Case Report

Giant solitary fibrous tumor pleura: Clinical dilemma and diagnosis

Amulyajit Kaur, Satyajit Singh Gill[1], Jaswinder Singh[2], Amandeep Singh[3], Narayanan Subramaniam Mani[1]

Department of Pathology, Military Hospital (Cardiothoracic Centre), Pune, [1]Department of Pathology, Armed Forces Medical College, Pune, Maharashtra, [2]Departments of Surgery, [3]Radiology, Military Hospital (Cardiothoracic Centre), Army Hospital (R and R), New Delhi, India

ABSTRACT

Solitary fibrous tumors in the lung are rare neoplasms with distinct clinicopathological and immunohistochemical features. We report a giant solitary fibrous tumor of the pleura in a young male which remained silent clinically till it assumed gigantic proportions. Histology and immunohistochemistry were classical of a solitary fibrous tumor. Inspite of its appalling size it proved to be benign in behavior. This case depicts the vulnerability of this lesion to a clinical bungle. The report highlights the significance of clinical suspicion of this rare neoplasm and reveals the diagnostic associations and procedures to avoid emperical therapy and delay in curative surgical treatment.

KEY WORDS: Clubbing, giant solitary fibrous tumour, pleura

Address for correspondence: Dr. Amulyajit Kaur, Department of Pathology, Command Hospital (Western Command), Chandimandir-134 107, India.
E-mail: amulyabhatti@rediffmail.com

INTRODUCTION

Solitary fibrous tumors (SFT) of the pleura are rare tumors, comprising only 5% of pleural neoplasms. Literature reveals only two case series and a few solitary reports.[1] We report a unique case of a gigantic SFT presenting with low grade fever. The shadow caused by the tumor on plain X-ray chest suggested a pleural effusion. The patient was initially placed on antitubercular treatment. It was only when attempted pleural taps failed and help from the ultrasonologist was sought that information of the presence of a solid neoplasm mimicking pleural effusion on chest X-ray could be revealed. This illusory picture on X-ray chest has not yet been reported in literature and has a possibility of occurring in our country where facilities of higher specialized imaging techniques are resorted to sparingly in the periphery, mainly due to non availability and exorbitant costs involved.

Access this article online

Quick Response Code:

Website: www.lungindia.com

DOI: 10.4103/0970-2113.95338

CASE REPORT

A well-built 39- year old male presented with low-grade continuous fever over a period of 1 month and mild discomfort left lower side of the chest. On examination he was found to have low-grade fever, clubbing of nails, and absent breath sounds in the left infra-axillary and infrascapular regions. On investigating, hemoglobin and the blood counts were in normal range. ESR was 44 mm in the first hour. He was euglycemic; however X-ray chest [Figure 1 inset] revealed features suggestive of a loculated pleural effusion with pleural thickening on the left side. Sputum for acid fast bacilli was negative. He was initially clinically diagnosed to have a tubercular effusion and antitubercular treatment was initiated. The patient however showed no response and guided tapping of the fluid was considered under ultrasonographic (USG) guidance. Ultra sonogram revealed a huge mass in the left pleural cavity abutting the chest wall. A USG-guided fine needle aspiration and trucut biopsy thereafter revealed a cellular but benign spindle cell neoplasm. A contrast-enhanced computerized tomogram demonstrated a large heterogeneous enhancing mass of soft tissue density in the left hemithorax. The mass appeared smooth, well-defined, and lobulated, with no evidence of chest wall or mediastinal invasion, or significant associated mediastinal shift [Figure 1]. The visualized angle of contact with the

pleura on the chest PA radiograph appears obtuse, while on the CT, the angle formed with the lateral chest wall is acute. An obtuse angle suggests that the organ of origin is pleura; however in large, benign masses this angle does not necessarily have to be obtuse. Subsequently the mass was resected via thoracotomy.

