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A Chest Wall Desmoid Tumor with Rib Encasement and Diaphragmatic Involvement

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Abstract

Background: Desmoid tumors (aggressive fibromatoses) are rare soft tissue tumors seen in abdominal and chest regions. We report a patient with a rare significantly larger desmoid tumor of the chest wall with bone encasement and diaphragmatic involvement. *Case Report*: We are reporting a 25-year-old lady who presented with complaints of pain and swelling over right lower chest since last 2 years. MRI and Trucut biopsy revealed it to be a spindle cell neoplasm. Histopathological and immunohistochemistry following wide surgical excision revealed it to be a desmoid tumor. *Conclusion*: This case is discussed because of the rarity of desmoid tumors of the chest wall, complexity of rib and diaphragmatic involvement and surgical challenges.

Keywords: Chest Wall, Desmoid, Diaphragm, Fibromatosis, Rib.

Introduction

Chest wall tumors have long represented a unique challenge for surgeons. In the past, an inability to perform successful reconstruction for large thoracic wall defects led to high peri-operative morbidity due to pleural cavity infection, respiratory failure and paradoxical breathing [1-4]. The role of surgical treatment for primary malignant tumors is now established following improvement in surgical reconstruction, antibiotics prophylaxis and anaesthesia [5].

Desmoid tumors (DT), a fibromatous proliferative disease, are defined by the World Health Organization as clonal fibroblastic proliferations that arise in the deep soft tissues and are characterized by infiltrative growth and a tendency toward local recurrence but an inability to metastasize [6]. Although surgery is the primary treatment modality, there remains significant controversy amongst surgeons regarding the management of disease. We report a patient with a rare, significantly large desmoid tumor of the chest wall with bone encasement and diaphragmatic involvement to discuss the rarity, complexity of structure involvement and surgical challenge in view of rib encasement and diaphragmatic involvement.

Case Report

A 25-year-old lady presented with pain over right lower chest since last 2 years. Swelling over right lower chest extended upto upper abdomen. Clinical examination revealed a hard 5×4×4 cm swelling over right lower chest fixed to 7th, 8th and 9th rib moving with respiration. CT scan of abdomen and chest revealed heterogeneously enhancing soft tissue 6.4×4.5×5 cm lesion noted in anterior abdominal wall of right hypochondrium enclosing the 8th and 9th ribs **[Fig.1]**.

Tru cut biopsy revealed spindle cell neoplasm having moderate differentiation spindly nuclei with bold chromatin. Intraoperatively there

