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Case Report

A rare mediastinal tumor-fat-forming solitary fibrous tumor

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ABSTRACT

Solitary fibrous tumor is uncommon spindle cell mediastinal tumor arising mainly from pleural surface. Authors discuss a clinic-radiological features of Fat forming SFT which is rare variant of SFT in 70yrs old male patients. Initially we kept differential of Liposarcoma, Lymphoma on clinico-radiologically features but later after histopathology and Immunohistochemistery turns out to be Solitary fibrous tumor. Here accurate classification is important as SFT classified as intermediate type (rarely malignant), require wide surgical excision with close follow-up.

Keywords: Immunohistochemistry, Mediastinum, Solitary fibrous tumor

INTRODUCTION

Solitary Fibrous Tumor (SFT) constitutes a heterogeneous group of rare spindle-cell tumors that include benign and malignant neoplasms. Their cell of origin is still debated. SFT is preferred by most pathologists as a better term than "hemangiopericytoma" that gathers numerous unrelated entities and is presently only employed by neuropathologists.[1] Most tumors present as well defined, slow growing masses, which can be cured by surgery. A small percentage of SFTs, between 10-20%, behave in a more aggressive way, with local recurrence and/or distant metastasis for which systemic therapy (chemotherapy or targeted treatment with e.g. sunitinib) can be given.[2] The fat-forming variant of SFT involving the thoracic cavity is very rare with only few cases reported involving the mediastinum, epicardium and lung.[3,4]

There are 3 typical primary locations: pleural, meningeal and extra-thoracic soft tissue, but we will restrict our discussion to pleural SFT as in present case. SFT of the pleura occurs mainly in adults. The sex incidence is equal, and they are seen in all age groups, although they most commonly present in the 60s and 70s.

CASE REPORT

A 77 years old male, non-smoker, ex-army personal presented with complain of dull pain in right side of chest, since 2 months, hoarseness of voice since 15 days, acute onset dyspnea which progressed to grade 4 (mMRC) since 8 days. There was no history of fever, cough, expectoration, weight loss. He is known Hypertensive since 10 years. On examination he was tachypneic with SpO2 of 95%, at rest. General examination was with in normal limit. In respiratory system air entry reduced on right side with occasional rhonchi.

Complete blood picture shows anaemia with normal leucocyte counts. Renal, liver function and bleeding profile is within normal limits. We asked for chest x-ray which shows mediastinal widening with homogenous soft tissue opacity extending into the both sides (right more than left, size 9.5cm x 7.5cm), involving the mid and lower zones (Figure 1).

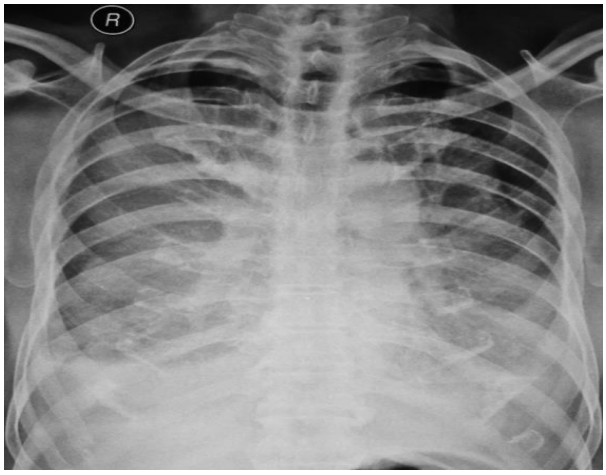

Figure 1: Plain frontal chest radiograph showing mediastinal widening with homogenous soft tissue opacity extending into the both sides (right more than left), involving the mid and lower zones. There is bilateral costophrenic angle blunting.

