International Journal of Surgery Science

E-ISSN: 2616-3470
P-ISSN: 2616-3462

https://www.surgeryscience.com
2024; 8(1): 39-43
Received: 05-10-2023
Accepted: 06-11-2023

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Fibro sarcomatous conversion of a recurrent Dermatofibrosarcoma protuberans with sternal involvement

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DOI: https://doi.org/10.33545/surgery.2024.v8.i1a.1047

Abstract
A fibrosarcomatous-dermatofibrosarcoma protuberans is a locally aggressive tumor necessitating en-bloc excision with 2-3cm negative margins. Inadequate resection is an independent factor determining recurrence. Here we present a case of FS-DSP involving the sternum. The location proved to be a challenge in achieving adequate margins. Emphasis on the importance of a multi-speciality approach that enabled en-bloc tumor excision with negative margins and an immediate functional and cosmetic reconstruction. Our goal was obtaining optimal oncological results to improve the quality of life of the patient, the overall survival and achieve suitable cosmesis.

Keywords: Dermatofibrosarcoma protuberans, Fibrosarcomatous- Dermatofibrosarcoma protuberans, sternal reconstruction, omental flap

Introduction

Case Report

71-year-old male, presented with an exophytic bleeding mass on the sternum. He is a known case of dermatofibrosarcoma (DFS) of the anterior chest wall diagnosed two years back. Then, a wide local excision of the DFS and reconstruction using a local rotational flap was done at another institute. This flap underwent necrosis requiring multiple debridements and negative pressure therapy adding to the morbidity. The defect was eventually skin grafted. He developed a recurrent lesion which demonstrated fibrosarcomatous conversion of the DFS on core-needle biopsy. The tumor was a 7×7 cm fungating mass with a bleeding ulcer (fig.1). The patient was on dual-antiplatelet cover for his IHD, which amplified the bleeding to require multiple blood transfusions and pressure dressings. CT scan with angiography of the thorax revealed that the mass involved deeper osseocartilaginous structures with encasement of the right internal mammary artery (fig.2). Patient was optimized for surgery from cardiac and diabetic standpoint.

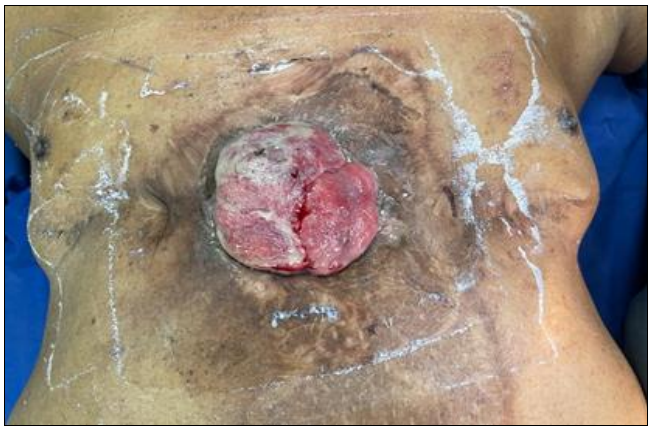

Fig 1: 7×7cm fungating tumor on the anterior chest wall

Fig 2: CT scan revealing the mass is adherent and involving underlying osseocartilaginous structures.

Surgery encompassed extended resection with reconstruction. An abdominal approach was adopted to enter virgin territory. An opening was made in the diaphragm to approach the thoracic cavity. Careful dissection was carried out in the substernal plane between the tumor and pericardium. En-bloc tumor excision included 4-7th costal cartilage and corresponding ribs partly on the left, and 3-7th costal cartilages and corresponding ribs on the right along with the body and xiphoid process of the sternum. Once the body of sternum was detached from the manubrium sterni, both internal mammary arteries were ligated close to its origin. This excision created a defect involving both thoracic and abdominal cavity with a midline bony defect and an open diaphragm (fig.3). Intercostal drains were placed in both pleural cavities and further reconstruction was done by the plastic surgery team.

