Case Report

Surgery Section

Desmoid Tumor of the Abdominal Wall: Report of a Case

SALMAN YOUSUF GURAYA, KHALID RIDA MURSHID

ABSTRACT

Desmoids tumours of the abdomen are uncommon benign tumours with varied presentations. The diagnosis is frequently established by histological examination. These tumours are known to recur even after surgical excision. A case report of a 41-year-old female with a painless abdominal mass has been presented. She was found to have a firm, non-tender, immobile, and regular mass in the left lumbar region of the abdominal wall. Her CT scan and FNAC revealed a benign-natured lesion. The patient's mass was completely excised and the histology confirmed desmoids tumour of the abdominal wall. The rarity of such lesions warrants that the treating physicians should have a high degree of suspicion while managing abdominal masses of a questionable nature.

Key Words: Surgery, Abdomen, Growth

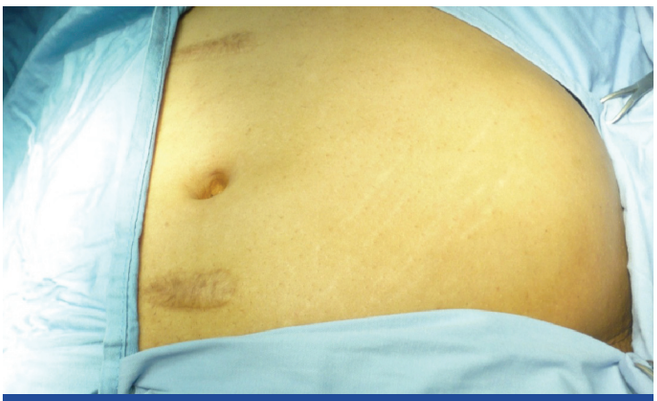
INTRODUCTION

Desmoids tumour, also known as aggressive fibromatosis, is a rare soft tissue neoplasm which is caused by a monoclonal proliferation which arises in musculoaponeurotic structures [1]. Although fibromatosis is thought to be a benign lesion that lacks metastatic potential, it can be characterized by a locally aggressive and infiltrating growth pattern with a high propensity towards local recurrence [2, 3]. Histologically, the tumour consists of a bland-appearing proliferation of spindle cells. This uncommon disease accounts for less than 3% of the soft tissue tumours, or 0.03% of all the neoplasms, and it is more common in women. The peak incidence is in the age range of 25-35 years [4], and most of the cases occur between puberty and 40 years of age [5].

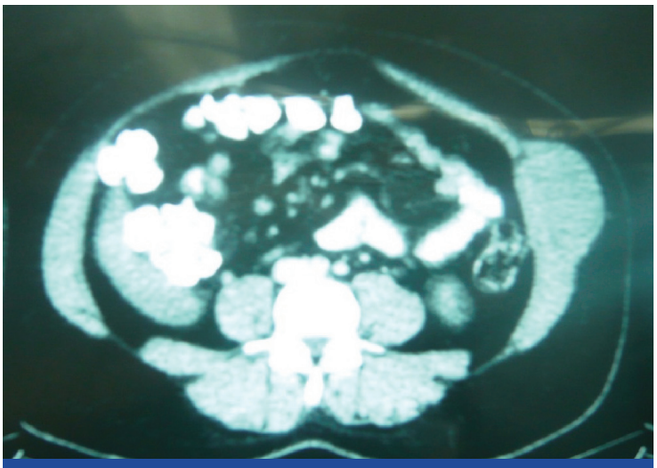
Abdominal desmoids tumours occur sporadically or in association with rare familial syndromes such as familial adenomatous polyposis coli [6]. This case report presents a rare case of desmoids tumour of the abdominal wall in a young female.

