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Case Report

A giant solitary fibrous tumor of the pleura: A case report

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Abstract. Solitary Fibrous Tumor of the Pleura (SFPT) are rare neoplasms and can have giant diameters. Immunohistochemical analysis has confirmed that SFPTs originate from the mesenchyme underlying the mesothelial layer of the pleura. SFPTs are usually asymptomatic. However, larger tumors occupying larger spaces in the thoracic cavity present more commonly with symptoms like dyspnea or chest pain. In this case report, we present a giant SFPT in a 71-year-old woman who presented with chest pain on the left side and progressive dyspnea over the previous 4 years.

Key words: Solitary fibrous tümör, pleura, neoplasm

1. Introduction

Solitary Fibrous Tumor of the Pleura (SFPT) is rare neoplasm, and giant SFPTs occupying the entire hemithorax are even more unusual. These tumors frequently originate from visceral pleura and hold on to the pulmonary parenchyma with a pedicle. Radiologically they can appear as a pleural thickness or a space-occupying lesion in the thorax. Peak incidence occurs in the sixth and seventh decades of life and both sexes are affected equally (1). Although most of the cases are asymptomatic, when they reach large sizes, they can lead to symptoms like shortness of breath and chest pain.

2. Case report

A 71-year-old male presented with left sided chest pain and progressive shortness of breath with a history of 4 years. Physical examination revealed decreased breath sounds on the left side. Opacity was present in the middle and lower zones of the left lung in t he Chest radiography (Figure 1). Computed tomography (CT) scan of the chest revealed a mildly hyperdense massive lesion in the left hemithorax with a high density and approximate size of 20x12 cm that displaces the left lung tissue superiorly, and producing mass effect on the mediastinum and diaphragm, and contains calcifications in the posterior portion masscompression (Figure 2,3). Fiberoptic bronchoscopy showed extrinsic compression of the main left bronchus with no endobronchial growth. The positron emission tomography scan we performed revealed low-grade diffuse fluorodeoxyglucose (FDG) uptake in the mass

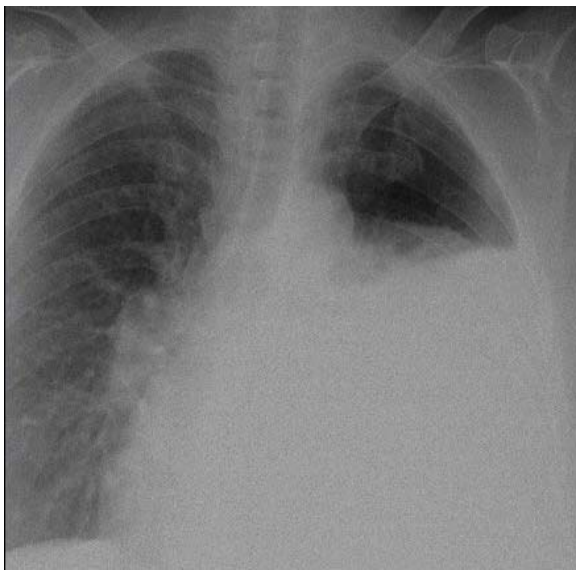

Fig. 1. Chest radiography: Opacity was present in the middle and lower zones of the left lung.

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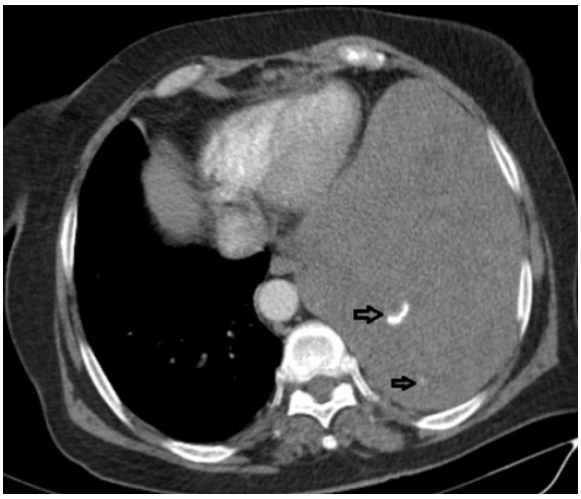

Fig. 2. Computed tomography scan of the chest revealed a mildly hyperdense massive lesion in the left hemithorax with a high density and approximate size of 20x12 cm that displaces the left lung tissue superiorly and contains calcifications in the posterior portion.