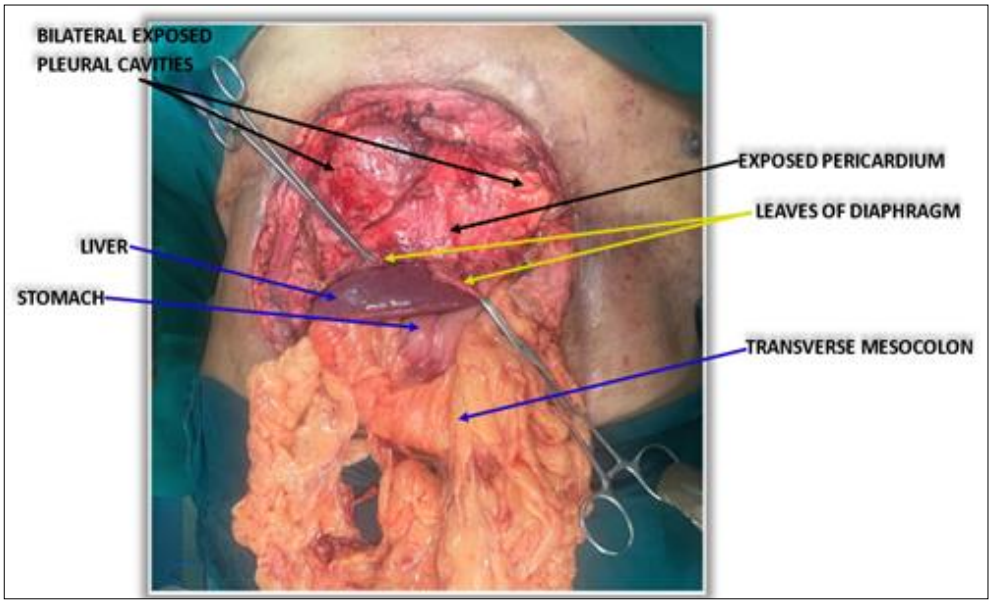

Fig 3A: The surgical defect on the right. The surgical defect encompasses exposed bilateral pleura and lungs, the pericardium, diaphragmatic leaves held by the artery forceps, the liver, stomach, transverse colon, and the omentum.

