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## 6 7 **Giant Mediastinal Myxoid Pleomorphic Liposarcoma**

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### 13 14 **Introduction**

15 An 18 years old girl with no past medical history was experiencing progressive shortness of breath  
16 for 3 months. At presentation, she was tachypneic, tachycardiac, hypotensive and was not able to  
17 lie flat.

18  
19 Her chest x-ray revealed abnormal contouring of the mediastinum (Figure 1A). Contrasted CT of  
20 the chest showed a large heterogenous mass occupying the anterior mediastinum, right and left  
21 hemithorax with posterior displacement of the heart and atelectasis of the lungs. (Figure B, C) The  
22 patient underwent urgent surgical resection. The mass was exposed thorough a clam shell incision  
23 exposing the anterior mediastinum and bilateral pleura (Figure 2A). The mass was yellow,  
24 encapsulated and lobulated. It was resected en-block with intra-operative frozen section  
25 confirming negative margins. The mass weighed 3208 grams and was 22 × 18 × 13 cms. (Figure  
26 2B)

27  
28 Post-operatively, the patient did well with complete resolution of her symptoms. The final  
29 histopathology proved the mass to be high grade malignant pleomorphic myxoid liposarcoma.  
30 (Figure 3 & 4). MDM2 and cyclin-dependent kinase 4 were both negative. She received adjuvant  
31 chemotherapy and is now on close surveillance by the oncologist.

32

33 Consent was taken from the patient for the publication of the case and images.

34

35 **Comment**

36 Liposarcoma is a relatively uncommon malignant tumor of adipose tissue that can occur at  
37 anybody site. <sup>1</sup> For rare instances it arises primarily in the mediastinum.<sup>2</sup> The recent World Health  
38 Organization (WHO) classification has classified adipocytic tumors into: 1) Benign 2)  
39 Intermediate (locally aggressive) and 3) Malignant. Malignant tumors include: 1) Well  
40 differentiated liposarcoma: lipoma-like, sclerosing, inflammatory 2) Dedifferentiated liposarcoma  
41 3) Myxoid liposarcoma 4) Pleomorphic liposarcoma 5) Myxoid pleomorphic liposarcoma. <sup>3, 2, 4</sup>

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43 Myxoid pleomorphic liposarcoma is a new entity in this classification. Unlike other liposarcomas,  
44 its most common anatomical site is in the mediastinum followed by the limbs and neck.  
45 Histologically, it shows a mixture of both myxoid and pleomorphic liposarcoma.<sup>3</sup> Mediastinal  
46 liposarcoma may grow to substantial size before causing any symptoms. The most common  
47 presentation is incidental finding on chest X-ray. Symptomatic patients may have cough, dyspnea,  
48 dysphagia, and chest pain.<sup>5</sup>

49

50 Radiologically, the tumor appears inhomogeneous in CT scan with a difficulty to differentiate it  
51 from other forms of sarcoma. MRI can differentiate lipoma from well differentiated liposarcoma  
52 (WDL).<sup>6</sup> In Lipoma, MRI shows high intensity in T1 and T2 weighted images as this represents  
53 the uniform structure with fatty tissue. In contrast, high grade liposarcoma show low intensity in  
54 T1 images. In addition, Liposarcomas tends to be larger and has more thick septa.<sup>7,8</sup> As the entity  
55 is rare, there are no randomized trials that assess different treatment modalities. Current practice  
56 is based on case reports and series.<sup>4</sup> Complete surgical resection remains the mainstay for the  
57 treatment of myxoid pleomorphic liposarcoma.<sup>8</sup> Wide resection with negative margins is the goal.  
58 However, anatomical location plays an important role whether this is possible. Patients with huge  
59 mediastinal mass that present late may pose high risk surgical candidates as the compression of  
60 vital structures, such as the heart and the lung, may cause critical hemodynamic issues during  
61 surgery.<sup>9</sup>

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63 Myxoid pleomorphic liposarcoma has an aggressive clinical presentation with high recurrence rate  
64 and distant metastasis following surgical resection. Approximately 40% recur after surgery.<sup>8</sup> Thus,  
65 discussion in a multidisciplinary tumor board is recommended to assess the need for adjuvant  
66 radiotherapy with or without chemotherapy after surgical resection.<sup>9</sup> Although 90% of mediastinal  
67 WDL show 12q12-15 amplicon that represents amplified oncogenes MDM2 and CDK-4, no  
68 genetic aberration has been associated with myxoid pleomorphic liposarcoma.<sup>3,9</sup>

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70 In conclusion, primary mediastinal myxoid pleomorphic liposarcoma is a rare entity and tend to  
71 present at an advanced stage. Recognition of the need for complete surgical resection followed by  
72 adjuvant therapy is very important.