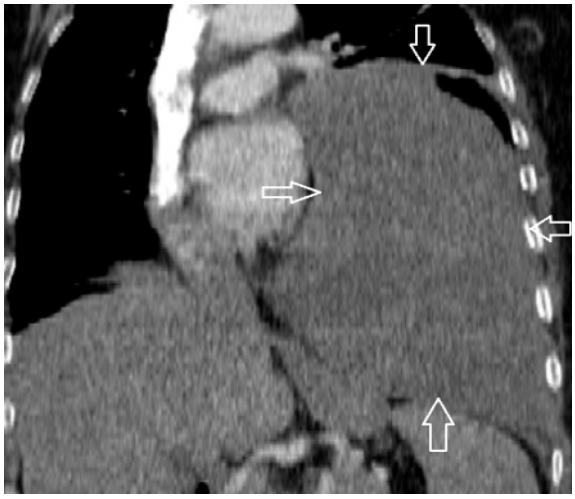

Fig. 3. Computed tomography scan shows a massive tumor occupying most of the left hemithorax, producing mass effect on the mediastinum, diaphragm.

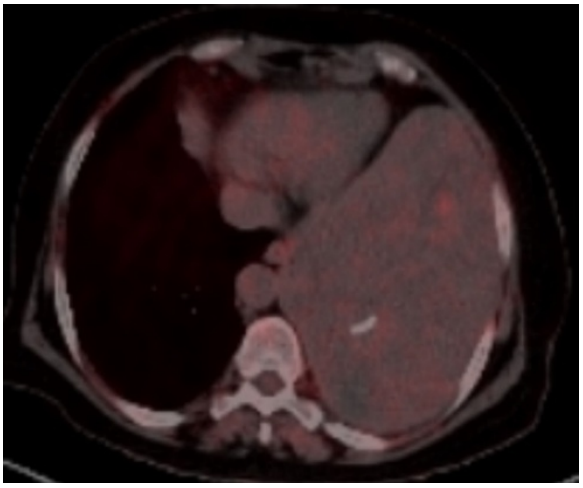

Fig. 4. The positron emission tomography scan revealed low-grade diffuse fluorodeoxyglucose (FDG) uptake in the mass (maximum standardized uptake value/max SUV 2.3).

(maximum standardized uptake value/max SUV 2.3) (Figure 4). The results of the laboratory tests were Hb 11.9 gr/dl, Hct 36.4%, leukocyte 7300/ mm³, thrombocyte 297000 mm³, serum glucose 88 mg/dl and sedimentation 50 mm/hour. A thoracic needle biopsy was performed with the guidance of chess computed tomography. The computed tomography-guided transthoracic biopsy revealed a collagen forming low-grade spindle cell lesion (Figure 5a). The spindle cells were negative for actin, Desmin, S-100. The immunochemical evaluations of all cases were CD34 positive (Figure 5b).

3. Discussion

SFPTs are rare and slow-growing lesions. The diameters range between 1 and 37 cm and they are mostly benign. These tumors frequently originate from visceral pleura and hold on to the pulmonary parenchyma with a pedicle. SFPT often have a silent clinical course over several years. Symptoms develop according to the size and location of the tumors (2). Coughing, chest pain and shortness of breath are the most common symptoms. Chest pain is more common in pleural tumors. Rarely, giant tumors cause bronchial compression, atelectasis symptoms and very rarely hemoptysis. Tumors might also cause signs and symptoms of paraneoplastic syndromes like weight loss, hypoglycemia, shivering or clubbed fingers.

