

## Case Report

# Management of the patient with fibromatosis of the left anterior chest wall

Saravanakumar Subbaraj, Sandeep Manohar\*

Department of General Surgery, Mahatma Gandhi Medical College and Research Institute, Pondicherry, India

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**\*Correspondence:**

Dr. Sandeep Manohar,

E-mail: [sm69154@gmail.com](mailto:sm69154@gmail.com)

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### ABSTRACT

Desmoid fibromatosis is a rare but locally aggressive tumour comprised of myofibroblasts. It is histologically benign but can behave aggressively. They do not have the ability to metastasize but can cause significant morbidity and mortality by local invasion. These tumours may occur anywhere on the body, but are commonly found on the abdominal wall and within the intestinal mesentery. Mutations in either the  $\beta$ -catenin or the adenomatous polyposis coli (APC) genes are usually the cause for the development of desmoid tumours with the former comprising the sporadic development of tumours and the latter being associated with familial adenomatous polyposis syndrome. Surgical resection with histologically negative margins has been the cornerstone of therapy for this disease, but this paradigm has begun to shift. It is now common to accept a microscopically positive margin after resection as recurrence rates may not be significantly affected. This case report intends to describe the clinical, diagnostic and pathologic features of a post-traumatic fibromatosis involving left side chest wall in a 45 years old female and causing worsening pain. The surgical management was successfully undertaken.

**Keywords:** Desmoid tumour, Fibromatosis, Post traumatic, Surgical resection

### INTRODUCTION

Term fibromatosis refers to group of benign soft tissue tumour, which have certain characteristics in common, including absence of cytological and clinical malignant features, histology consistent with proliferation of well-differentiated fibroblasts, an infiltrative growth pattern and an aggressive clinical behavior with frequent local recurrence. Other names include musculo-aponeurotic fibromatosis and desmoid tumour.<sup>1</sup>

Desmoid tumour is a rare and aggressive soft tissue tumour with unique biologic behaviour of recurrence even after complete resection. Three main anatomic sites are described: trunk or extremity, abdominal wall, and intra-abdominal (bowel and mesentery). They are more common in women and occur commonly in the fourth

decade of life. It can be associated with familial adenomatous polyposis.<sup>1</sup> Intra-abdominal and extremity desmoid represent extra-colonic manifestations of disease in Gardner syndrome. The origin of the tumour was poorly understood and warrants specific treatment strategies based on anatomical sites.<sup>2</sup> Although surgery is the primary treatment modality, there is poor consensus amongst surgeons in the management of desmoid tumour. Here we report a case of a female with fibromatosis of the left inferior chest wall.

### CASE REPORT

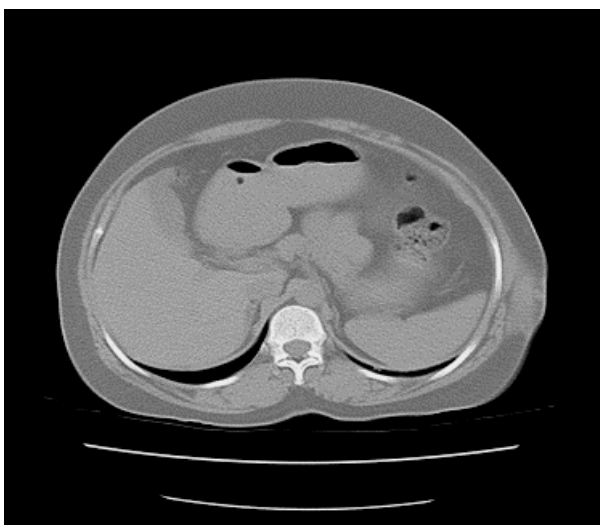
We encountered a 45 years old married female, who presented to our hospital surgery OPD with worsening pain of the left chest wall region with obvious mass in the left costal region for one and half years. Two years prior to

her presentation she had a blunt trauma over the same region.