CASE REPORT

A 41-year-old female presented to the surgical clinic of Ohud Hospital, Al Madina Al Munawara Kingdom of Saudi Arabia, with the complaint of a painless left sided abdominal mass of 6 months duration. The patient denied any systemic illness, familial disease or trauma in the region. She was found to have normal vital signs. On examination of her abdomen, a firm, globular, non-mobile, non-tender mass was found in the left lumber region, which measured 8 × 8 cm [Table/Fig-1]. All the baseline blood results were normal. Her CT scan revealed a homogenous, benign looking mass in the left lumbar region with local infiltrations to the surrounding abdominal wall muscles [Table/Fig-2]. The mass was reported to be superficial to the peritoneum, lying among the layers of the abdominal wall muscles. The FNAC of the mass showed few spindle cells but they remained essentially equivocal. The patient was subjected to exploration of the mass under general anaesthesia through a left oblique incision. The mass was found to infiltrate the peritoneum which was opened for a clear resection of

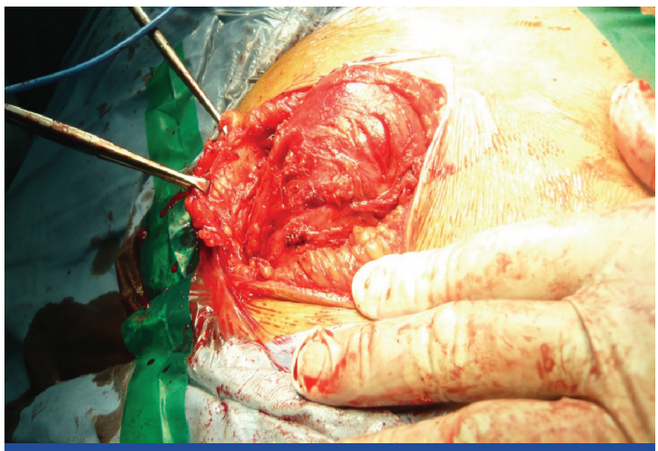


[Table/Fig-1]: Left sided abdominal mass

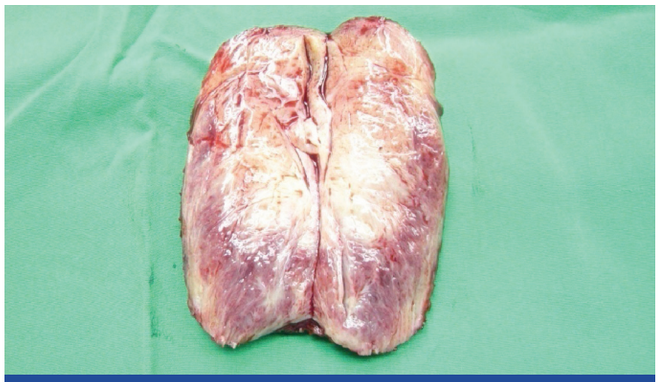


[Table/Fig-2]: Contrast-enhanced CT scan of the abdomen showing left sided soft tissue homogenous mass attached to the abdominal wall musculature

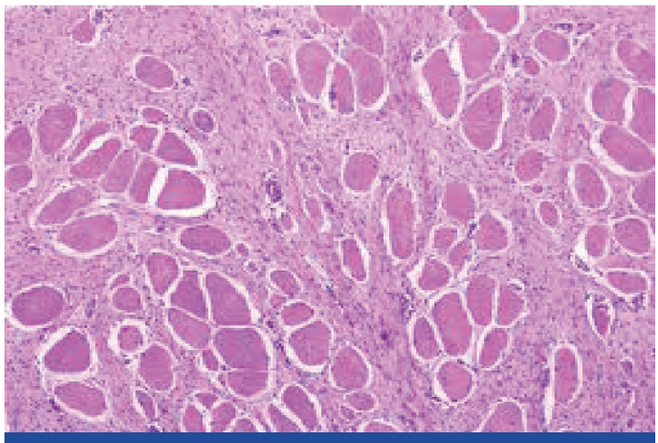
the lesion [Table/Fig-3]. The tumour was completely excised with a safety margin of 2 cm all around [Table/Fig-4]. The abdominal wall defect was approximated with a layered closure after placing a drain. The histological 9 report confirmed a desmoids tumour which was composed of scattered spindle cells and fibroblasts



[Table/Fig-3]: Preoperative view of the abdominal wall mass



[Table/Fig-4]: The cut-open view of the excised specimen



[Table/Fig-5]: 625×450 HPF showing fibroblasts and spindle cells among a background of mesenchymal cells

among few skeletal and mesenchymal cells [Table/Fig-5]. There was no evidence of malignant transformation in the lesion. The patient made an uneventful recovery and remained well during a 1-year follow up.