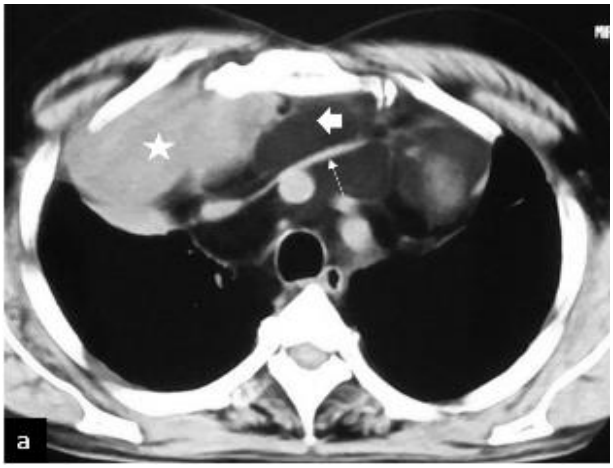

Figure 2: CT thorax a) Axial NECT section at superior mediastinal level showing a fatty mass (thick arrow) encasing the large vessels (arrow points to the stretched attenuated left brachiocephalic vein); towards the right side is a soft tissue density peripheral mass (star).

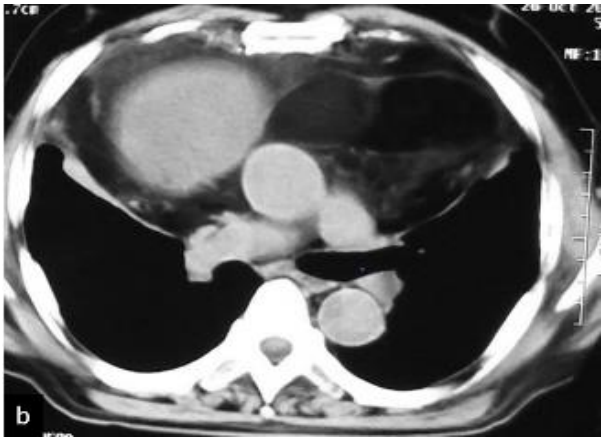

Figure 2: b) Axial NECT section image at slightly inferior level.

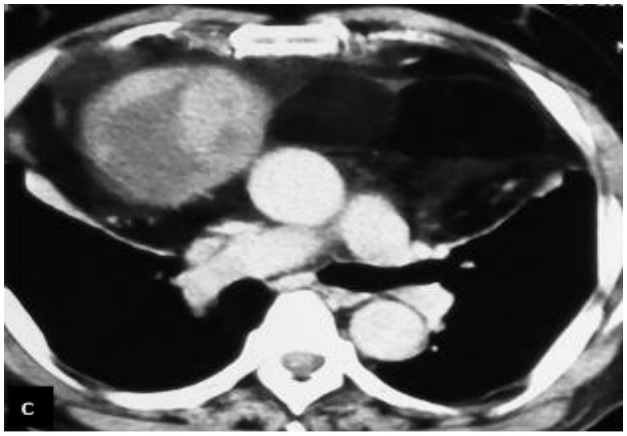

Figure 2: c) corresponding axial CECT section showing heterogeneous enhancement of the soft tissue portion; there is large fatty component on the left side abutting the great vessels.

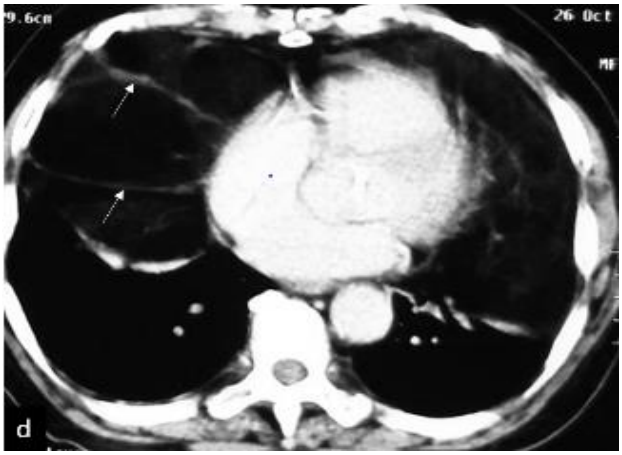

Figure 2: d) CECT section further inferiorly shows the fatty mass abutting the pericardium; multiple thin soft tissue density strands are seen passing through the mass (thin arrows).

