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A case of a rare primary tumor of the mediastinum presenting with superior vena cava syndrome

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CASE REPORT

A 44-year-old active man presented with a two-month history of shortness of breath on exertion with an insidious onset. This was associated with swelling of his face and eyes that was worse in the mornings. There were no other symptoms. All his vital signs were normal.

Chest X-ray demonstrated a large soft tissue density in the mediastinum extending into the right upper zone, measuring 10.7 cm across, resulting in tracheal displacement. This was confirmed on CT imaging (Figure 1) which showed a mixed density mass filling the mediastinum and extending into both pleural cavities. The superior vena cava and innominate vein was laterally displaced alongside right tracheal deviation and bilateral compression of upper lung lobes. Percutaneous biopsy suggested a benign soft tissue tumour with features suggestive of a lipoma variant and no evidence of malignancy.

After multidisciplinary discussion, initial surgical therapy was performed. Resection via median sternotomy was performed. The excised tumor (Figure 2) was large in nature and was completely excised including tumor extensions into the neck. Full mediastinal clearance was performed due to the indurated and inflamed appearance of the mediastinal fat. Histology demonstrated well-differentiated liposarcoma with areas of low grade fibrosarcoma-like dedifferentiation (Figures 3 and 4). Recovery was uneventful and the patient underwent adjuvant radiotherapy, remaining symptom free at immediate follow-up.

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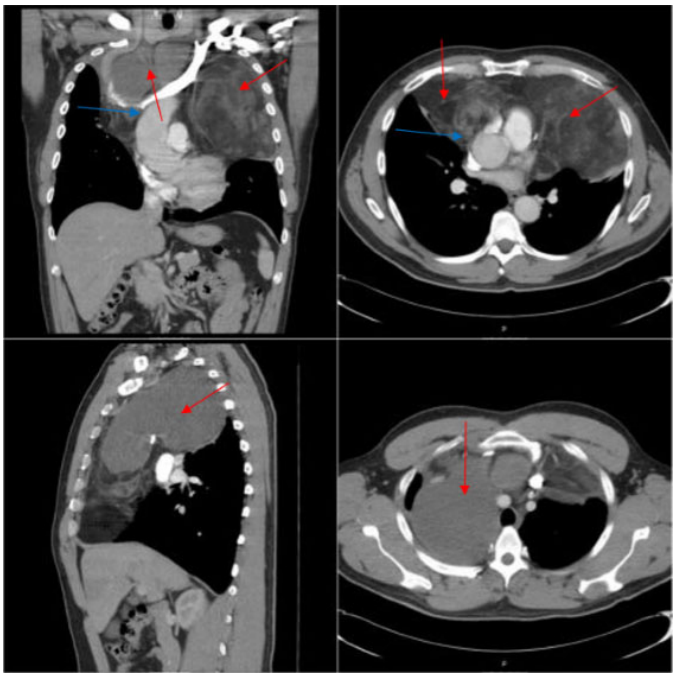

Figure 1: Multi-planar CT images demonstrating a large mediastinal tumor (red arrows) with SVC and innominate vein compression. There is complete obliteration of the SVC in the coronal and axial planes (blue arrows).

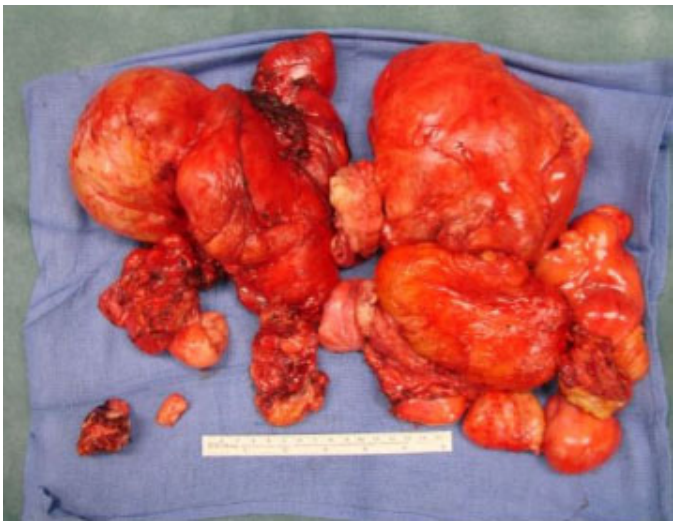

Figure 2: Large excised mediastinal tumor with 15 cm ruler as reference size.

