Giant Mediastinal Myxoid Pleomorphic Liposarcoma

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Introduction

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

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

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

**Comment**

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

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

Radiologically, the tumor appears inhomogeneous in CT scan with a difficulty to differentiate it from other forms of sarcoma. MRI can differentiate lipoma from well differentiated liposarcoma (WDL). In Lipoma, MRI shows high intensity in T1 and T2 weighted images as this represents the uniform structure with fatty tissue. In contrast, high grade liposarcoma show low intensity in T1 images. In addition, Liposarcomas tends to be larger and has more thick septa. As the entity is rare, there are no randomized trials that assess different treatment modalities. Current practice is based on case reports and series. Complete surgical resection remains the mainstay for the treatment of myxoid pleomorphic liposarcoma. Wide resection with negative margins is the goal. However, anatomical location plays an important role whether this is possible. Patients with huge mediastinal mass that present late may pose high risk surgical candidates as the compression of vital structures, such as the heart and the lung, may cause critical hemodynamic issues during surgery.
Myxoid pleomorphic liposarcoma has an aggressive clinical presentation with high recurrence rate and distant metastasis following surgical resection. Approximately 40% recur after surgery. Thus, discussion in a multidisciplinary tumor board is recommended to assess the need for adjuvant radiotherapy with or without chemotherapy after surgical resection. Although 90% of mediastinal WDL show 12q12-15 amplicon that represents amplified oncogenes MDM2 and CDK-4, no genetic aberration has been associated with myxoid pleomorphic liposarcoma.

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

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

References


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

Figure 2: A: Clam-shell incision exposing the anterior mediastinum and bilateral pleura. B: The mass was yellow, encapsulated and lobulated measuring was 22 × 18 × 13cms.
Figure 3: Microscopic examination showed an encapsulated multinodular malignant neoplasm of variable cellularity (A). There were lipomatous areas showing atypical multivacuolated lipoblasts (arrows) (B). Other areas showed myxoid stroma containing plump spindle to stellate cells (C). There were highly cellular nodules formed of fascicles of pleomorphic spindle cells with frequent mitotic figures (arrow) (D).
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).