Gross examination revealed an encapsulated solid tumor with the bosselated nodular surface weighing 2 kg. The cut section showed a solid fleshy mass with vague whorling measuring 23 × 21 × 09 cm [Figure 2]. The peripheral subcapsular area showed an increased density of blood vessels. Pericapsular adherent lung and pleural tissue was present in some areas.

Microscopy revealed an encapsulated spindle cell neoplasm of varying cellularity [Figure 3]. There were occasional focal areas of ischemic subcapsular hemorrhage and necrosis. The less cellular areas contained spindle cells enmeshed in collagen bundles and the cellular areas comprising cells with round to ovoid nuclei with a fine chromatin pattern and pale eosinophilic cytoplasm. Occasional foci of typical mitosis were seen (maximum number of 03-4/HPF). Perivascular cuffs of hyalinization and areas with increased vascularity were noticed. A sparse sprinkling of lymphomononuclear cells was also observed.

Immunohistochemistry revealed a strong diffuse positivity for CD34 (+++), Vimentin (++) and SMA (++) [Figure 4]. F-VIIIAg showed mild (+) focal staining in some tumor cells. Epithelial membrane antigen and bcl-2 were negative (labvision, prediluted). This confirmed the diagnosis of a solitary fibrous tumor.

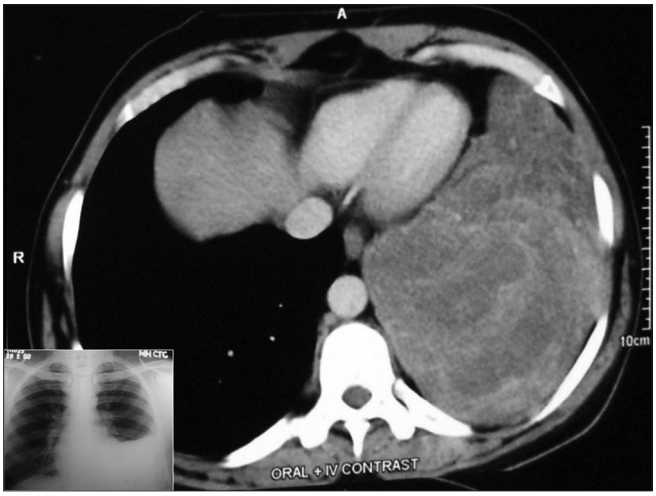

Figure 1: Contrast enhanced computed tomogram showing a huge mass in the left hemithorax. X-ray chest PA view (inset) providing an impression of a loculated pleural effusion

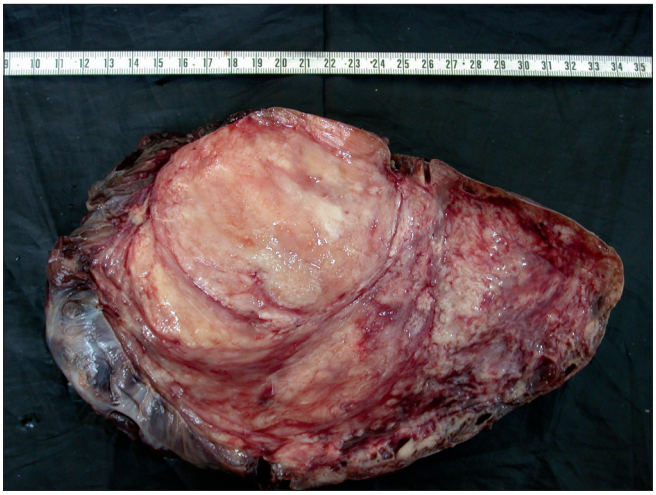

Figure 2: Cut section of the excised tumor revealing an encapsulated solid fleshy mass with whorling