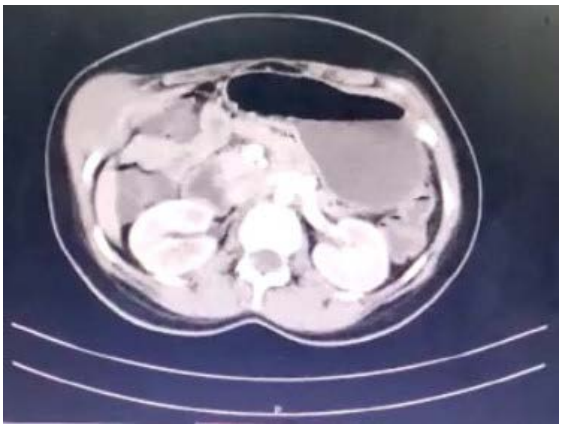

Fig.1: *CT scan showing parietal tumor chest wall with rib encasement.*

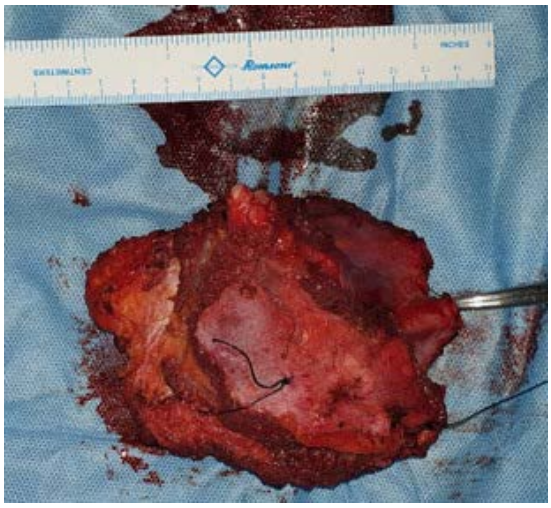

Fig.2: *Excised specimen with 2 cm margin with lower ribs partial excision.*

was a right chest tumor 6×8 cm over chest wall encircling 10th, 9th, and 8th rib abutting 11th and 7th extending to pleural cavity and diaphragm. Wide local excision and resection of diaphragm and rib and reconstruction of abdominal mesh plasty was done **[Fig.2,3]**. Patient had uneventful post-operative recovery. Histopathological evaluation and immunohistochemistry (IHC) revealed it as a case of desmoid type fibromatosis (5.5×5× 3.4 cm) with the cells are focally positive for SMA, negative for CD34, S100, Desmin and CD117, Ki 67- 3-4% **[Fig.4]**.

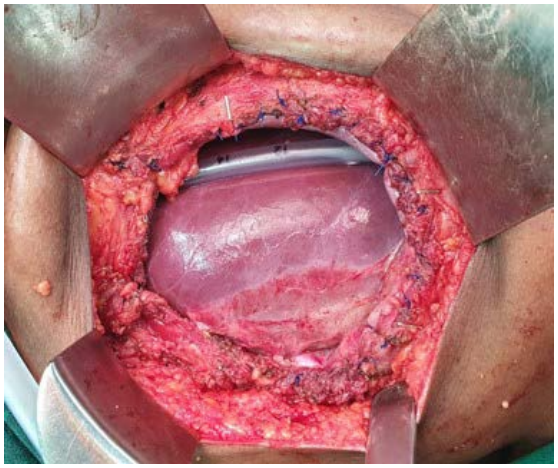

Fig.3: *Excised area with defect showing liver and diaphragm.*

Discussion

Desmoid tumors arise from fascia and connective tissue of the muscular layers and invade surrounding structures. They are mainly seen in the abdominal region [7]. Of the extra-abdominal sites, the chest wall is the most common [8], whereas the other sites are the shoulder girdle and inguinal regions. There is also a close association with familial adenomatous polyposis and Gardner's syndrome, suggesting the role of an intrinsic genetic defect in the development of desmoid tumors.

Treatment of these tumors involves wide local resections with stressing of the involved soft tissues and bony structures. Radiation therapy is usually used when a wide local excision cannot be accomplished. Chest wall tumors have long

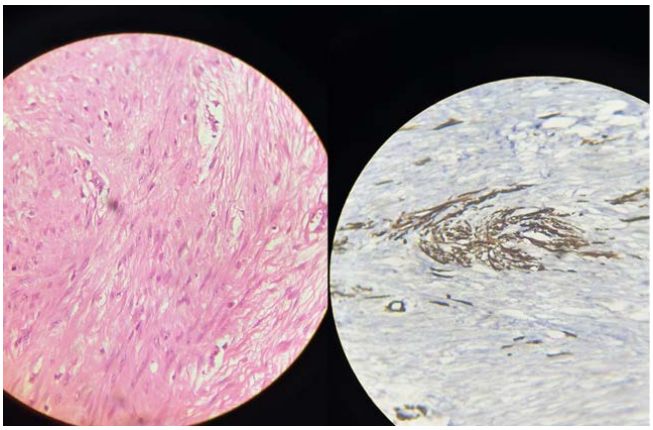

Fig.4: *Histopathology and IHC of the lesion.*

represented a unique challenge for surgeons and inability to perform successful reconstruction for large thoracic wall defects led to high peri-operative morbidity especially when it involves pleura and diaphragm. The advancement of surgical gadgets, modalities for surgical reconstruction, antibiotics prophylaxis and anaesthesia has now established the role of surgical treatment for primary malignant tumors.

Extra-abdominal desmoid tumors have a high local recurrence rate after resection [9]. In a recent series of chest wall desmoid tumors, although the overall 5-year survival rate was 93%, the 5-year local recurrence rate despite aggressive surgical intervention was 29%. [10], pointing to stress the importance of continued follow up in these patients even after attaining optimal wide tumor-free surgical resection margins, but these tumors rarely metastasise. This case was presented due to the complexity of the tumor, rarity of location and successful post-operative outcome of a rare complex presentation of desmoid tumors.

Conclusion

A rare large desmoid tumor of the chest wall is reported to discuss the complexity of structure involvement and surgical challenge in view of rib encasement and diaphragmatic involvement.

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