermluk N. A large abdominal desmoid tumor associated with pregnancy. J Med Assoc Thai 2009;92 Suppl 3:S72-5.
12. Krentel H, Tchartchian G, De Wilde RL. Desmoid tumor of the anterior abdominal wall in female patients: Comparison with endometriosis. Case Rep Med 2012;2012:725498.
13. Huang K, Fu H, Shi YQ, Zhou Y, Du CY. Prognostic factors for extra-abdominal and abdominal wall desmoids: A 20-year experience at a single institution. J Surg Oncol 2009;100:563-9.
14. Kumar V, Khanna S, Khanna AK, Khanna R. Desmoid tumors: Experience of 32 cases and review of the literature. Indian J Cancer 2009;46:34-9.
15. Adigun IA, Olabanji JK, Oladele AO, Lawal OO, Alabi GH. Early experience with the use of prosthetic mesh as fascia replacement in structural abdominal reconstruction. Ind J Plast Surg 2007;40:164.
16. Wilcken N, Tattersall MH. Endocrine therapy for desmoid tumors. Cancer 1991;68:1384-8.
17. Patel SR, Evans HL, Benjamin RS. Combination chemotherapy in adult desmoid tumors. Cancer 1993;72:3244-7.
18. Chao AS, Lai CH, Hsueh S, Chen CS, Yang YC, Soong YK, *et al.* Successful treatment of recurrent pelvic desmoid tumour with tamoxifen: Case report. Hum Reprod 2000;15:311-3.
19. Seoud M, Abbas J, Kaspar H, Khalil A, Geara F. Long-term survival following aggressive surgery and radiotherapy for pelvic fibromatosis. Int J Gynecol Cancer 2005;15:1112-4.