Primary Mediastinal Liposarcoma of the Superior, Middle, and Anterior Mediastinum

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ABSTRACT

Primary liposarcomas constitute less than 1% of all mediastinal tumors. The posterior mediastinum is the location in 95% of cases. Literature search did not show a case that encompassed three mediastinal compartments.

A 32-year old man was diagnosed with high grade primary mediastinal liposarcoma (80% myxoid plus 20% round cell), after presenting with superior vena cava syndrome. CT-scan revealed lobulated masses on the superior, anterior, and the right middle mediastinum measuring 7.8 x 9.0 x 7.5 cm and compressing the superior vena cava. He underwent surgical debulking and palliative 3 cycles doxorubicin-based chemotherapy, but later succumbed to pneumonia.

Key Words: primary, mediastinal, liposarcoma

Introduction

Liposarcomas are the most common soft tissue sarcoma of adults, usually arising in the extremities or the retroperitoneum.[1] Primary mediastinal liposarcomas are extremely rare, with only less than 100 cases reported in literature.[2] Because of its lower frequency, documentation in literature is limited and the clinical features of primary mediastinal liposarcomas still remain unclear.[3,4]

A liposarcoma case primary on the superior, anterior, and the right middle mediastinum, who presented with superior vena cava syndrome is presented.

Case Report

A 32-year old man from the Philippines presented with a three-month history of right facial and extremity edema. His chest radiograph revealed a right suprahilar mass, which CT-scan read as lobulated masses on the superior, anterior, and the right middle mediastinum measuring 7.8 x 9.0 x 7.5 cm and compressing the superior vena cava. Open biopsy of the mass with frozen section revealed a high grade liposarcoma.

Patient only followed-up after 2 months, dyspneic with worsening edema. Repeat CT-scan revealed 18.2 x 8.9 x 9.7 cm mass encasing and compressing the azygous vein, superior vena cava, ascending aorta, and the aortic arch (Figure 1). A pleural nodule was now seen in the right lower lung lobe measuring 1.0 x 2.0 x 1.1 cm.

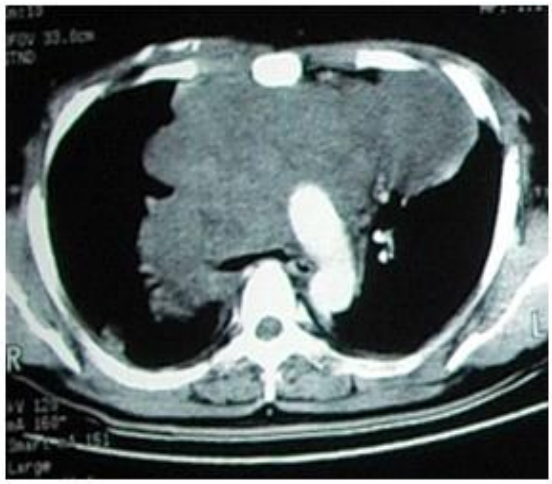

Figure 1. Chest CT-scan with heterogeneously enhancing mass in the superior, anterior, middle mediastinum encasing major vessels.

Patient underwent surgical debulking of the mass, reducing it by more than 50%. Histopathology revealed predominantly myxoid liposarcoma (80%) with round cell liposarcoma component (20%) (Figure 2).

The patient was given 3 cycles of doxorubicin but the mass increased in size (Figure 3). Three weeks after the 3rd cycle chemotherapy, the patient developed pneumonia and expired.

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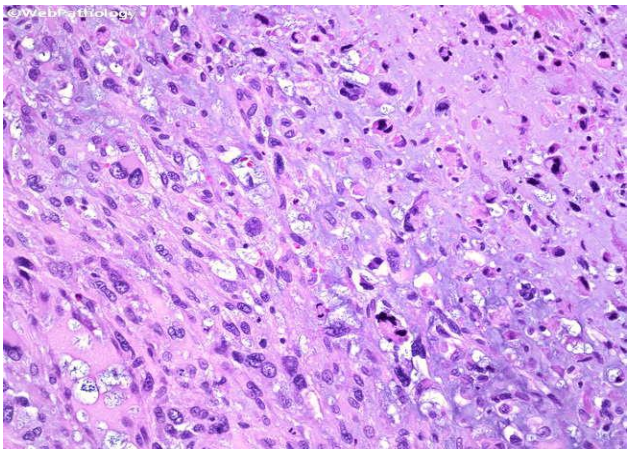

Figure 2. Histopathology with 80% myxoid liposarcoma and 20% round cell liposarcoma