DISCUSSION

Due to the rarity of desmoids tumours, randomized controlled trials are lacking, but in reported retrospective studies, it is evident that although desmoids tumours are considered to be benign, local recurrence is not uncommon [7]. Local recurrence rates of 25-65% have been reported [4]. These tumours have been reported at various sites including the abdominal wall and the breast [8]. Despite attaining huge sizes, desmoids tumours remain asymptomatic. The diagnosis is generally established by a combination of clinical examination, imaging by CT and MRI, and Fine Needle Aspiration and Cytology (FNAC). There is no striking radiological feature of the desmoids tumours which can easily clinch the diagnosis.

Regardless of the site, an aggressive wide local excision of the desmoid tumours has been popularly advocated in the literature [9, 10]. The factors that support this aggressive surgical approach, include the potential of fibromatosis in displaying a locally infiltrating growth pattern into the surrounding structures, the resultant high propensity for local recurrence when the tumours are incompletely excised, and the lack of convincing evidence for a proven beneficial role of radiation therapy, chemotherapy, or a combination of therapies [11, 12]. However, other authors have been less enthusiastic and have given guarded support to an aggressive surgical approach [13, 14]. The factors that these authors stress are the increased potential for a less than optimal cosmetic outcome, the risk of loss of function, rare reported cases of spontaneous regression of fibromatosis, and a high incidence of recurrence after an apparent complete excision with negative surgical margins.

In a small series of seven cases, one patient developed recurrent disease that was surgically resected and the patient remained disease-free during a 6-year follow-up [15]. Also, in another study of 23 cases, the local recurrence rate of 13% was recorded after a median follow-up of 30 months [16]. In a small series of four patients with desmoids tumours of the abdominal wall, who underwent surgical excision after neoadjuvant therapy with preoperative doxorubicin and radiotherapy, no long-term recurrences were reported after a median follow-up of 6 years [17]. Most of the recurrences are observed within 3 years, and nearly all were observed by 6 years [18]. Age may affect the recurrence rate; although the information is unclear, it may be more likely in younger patients with extra-abdominal desmoids tumours [18]. Bertani et al [7] have stressed that a disease-free resection margin was the most crucial factor for reducing the recurrence rate. Moreover, a one-stage treatment with immediate mesh reconstruction was cost-effective, and it increased the chances of a definite cure. This concluded that the mesh placement has to be performed in the presence of microscopically negative margins, and for this reason, they recommended routine frozen sections of the specimen. Reconstruction of the large abdominal wall defects is complicated and demanding [19]. Abdominal wall reconstruction can be achieved by direct repair by suturing, and by the use of synthetic material (mesh) or myocutaneous flaps, in case of extensive defects. In addition to the surgical option, there are claims which support the efficacy of radiation therapy. However, it is rarely used for abdominal wall and intra-abdominal disease because of the potential risk of radiation enteritis [20].

To conclude, abdominal wall desmoids tumours are rare and they necessitate wide surgical excision for a definite cure. A close follow-up is warranted due to the high recurrence rate.

REFERENCES

[1] Li M, Cordon-Cardo C, Gerald WL, Rosai J. Desmoid fibromatosis is a colonal process. *Hum Pathol.* 1996; 27: 939-43.
[2] Enzinger FM, Weiss SW. Fibromatosis. In soft tissue tumours 4th edition. Edited by: Enzinger FM, Weiss SW. St Louis, MO, *Mosby*; 2001; 320-29.
[3] Wargotz ES, Norris HJ, Austin RM, Enzinger FM. Fibromatosis of the breast. A clinical and pathological study of 28 cases. *Am J Surg Pathol* 1987; 11: 38-45.
[4] Enzinger FM, Weiss SW. Soft tissue tumours. 3rd ed. St Louis: *Mosby*; 1995; 201-29.