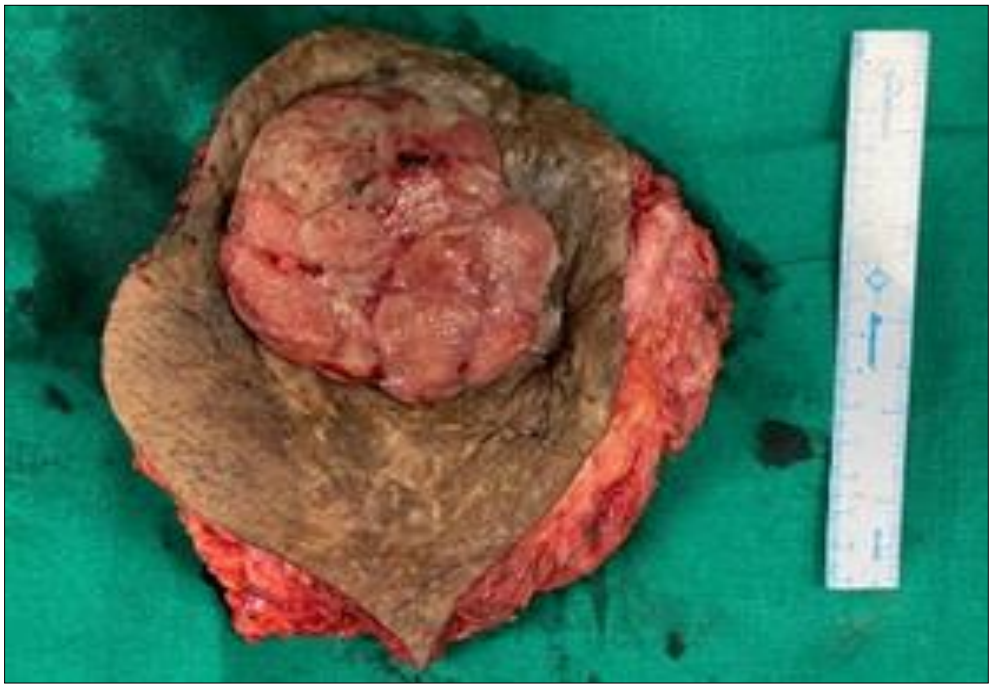

Fig 3B: The resected specimen on the left

The bony defect could have been reconstructed by a customised Titanium framework, approach that precluded the 6-week wait. Regardless of the wide defect, circumferential continuity of the thoracic cage was maintained as the manubrium was preserved along with its the attachments of the first three ribs. Skin of the anterior chest wall had been utilised during previous reconstruction. Since both internal mammary arteries had to be ligated, a TRAM flap was eliminated. In the background of multiple comorbidities, it was eminent to limit the time of surgery and hence latissimus dorsi flaps were avoided as it required repeated change of positions. Reconstruction with a fold-over omental flap and a dual mesh sandwiched between the two layers was opted, as the abdominal cavity was already open. Omentum was released from the transverse colon to gain length and turned over cephalically. The part attached to the colon was used to seal open pleural cavities and form a base for the mesh. A composite mesh was placed and anchored with its acellular layer facing the pleural and pericardial cavities. It was additionally hitched to the diaphragm to close its opening. This mesh along with subsequent fibrosis made a semi-rigid structure providing stability and adequate elasticity. The distal part of the omentum was now folded over the mesh, thus covering it, and preventing mesh exposure. Omentum was covered with an autologous split thickness skin graft and secured with negative pressure therapy (Fig.4).

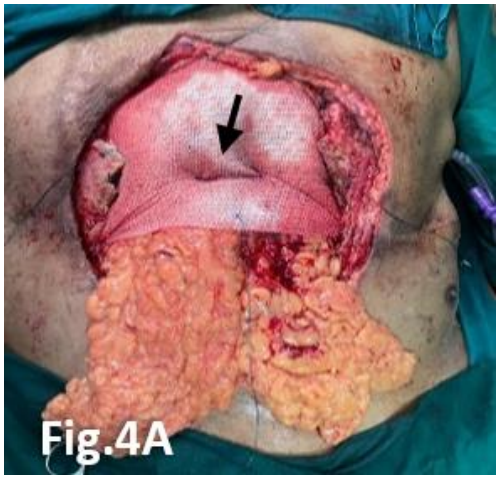

Fig 4A: One layer of omentum enforced with a composite mesh. Diaphragm hitched to the mesh – arrow.

Fig 4B: Self folding omental flap to cover the mesh.

Fig 4C: A split thickness skin graft

Fig 4D: Negative pressure therapy.

Post-operatively the patient was mechanically ventilated for 48 hours. After extubation, routine post-operative care given. Vacuum therapy was discontinued after 5 days once graft take was confirmed. Histopathology and immunohistochemistry confirmed a high-grade fibrosarcoma with negative cut margins on all sides. Patient was discharged once all parameters were met. On follow up, wound had healed satisfactorily (fig.5).

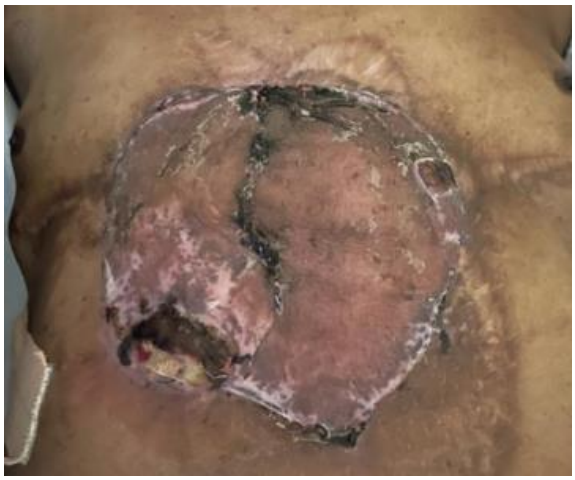

Fig 5: Post-operative cosmesis on 30-day follow-up.

Discussions

Sternal involvement by tumors is rare, with the most common

being chondrosarcoma (0.5-1% of all bone tumors). Overall, <1% cases of sternal involvement by malignancy are due to fibrosarcoma [1].