73

#### 74 **Authors' Contribution**

75 AHK performed the surgery of this case. EK assisted in the surgery. FAK prepared and reported  
76 the radiological images. MR prepared the pathology of the patient and reported the pathological  
77 images. AHK and EK drafted and edited the manuscript. All authors approved the final version of  
78 the manuscript.

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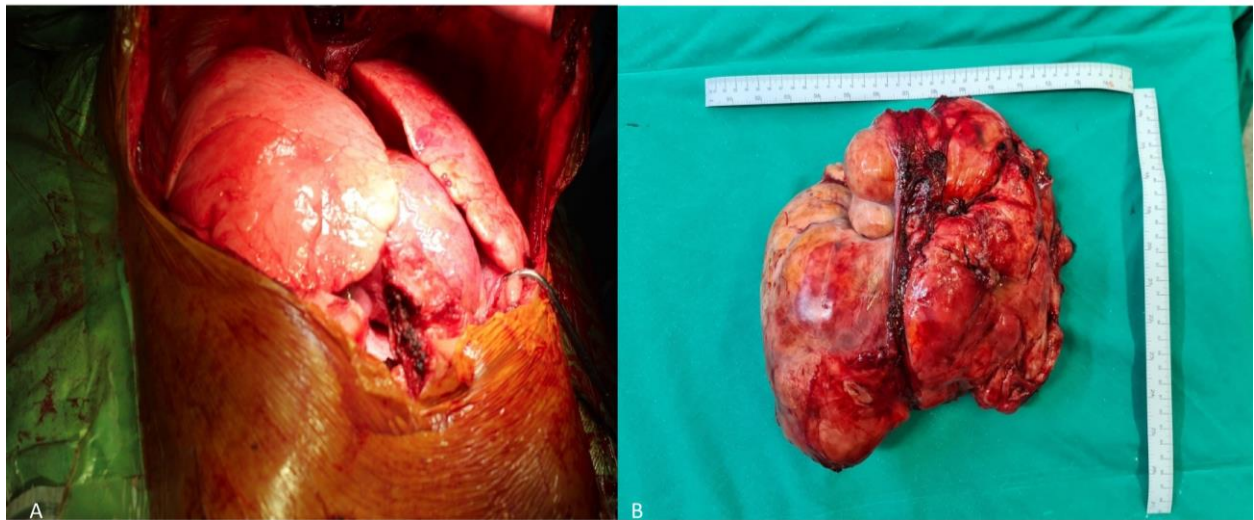
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105 **Figure 1: A:** Anteroposterior chest radiograph showing an abnormal contouring of the  
 106 mediastinum with a large lesion in the anterior mediastinum and the left lower hemithorax. Hilar  
 107 overlay sign is noted with both hilar shadows identified through the opacity of the abnormal  
 108 lesion. **B & C:** CT-scan Images of the chest with IV contrast, **(B)** Axial **(C)** Coronal , showing a  
 109 large heterogenous mass occupying the anterior mediastinum, right and left hemithorax with  
 110 posterior displacement of the heart and atelectasis of the lungs . The mass shows areas of low  
 111 density (\*) likely representing fat content and other areas of high density (\*\*) representing  
 112 calcification.