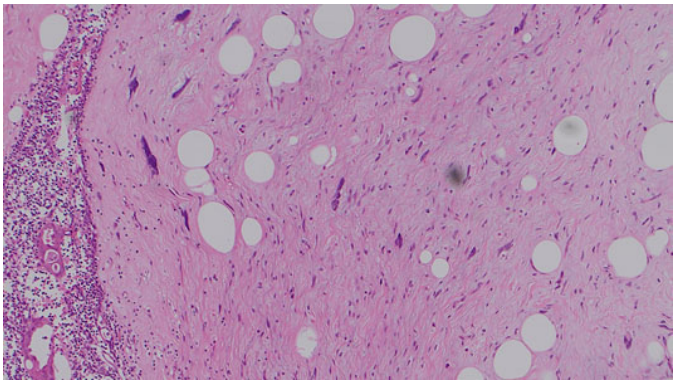

Figure 3: Area of dedifferentiated liposarcoma showing nuclear pleomorphism with scattered adipocytes and adjacent residual normal thymus (left).

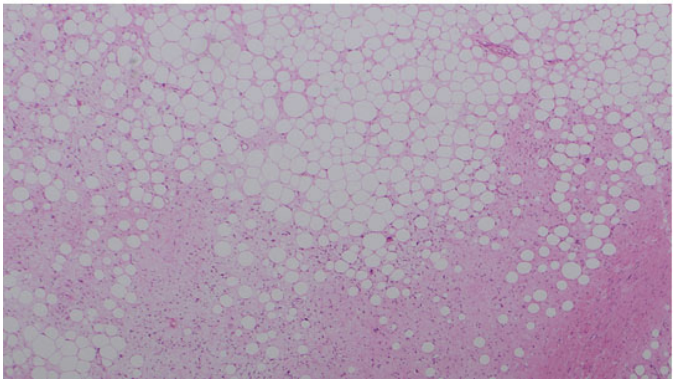

Figure 4: Well differentiated liposarcoma with variably sized adipocytes (upper half) with area of dedifferentiation (lower half).

Unfortunately the patient returned two years later and CT scan showed recurrence involving the supraclavicular and retroclavicular area with right paratracheal extension. The tumour was re-excised but was found to be adherent to the thyroid with infiltration into one of the lobes. The patient underwent post-operative radiotherapy. Unfortunately two years later further recurrence of the liposarcoma was found. After multidisciplinary discussion the patient was deemed not for further surgery, and had palliative chemotherapy before succumbing to the disease process.

DISCUSSION

Mediastinal tumours after frequently symptomless until they reach a size resulting in mass effects [1]. Superior vena cava (SVC) obstruction is generally a late sign as the tumor will have taken up a large portion of the mediastinum. Superior vena cava syndrome is highly suggestive of thoracic malignancy and is generally associated with poor prognosis [2]. Management of SVC syndrome is determined largely by the underlying pathology and can include chemo/radiotherapy as well as stenting or surgical bypass in selected cases [2]. In the above case a mediastinal liposarcoma resulted in SVC syndrome and after surgical resection, the symptoms rapidly subsided.

Liposarcomas, malignant tumors arising from adipose tissue are rare in comparison to other adipose tissue derived tumors and have a range of malignant potential [1]. Four histological entities are described: myxoid, round-cell, well-differentiated, and pleomorphic [1]. Treatment for all types include surgical resection followed by adjuvant radiotherapy. The role of chemotherapy in these mediastinal tumors is still uncertain [1]. Poor prognostic factors for liposarcomas irrespective of histological subtype are increasing tumor size (especially >10 cm), higher tumor grade (Grades II and III), involved surgical resection margin and if clinical presentation was in the recurrent stage of the disease [3]. However, differences in outcome according to histologic variance is harder to quantify as outcomes can be dependent on tumor location and administration of radiotherapy which is likely a reflection on the ability to achieve adequate surgical margins. What is known from large case series is that myxoid liposarcomas are likely to be classified as grade I and 10-year incidence of distant metastasis was only 5% which confers a more favorable prognosis [3]. The presence of >5% round cells on histology can result in a >1.5 risk of dying from the disease but has a similar distant-metastasis rates to that of the pleomorphic and well-differentiated liposarcoma variant [3]. Well-differentiated liposarcomas as in the above case typically have a high mortality with a five-year survival rate of 65% [3]. Recurrence is possible; our case is highlighting the importance of follow-up despite multiple surgical resections.

CONCLUSION

Primary mediastinal liposarcomas are extremely rare mesenchymal tumors that have high potential of recurrence even after surgical excision. This case describes a classic presentation of a rare mediastinal liposarcomas which account for approximately 1% of mediastinal tumors. These tumors often present slowly with symptoms due to mass effect. Superior vena cava syndrome should prompt expedited investigation for the patient's presentation as it is often associated with malignancy and often in the later stages of the disease. There is limited research, and more studies should be directed toward the best management course of this disease.

Keywords: Mediastinal liposarcoma, Primary mediastinal malignancy, Superior vena cava (SVC) syndrome

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Author Contributions

Ramanen Sugunesegran – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Christina Kumate – Conception of the work, Design of the work, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Cody Ross – Acquisition of data, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.