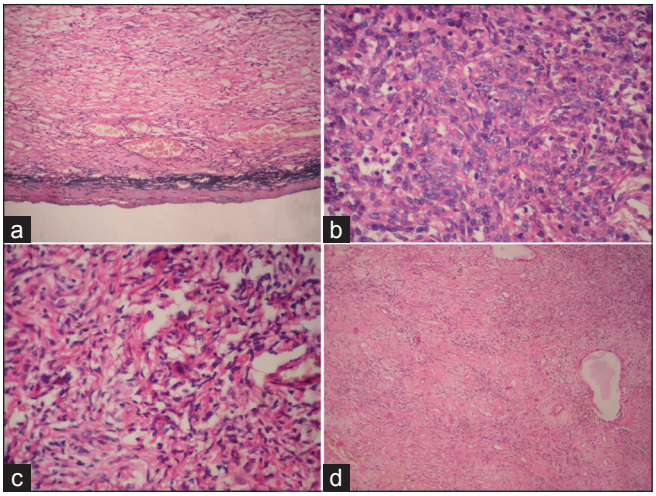

Figure 3: Photomicrographs of the neoplasm showing (a) Subcapsular increased vascularity (H and E, ×200), (b) Cellular areas with relatively plump spindle cells and an occasional mitosis (H and E, ×400), (c) Parvicellular spindle cell areas, (H and E, ×400), (d) Variably hyalinised collagenous stroma, hyaline cuffs around vessels, and microcystic degeneration, (H and E, ×200)

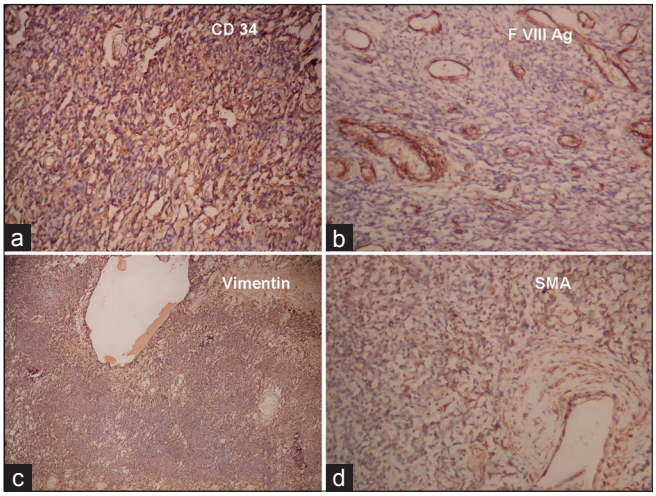

Figure 4: Immunohistochemical profile showing (a, c, d) Diffuse positivity for CD34, vimentin, and SMA. (b) FVIIIAg immunohistochemistry revealed mild-sparse positivity with normal staining of vessels within the tumor. (DAB×200)

DISCUSSION

Solitary fibrous tumors of the pleura (SFTP) are rare primary tumors, histogenesis of which has been a controversial issue. They have been given varying nomenclatures such as localized pleural mesothelioma, pleural fibroma, localized fibrous mesothelioma, submesothelial fibroma, and localized fibrous tumor.[2] However they are now considered to arise from the submesothelial areolar mesenchyme. Immunohistochemistry and electron microscopy have led to these neoplasms being named as solitary fibrous tumors.[3] Cytogenetic data, although limited, have shown various abnormal karyotypes and supernumerary chromosome 8 has suggested malignant behavior of the tumor.[4,5] SFTP occurs in a wide age range (5–87 years) with a fairly equal frequency in both sexes.[6] Most of these arise from the parietal pleura and are exophytic, sometimes attached by a stalk. Occasionally those that are sessile appear to grow inward into the lung parenchyma leading to a clinical suspicion of a malignant mass. Most patients are asymptomatic and come to notice incidently. Our patient had developed atelectasis of the surrounding lung with low to moderate grade fever possibly due to supervening infection. He was suspected to have tuberculosis and was being investigated for the same when radiological imaging beyond an X-ray chest confirmed the mass. These tumors are now known to have accompanying paraneoplastic syndromes. Hypertrophic pulmonary osteoarthropathy (HPO) has been found to be the most common paraneoplastic syndrome in SFTP.[7] Hypertrophic pulmonary osteoarthropathy, possibly caused by ectopic GH like substance, is reported in up to 22% of patients (especially in tumors over 7 cm in diameter) compared to a 5% incidence in lung carcinoma. The patient being reported upon had prominent clubbing. Osseous radiographic changes and the HPO may take months to resolve after tumor resection. Recurrence of symptoms may herald recurrence of the tumor. Severe symptomatic hypoglycemia has also been reported in these patients incidence of which ranges from 3% to 4%.[1,2]