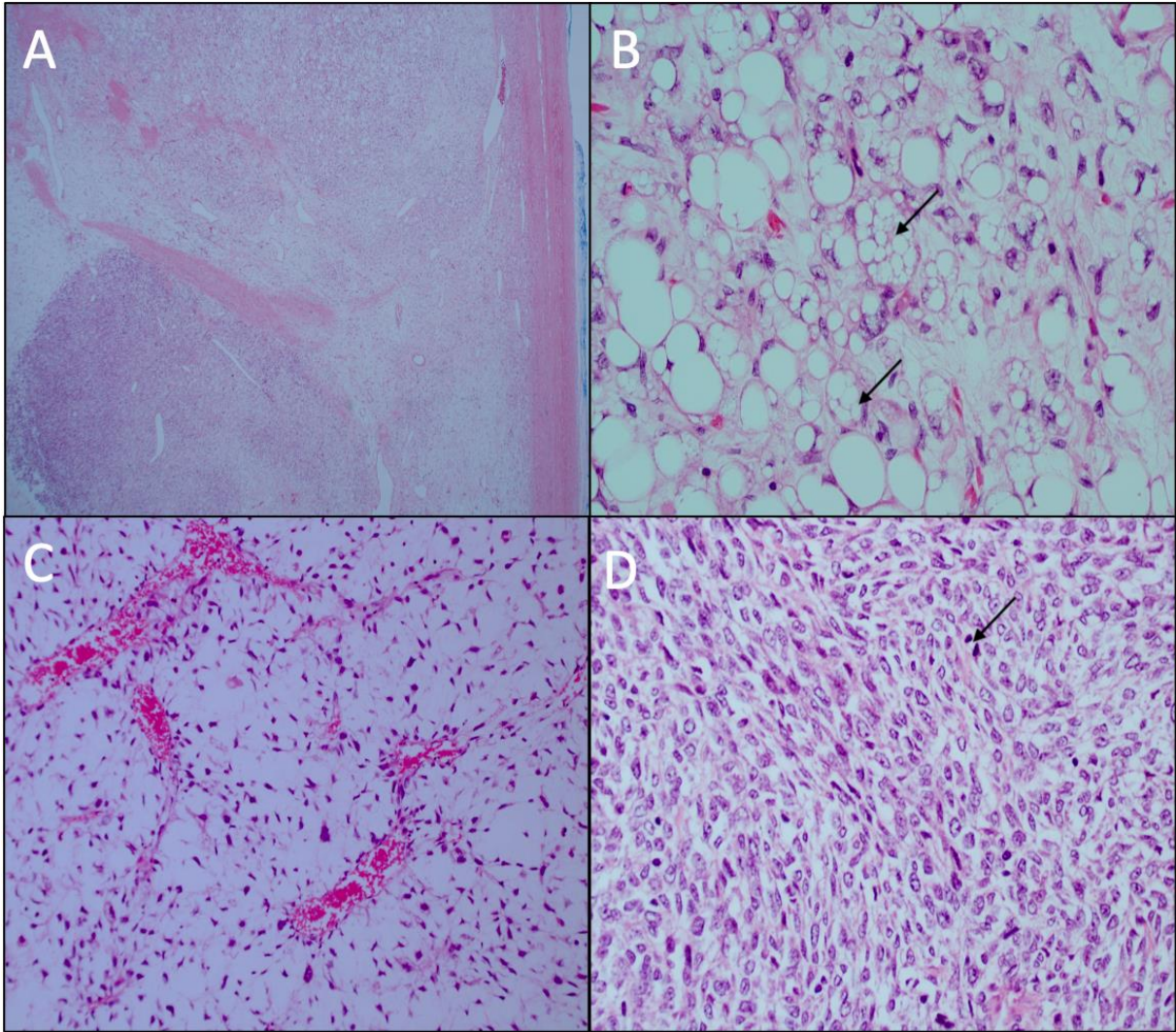
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115 **Figure 2: A:** Clam-shell incision exposing the anterior mediastinum and bilateral pleura. **B:** The  
 116 mass was yellow, encapsulated and lobulated measuring was 22 × 18 × 13cms.