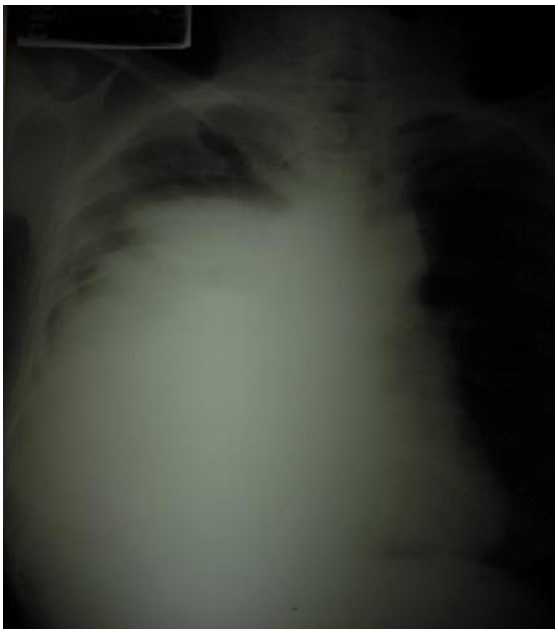

Figure 3. Repeat chest x-ray showing interval increase in mass size

Discussion

Primary liposarcoma represents less than 1% of all mediastinal tumours.[2] It usually occurs in adults, with most cases in patients more than 40 years old.[5-6] Malignant liposarcoma develops more commonly in the posterior mediastinum and its occurrence in anterior mediastinum is very rare with only a few cases that have been reported in international literature.[7] This case presented had a CT-scan revealing involvement of 3 mediastinal compartments. This is extremely rare and literature review did not report such 3 mediastinal compartment involvement.

Mediastinal liposarcomas may, on account of excessive size or direct invasion of contiguous vital structures, such as heart, trachea, bronchi, and esophagus etc., cause functional limitations, such as pain, cough, shortness of breath with exertion or dyspnea or dysphagia.[8-9] They are usually identified by routine chest radiograph or are discovered after the patient presents with symptoms secondary to the intrathoracic structure compression.[10] In the patient's case, there was encasement of the superior vena cava, and with its continued growth, caused compression of this major vessel. On review of literature, superior vena cava syndrome was the presenting symptom of patients with mediastinal sarcomas in 37%.[11]

The radiologic features of mediastinal liposarcomas are nonspecific but are suggestive of the diagnosis. The predominant finding on conventional chest radiography is a widened mediastinum. On CT-scan, the appearance of mediastinal liposarcomas, the same as those located in other sites of body, varies from a predominantly fat-containing mass to a solid mass. Low attenuation values between –50 to –150 Hounsfield Unit are consistent with a tissue composed of fat. Greater values are related to the necrosis, heterogenity and soft tissue component in liposarcomas. A differential diagnosis should be made between lipoma, thymolipoma, teratoma, lymphoma, or germ cell tumor.[12]

Liposarcomas are characterized by their large size and their variable histologic subtypes, which correlate with the clinical behavior and the prognosis. The most recent World Health Organization classification of soft tissue tumors recognizes 5 categories of liposarcomas: (1) well differentiated, which includes the adipocytic, sclerosing, and inflammatory subtypes; (2) dedifferentiated; (3) myxoid; (4) round cell; and (5) pleomorphic. Patients with dedifferentiated or pleomorphic liposarcomas have a significant poor survival and prognosis than those with myxoid or well-differentiated liposarcomas.[13] Well differentiated low-grade liposarcomas, also known as atypical lipomatous tumors, have histologic features in many areas resembling mature adipose tissue. Evans reported that atypical lipomatous tumors may transform to dedifferentiated liposarcomas and usually do not metastasize.[12] The patient had predominantly myxoid with some round cell component, and these subtypes are considered to be less aggressive than the dedifferentiated subtype.

Surgical removal is the optimal treatment for a mediastinal liposarcoma. According to literature, aggressive surgical intervention seems to favour the quality of life of patients and prolong survival, being considered the most effective treatment.[14] If the entire tumor cannot be resected, surgical debulking often results in symptomatic relief. Chemotherapy and radiotherapy are still to be explored for effective strategies. These may be added as adjuncts to

surgical excision in unresectable or incompletely resected tumors, but liposarcomas seem to have low sensitivity.[15-16]

Prognosis depends on the histological subtype and on surgical resection with wide margins of safety.[17] Recurrence is common in deep-seated liposarcomas and it becomes apparent within the first 6 months in most cases, but it may be delayed for 5 or 10 years following the initial excision.[18]

Summary

A 32-year old man presented with SVC syndrome and was diagnosed with a primary mediastinal liposarcoma, predominantly myxoid that involved three mediastinal compartments. He underwent surgical debulking with interim relief of symptoms. Palliative chemotherapy with an anthracycline was attempted but was not effective. The patient later succumbed to a nosocomial infection. Mediastinal liposarcomas, because of their innate resistance to chemotherapy and radiotherapy, are very challenging to treat especially in the locally advanced and metastatic setting.

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