Histologically these exhibit a morphology of a low-grade spindle cell neoplasm.[8] The main differential diagnoses in this case were of an inflammatory myofibroblastic tumor (IMT) and a sclerosing hemangioma (SH). The inflammatory component classical of an IMT was not seen. Focal areas possessing an apparent vascular pattern and microcystic degeneration led to the suspicion of a SH. Immunohistochemistry however confirmed a solitary fibrous tumor.

These tumors prove to be benign in behavior and resection is considered curative with cure rates ranging from 88% to 92%. About 8% of these are reported to recur which has been related to an increased mitosis (more than 4/10 HPF).[9] These tumors may assume alarming proportions; however, the size has been reported to not to have any bearing on its being malignant, resectable or curable.[10]

The case being reported is unique for the purpose of rarity of these neoplasms, occurrence in a relatively young age, and the accompanying HPO. The case also highlights the significance of imaging modalities beyond a simple X-ray chest to avoid an erroneous diagnosis and therapy which could lead to unnecessary increased morbidity for the patient. This would only be possible by keeping a high index of suspicion in an appropriate clinical background.

REFERENCES

1. Cardillo G, Facciolo F, Cavazzana AO, Capece G, Gasparri R, Martelli M. Localized (solitary) fibrous tumors of the pleura: An analysis of 55 patients. Ann Thorac Surg 2000;70:1808-12.
2. De Perrot M, Fischer S, Brundler MA, Sekine Y, Keshavjee S. Solitary fibrous tumors of the pleura. Ann Thorac Surg 2002;74:285-93.
3. Yousem SA, Flynn SD. Intrapulmonary localized fibrous tumor. Intraparenchymal so-called localized fibrous mesothelioma. Am J Clin Pathol 1988;89:365-9.
4. Torabi A, Lele SM, DiMaio D, Pinnt JC, Hess MM, Nelson M, *et al*. Lack of a common or characteristic cytogenetic anomaly in solitary fibrous tumor. Cancer Genet Cytogenet 2008;181:60-64.
5. De Leval L, Defraigne JO, Hermans G, Dôme F, Boniver J, Herens C. Malignant solitary fibrous tumor of the pleura: Report of a case with cytogenetic analysis. Virchows Arch 2003;442:388-92.
6. Shields TW, Yeldandi AV. Localized fibrous tumors of the pleura. In: Shields TW, Locicero III J, Ponn RB, Rusch VW, editors. General thoracic surgery. 6th ed. Philadelphia, PA: Lippincott Williams and Wilkins; 2005:889-900.
7. Briselli M, Mark EJ, Dickersin GR. Solitary fibrous tumors of the pleura: Eight new cases and review of 360 cases in the literature. Cancer 1981;47:2678-89.
8. Granville L, Laga AC, Allen TC, Dishop M, Roggli VL, Churg A, *et al*. Review and update of uncommon primary pleural tumors: A practical approach to diagnosis. Arch Pathol Lab Med 2005;129:1428-1443
9. Corrin B, Nicholson AG, Burke MM. Pathology of the lung. 2nd ed. Philadelphia: Churchill Livingstone Elsevier; 2006:714-16.
10. Bar I, Papiashvilli M, Zukerman B, Stav D, Sandbank J. Large solitary fibrous tumour of the pleura: analysis of six cases. Heart Lung Circ 2007;16:282-4.

How to cite this article: Kaur A, Gill SS, Singh J, Singh A, Mani NS. Giant solitary fibrous tumor pleura: Clinical dilemma and diagnosis. Lung India 2012;29:179-81..

Source of Support: Nil, **Conflict of Interest:** None declared.