DFSP is a rare, fibrohistiocytic tumor of low-to-intermediate grade malignancy originating from dermal fibroblasts [2]. Incidence of DFSP is <0.1% of all malignancies and 1-6% of all soft tissue sarcomas [3]. They are common in the middle-age with an equal gender preponderance (3). They occur de novo but are also correlated with irradiated areas, scarred tissue, or tattooed skin [2, 7]. They are locally aggressive tumors with 20-50% recurrence rate (2) and 2-5% metastatic potential [4]. Achieving local control is key to treating DFSP. It has been observed that tumor spread due to insufficient previous surgery becomes more aggressive and invasive. Adjuvant radiotherapy is given for close or positive margins not amenable to resection. Imatinib has been proven to benefit in metastatic and inoperable disease [4].

Fibrosarcomatous areas in DFSP were first described by Penner in 1951 [7]. Now, Fibrosarcomatous-Dermatofibrosarcoma protuberans (FS-DFSP) is a known histological change documented in 5-15% of DFSP cases. Clinically there is no differentiating factor between the two. The risk of local recurrence (29.8% vs 13.7%), metastasis (14.4% vs 1.1%) and death (14.7% to 0.8%) are all higher in FS-DFSP as compared to DFSP alone [5]. The local recurrence rate has a strong correlation with surgical approach and defined surgical margins. In studies conducted by Goldlum *et al*. and Szollosi *et al*., recurrence was lowest (20%) in a group with margins ≥2cms as compared with no defined margins (38.8%) and highest (63.6%) where the surgical procedure was not specified [6, 7]. Tumor size does not correlate with prognosis or degree of recurrence [5]. Hence, en-bloc tumor excision with 2-3cm margin is of utmost importance. However, such extensive resections may lead to major defects with cosmetic and functional deficiencies mandating complex reconstruction. Reconstruction may be single or multiple-stage procedure to address lost components of the anatomy during excision. Difficulties in reconstruction though, should not influence the primary wide local resection, as cure holds precedence.

Reconstruction is paramount to establish cover for the underlying vital structures, maintain adequate cardiac and respiratory function, and prevent flail chest. Reconstruction of the chest wall encompasses soft tissue and bony components. Soft tissue defects are closed by local/distant skin flaps, myocutaneous flaps and/or omentum. Skeletal reconstruction by bone allografts/autografts, plates, titanium meshes or 3D-printed templates [9]. Use of locoregional/distant flaps are already described for reconstruction in post-CABG mediastinitis. Pectoralis Major Myocutaneous Flap is commonly used and Latissimus dorsi or rectus abdominis are used for larger defects. Their use is dictated by their availability, and blood supply.

In an oncological setting unlike infection, a non-biologic skeletal reconstruction can be considered. Customized titanium mesh is light weight, with high strength, biocompatibility, and an intrinsic diamagnetic character [9]. This must be anchored to the nearby ribs which reduces elasticity. Mesh reconstructions using PTFE, methyl methacrylate or a sandwich approach provide good rigidity and are preferred for large chest wall deficiencies. Mesh alone is not favourable as limited fluid permeability can cause infections and rigid chest wall pain [8]. Both the titanium plates and mesh require a viable soft tissue cover. Newer options like carbon-fiber, alumina ceramic molds are reproduced in 7 days and conform stability but have a high complication rate with increasing age and comorbidities [10]. A tailored approach is hence necessary to provide desired functional and cosmetic results. Post-operative infections in implanted material, local recurrences, technical and economic feasibility are some of the factors that influence the best reconstructive option.

Conclusion

Although, DFSP and FS-DFSP most commonly occur in the trunk those involving the sternum are very rare. Sternal involvement should not become a factor for incomplete resection, as wide excision with negative margins is potentially curative. A tailored approach should be employed to determine the best reconstructive option. Reconstruction should address functional stability and acceptable cosmesis. With the world literature and intellectual access available, a unique case can be tackled with various approaches and recourses at hand.

Acknowledgements

Special thanks to Dr. Vinod Vij for his valuable inputs. Special thanks to Dr Atul Limaye, cardiologist and Dr. Sweta Budyal, endocrinologist to provide a holistic cardiac and diabetic optimization in the peri-operative period.

The authors thank the members of the Editorial Advisory Board for their careful review and approval of the case report.

Conflicts of Interests

The Authors declare that there are no competing interests.

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How to Cite This Article
Chheda S, Behl A, Prasad S, Bangera S, Tehere A. Fibro sarcomatous conversion of a recurrent Dermatofibrosarcoma protuberans with sternal involvement. International Journal of Surgery Science. 2024;8(1):39-43.