[5] Lefevre JH, Parc Y, Kerneis S, et al. Risk factors for the development of desmoids tumours in familial adenomatous polyposis. *Br J Surg* 2008; 95: 1136-39.
[6] Lips DJ, Barker N, Clevers H, Hennipman A. The role of APC and beta-catenin in the aetiology of aggressive fibromatosis (desmoids tumours). *Eur J Surg Oncol* 2009; 35: 3-10.
[7] Bertani E, Chiappa A, Testori A, et al. Desmoid tumours of the anterior abdominal wall: results from a monocentric surgical experience and review of the literature. *Ann Surg Oncol* 2009; 16: 1642-49.
[8] Povoski S, Marsh Jr W, Spigos D, Abbas A, Buchele B. Management of a patient with multiple recurrences of fibromatosis (desmoids tumour) of the breast involving the chest wall musculature. *World Journal of Surgical Oncology* 2006; 4(32): 1-8.
[9] Pearson GD, Vasko SD. Chest wall reconstruction after desmoids tumour resection. *Surg Rounds* 2005; 28: 385-88.
[10] Devouassoux-Shisheboran M, Schammel DP, Man YG, Tavassoli FA. Fibromatosis of the breast. Age-correlated morphofunctional features of 33 cases. *Arch Pathol Lab Med* 2000; 124: 276-80.
[11] Matherne TH, Green A, Tucker JA, Dyess DL. Fibromatosis: the breast cancer imitator. *South Med J* 2004; 97: 1100-03.
[12] Schwarz GS, Drotman M, Rosenblatt R, Milner L, Shamonki J, Osborne MP. Fibromatosis of the breast: case report and current concepts in the management of an uncommon lesion. *Breast J* 2006; 12: 66-71.
[13] Godwin Y, McCulloch TA, Sully L. Extra-abdominal desmoid tumour of the breast: review of the primary management and implications for breast reconstruction. *Br J Plast Surg* 2001; 54: 268-71.
[14] Gump FE, Sternschein MJ, Wolff M. Fibromatosis of the breast. *Surg Gynecol Obstet* 1981; 153: 57-60.
[15] Sutton RJ, Thomas JM. Desmoid tumours of the anterior abdominal wall. *Eur J Surg Oncol* 1999; 25: 398-400.
[16] Phillips SR, Hern RA, Thomas JM. Aggressive fibromatosis of the abdominal wall, limbs and girdles. *Br J Surg* 2004; 91: 1624-29.
[17] Baliski CR, Temple WJ, Arthur K, Schachar NS. Desmoid tumours: a novel approach for local control. *J Surg Oncol* 2002; 80: 96-99.
[18] Reitamo JJ, Scheinin TM, Hayry P. The desmoid syndrome: new aspects in the cause, pathogenesis and treatment of the desmoid tumour. *Am J Surg* 1986; 151: 230-37.
[19] Rorhich RJ, Lowe JB, Hackney FL, Bowman JL, Hobar PC. An algorithm for abdominal wall reconstruction. *Plast Reconstr Surg* 2000; 105: 202-16.
[20] Guadagnolo BA, Zagars GK, Ballo MT. Long-term outcomes for desmoids tumours which were treated with radiation therapy. *Int J Radiat Oncol Biol Phys* 2008; 71: 441-47.

AUTHOR(S):
1. Dr. Salman Yousuf Guraya
2. Dr. Khalid Rida Murshid

PARTICULARS OF CONTRIBUTORS:
1. Corresponding Author.
2. FRCS(C), Associate professor of Surgery and Consultant Surgeon, College of Medicine, Taibah University Al Madina Al Munawara, Saudi Arabia.

NAME, ADDRESS, TELEPHONE, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Salman Yousuf Guraya FRCS
Professor of Surgery and Consultant Surgeon
College of Medicine, Taibah University
Al Madina Al Munawara Saudi Arabia
E-mail: syousuf@taibahu.edu.sa

DECLARATION ON COMPETING INTERESTS:
No competing Interests.

Date of Submission: **Jul 07, 2011**
Date of peer review: **Sep 27, 2011**
Date of acceptance: **Oct 04, 2011**
Date of Publishing: **Dec 25, 2011**