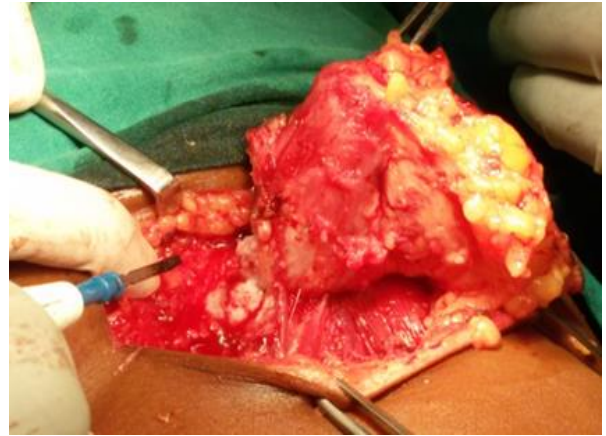
Physical examination showed a 5 cm swelling in left anterior chest wall, which was hard in consistency. The mass was partly mobile and tethered to the skin (Figure 1). Ultrasonography (USG) shows a well-defined hypoechoic lesion measuring 3.6×1.4 cm seen arising from the left side of the chest wall in the intermuscular plane which extends into subcutaneous plane. X-ray ribs excluded bone involvement. Doppler showed no evidence of increased vascularity. Computed tomography (CT)-thorax (Figure 2) showed, a well-defined hyper dense lesion measuring 4.2×2.4 cm seen in the left lateral aspect of the thorax in the intermuscular plane. The T value of the lesion varies from 25 to 50 HU (Hounsfield units). The rib was separate from the lesion, lung field appears normal and mediastinal structure appears normal. As diagnosis was established, surgery was scheduled under general anaesthesia. She underwent an en bloc resection of the tumour along with infiltrated surrounding intercostals muscles fibres (Figure 3 and 4).



**Figure 1: Fibromatosis of left chest wall with skin tethering.**



**Figure 2: CT picture of the tumour.**



**Figure 3: Plane of the tumor is predominantly subcutaneous with infiltration of the adjacent skeletal muscle.**

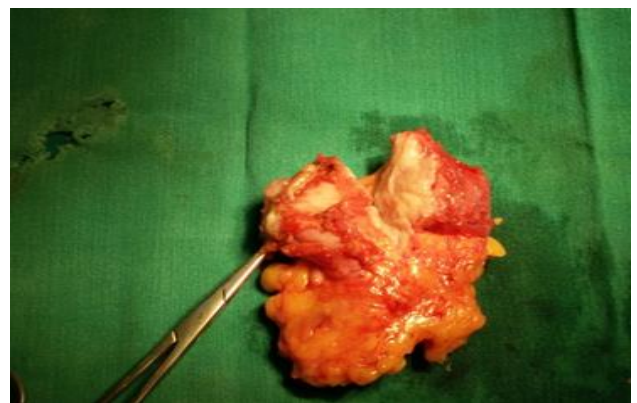


**Figure 4: After en bloc resection of the tumour.**

#### **Histopathology report**

##### *Gross*

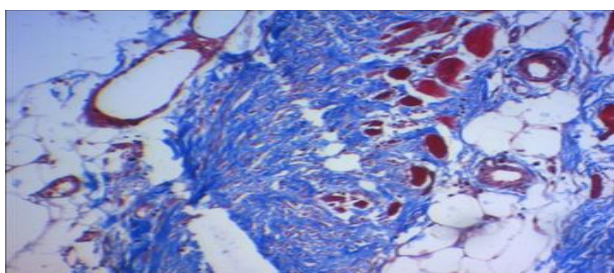
Cut section shows a grey white nodule measuring 4×3 cm with an island of skin surrounding fatty strands and few muscle fibres. Skin appears normal (Figure 5).



**Figure 5: Gross picture of the tumour.**

### Microscopic

Section shows bland looking elongated spindle cells resembling fibroblast and fibrocytes arranged in poorly circumscribed bundles. No evidence of mitotic activity or cytological features of malignancy seen. There is an infiltrative pattern seen in the periphery involving fat and entrapped skeletal muscle bundles. The vessels in the periphery shows perivascular lymphocytic infiltrate which is suggestive of fibromatosis (Figure 6).