There is bilateral costophrenic angle blunting. CT thorax a) Axial NECT section at superior mediastinal level showing a fatty mass (thick arrow) encasing the large vessels (arrow points to the stretched attenuated left brachiocephalic vein); towards the right side is a soft tissue density peripheral mass (star) b) Axial NECT section image at slightly inferior level and c) corresponding axial CECT section showing heterogeneous enhancement of the soft tissue portion; there is large fatty component on the left side abutting the great vessels d) CECT section further inferiorly shows the fatty mass abutting the pericardium; multiple thin soft tissue density strands are seen passing through the mass (thin arrows) (Figure 2). CT guided biopsy shows densely composed stroma with scattered spindle shaped cell with bland nuclei, finding supportive for fibrosing lesion (sclerosing mediastinitis/ myofibroma). Immunohistochemistry came out to be positive for CD34 and favours fat-forming solitary fibrous tumor (lipomatous hemangiopericytoma). In most cases surgery

is the only treatment necessary. Surgeons remove the tumor and a small margin of healthy tissue that surrounds it. It tumour is very large like in present case radiation therapy given to reduce the size followed by operation. In few case chemotherapy has been also given in patients in which tumor spread to other parts of body. Present case refers to radiation oncology at higher center after that patient loss to follow.

DISCUSSION

Solitary fibrous tumors are rare growths of soft tissue cells that can develop nearly anywhere in the body. It mostly often occurs in the lining around the outside of the lungs (pleural solitary fibrous tumors). Fat-forming solitary fibrous tumor is a rare variant of solitary fibrous tumor, usually occurring in middle-aged patients commonly between 40-60 years. Although there are very few cases report available in literature, the small case series shows that it is common between 20 to 93 years.[5] Fat-forming SFT often presents as a heterogeneous lesion, with solid enhancing regions and scattered low-attenuating adipose areas on CT examination.[6] Present case is 70 years old male having similar radiological presentation. Clinical presentation is highly unpredictable usually present in larger tumors and occur more frequently in malignant subtypes.[7] Symptoms occur mainly by either local compression/invasion of adjacent thoracic structures (e.g., dyspnea, angina, and/or cough) or paraneoplastic syndromes (e.g., hypoglycemia, weight loss, and/or pulmonary osteoarthropathy).[8,9] Present case presented with Dyspnea, chest pain and hoarseness of voice as described in literature. In our case tumour arose from the pleura, authors also found large area of fatty component within the tumour, which may have led to a differential diagnosis that included soft-tissue tumours containing a lipomatous component, such as liposarcoma and atypical lipomatous tumour. Authors did CT guided biopsy which shows densely composed stroma with scattered spindle shaped cell with bland nuclei, finding supportive for fibrosing lesion. Authors did immunohistochemistry of sample that came out to be positive for CD34 and favours fat-forming solitary fibrous tumor (lipomatous hemangiopericytoma). SFTs commonly express CD34, CD99, and Bcl-2; epithelial membrane antigen and smooth muscle actin may also be expressed. They are usually negative for S-100 protein, desmin, and cytokeratin. World Health Organization (WHO) classify this tumor in intermediate category, considering potential aggressive behaviour and so wide surgical excision with long term follow-up required.[10]

CONCLUSION

Clinico-radiological and histiopathological finding are not enough for treatment decision in many tumor. Iminohistochemistery of tissue sample also required to reach final diagnosis. Like in our case we initially suspected liposarcoma but histopathology favour for myofbroma and immunohistochemistery favour solitary fibrous tumor.

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