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119 **Figure 3:** Microscopic examination showed an encapsulated multinodular malignant neoplasm

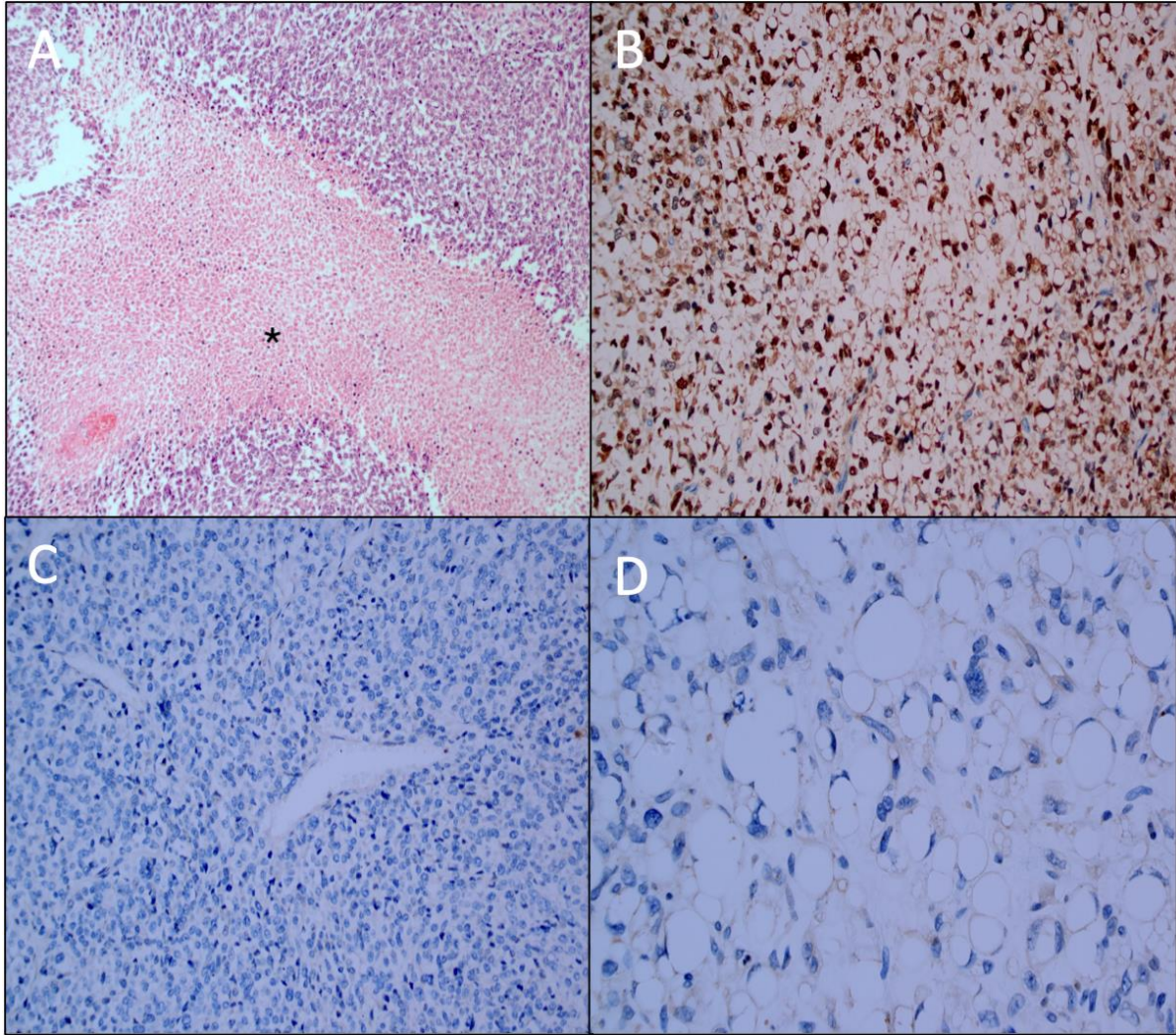
120 of variable cellularity (A). There were lipomatous areas showing atypical multivacuolated

121 lipoblasts (arrows) (B). Other areas showed myxoid stroma containing plump spindle to stellate

122 cells (C). There were highly cellular nodules formed of fascicles of pleomorphic spindle cells

123 with frequent mitotic figures (arrow) (D).

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**Figure 4:** Areas of geographic coagulative type necrosis were seen (asterisk) (**A**). The tumor cells in the lipomatous areas were strongly positive for s-100 (**B**) but this was negative in the cellular pleomorphic areas (**C**). Tumor cells were negative for MDM2 and CDK4. CDK4 is shown here (**D**).