**Figure 6: Fibrocytes arranged in poorly circumscribed bundles.**

The post-operative period was uneventful and she was discharged to home after 10 days. She is currently under follow-up.

### DISCUSSION

Desmoid tumours are soft tissue neoplasms arising from fascial or musculoaponeurotic structures, they consist of proliferations of benign-appearing fibroblastic cells, abundant collagen, and few mitoses. Desmoid tumours possess alterations in the adenomatous polyposis coli (APC)/ $\beta$ -catenin pathway. Cyclin D1 dysregulation is thought to play a significant role in their pathogenesis.<sup>7</sup> Associations with other diseases and conditions are well documented, especially those with similar alterations in APC pathway, such as familial adenomatous polyposis (Gardner's syndrome). Other conditions with increased risk of desmoid tumour formation include increased estrogen states (pregnancy) and trauma. Surgical incisions (abdominal and thorax) have been a site of desmoid development, either in or near the scar. Clinically, patients are usually in the third to fourth decade of life and have pain, a chest wall mass, or both. The tumour can sometimes be fixed to the chest wall, and skin involvement may not be present. Radiologically, magnetic resonance imaging (MRI) can be useful to delineate muscle or soft tissue infiltration. Desmoid tumours do not metastasize, but have the ability to recur locally, with recurrence rates ranging between 5-50%. This locally aggressive behaviour is secondary to microscopic tumour infiltration of muscle and surrounding soft tissues and prompts some to consider them a low-grade form of fibrosarcoma.

Because these lesions have a low cellularity and poor yield with fine needle aspiration, an open incisional biopsy for lesions over 3 to 4 cm is necessary. The surgery of choice is a wide local excision. A margin of less than 1 cm results in much higher local recurrence rates. Survival after a wide local excision with negative margins is 90% at 10 years.<sup>8</sup>

### CONCLUSION

Desmoid tumours are a mesenchymal neoplasm. The suggested treatment strategy is surgical resection with safe margin. There is disagreement amongst surgeons with resection protocol due to high rate of recurrence due to the aggressive and local invasive nature of desmoid tumour. We followed a safe and conservative resection.

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### REFERENCES

1. Latchford AR, Sturt NJH, Neale K, Rogers PA, Phillips RKS. A 10-year review of surgery for desmoid disease associated with familial adenomatous polyposis. *Br J Surg.* 2006;93(10):1258-64.
2. Smith AJ, Lewis JJ, Merchant NB, Leung DHY, Woodruff JM, Brennan MF. Surgical management of intra-abdominal desmoid tumours. *Br J Surg.* 2000;87:608-13.
3. Enzinger FM, Shiraki M. Musculo-aponeurotic fibromatosis of the shoulder girdle (extra-abdominal desmoid): analysis of thirty cases followed up for ten or more years. *Cancer.* 1967;20:1131-40.
4. MacKinnon JG, Neifeld JP, Kay S, Parker GA, Foster WC, Lawrence Jr W. Management of desmoid tumors. *Surg Gynecol Obstet.* 1989;169(2):104-6.
5. Merchant NB, Lewis JJ, Woodruff JM, Leung DH, Brennan MF. Extremity and trunk desmoid tumors: a multifactorial analysis of outcome. *Cancer.* 1999;86:2045-52.
6. Allen PJ, Shriver CD: Desmoid tumors of the chest wall. *Semin Thorac Cardiovasc Surg.* 1999;11:264-9.
7. Andino L, Cagle PT, Murer B, Lu L, Popper HH, Salle FG, Sienko AE, Barrios R, Zander DS. Pleuropulmonary desmoid tumors: immunohistochemical comparison with solitary fibrous tumors and assessment of beta-catenin and cyclin D1 expression. *Arch Pathol Lab Med.* 2006;130(10):1503-9.
8. Baliski CR, Temple WJ, Arthur K, Schachar NS. Desmoid tumors: a novel approach for local control. *J Surg Oncol.* 2002;80(2):96-9.

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