Imaging studies are useful in determining the tumor location and size. Even though the tumor can be incidentally discovered on Chest radiography, CT demonstrate a well-circumscribed, lobular, homogenous, soft-tissue mass (3). However, large tumors are frequently heterogeneous due to necrosis, hemorrhaging or cystic changes. Atelectasis, displacement of adjacent structures and pleural effusion were more common in giant SFPTs. Positron emission tomography is not commonly performed in the workup of fibrous tumors, but a high level of 18-FDG metabolism can indicate a malignant lesion (4,5).

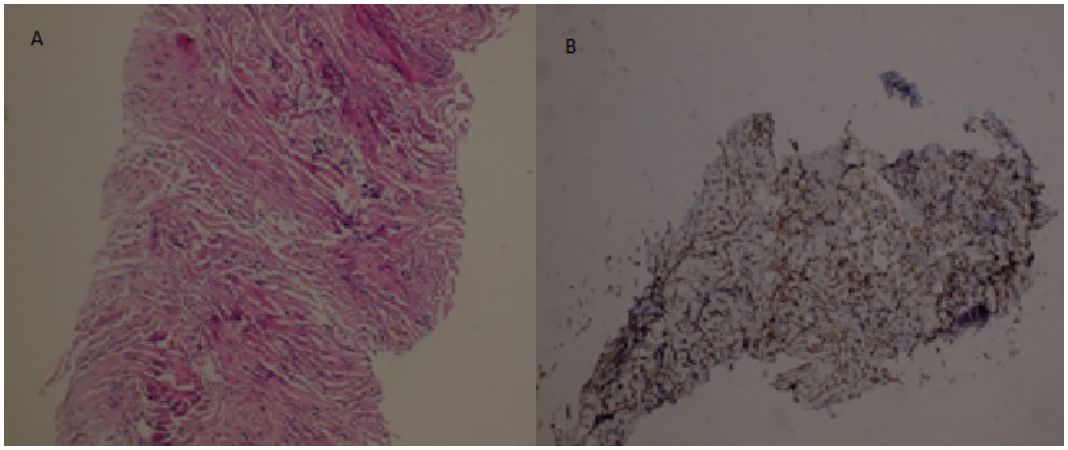

Fig. 5. Microscopic examination of the solitary fibrous tumor of the pleura (SFTP). (A) Microscopic specimen of the tumor shows a collagen forming low-grade spindle cell lesion (hematoxylin & eosin [H&E],X200). (B) Spindle cells show diffuse positivities for immunohistochemical staining with CD34 (DAB ×100).

The definite diagnosis can be made before surgery by obtaining histological material through percutaneous transthoracic needle biopsy or fine needle aspiration. Because the tumor consists of hypercellular and acellular components, in most cases the fine needle aspiration biopsy material is not sufficient to finalize the diagnosis. Sung et al. (6) have shown a 43% success rate of definite diagnosis by fine needle aspiration. Drachenberg et al. (7) reported two cases diagnosed by fine needle aspiration biopsies and Weynand et al. (8) reported 5 cases diagnosed by trucut biopsies. As our patient did not give consent for surgery, the diagnosis was made by fine needle biopsy. Macroscopically, SFPTs are well defined and lobulated and morphologically they are defined as patternless and contain fusiform cells rich in collagen (9). Most of these tumors are benign and the criteria suggested by WHO (more than 4 mitoses in 10x microscopy, the presence of necrosis, pleomorphism, significant cellularity) are important in the identification of malignity (10). Histochemical studies reveal Bcl-2 and CD 34 positivity and S100, cytokeratin, actin, desmin negativity. Positive immunoreactivity for CD34 has been used as a diagnostic marker for SFPT, and loss of CD34 immunoreactivity in high-grade CTs following malignant transformation has been also described (11). Bcl-2 and CD34 positivity and S100, actin and desmin negativity was identified in our patient. The treatment recommended for SFPT is surgery. The purpose is to remove the mass completely with negative surgical borders and minimal parenchymal tissue.

As a result, solitary Fibrous Tumor of the Pleura are rare and generally benign tumors and they should be considered in the differential diagnosis of massive lesions of the thorax.

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