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Case Report

Cinematic rendering of pleural solitary fibrous tumor ☆

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ABSTRACT

Solitary fibrous tumors are rare and typically benign fibroblastic neoplasms with a mean age of onset ranging from 60 to 70 years. Solitary fibrous tumors may arise anywhere within the body, however the pleura is the most common site of origin for these tumors, with approximately 30% of tumors arising from the pleura. In this report we highlight the case of a 62-year-old woman who presented with gradually progressive left sided chest pain that was eventually diagnosed as a pleural SFT. We highlight the appearance of solitary fibrous tumor of the pleura on computed tomography and positron emission tomography imaging, as well as providing detailed cinematic rendering images of these rare neoplasms.

Introduction

Solitary fibrous tumors of the pleura (SFTP) are typically benign fibrotic mesenchymal neoplasms. SFTPs are rare tumors, accounting for only 5% of all pleural tumors [1]. The mean age of presentation of SFTP is between 60 and 70 years of age with no sex predilection [1]. While typically benign, up to 30% of SFTPs can harbor malignant histological characteristics and recur following complete resection [2]. Given how SFTPs are often incidentally detected, cross sectional imaging is essential in detection, characterization as well as risk stratification of these tumors [3]. In this report we highlight the case of a 62-year-old woman who presented with gradually progressive left sided chest pain that was eventually diagnosed as a SFTP.

Case report

A 62-year-old woman presented with gradually progressive left sided chest pain. The patient denied fevers, night sweats, cough, or hemoptysis. Subsequent outpatient computed tomography (CT) imaging revealed a large intrathoracic lung

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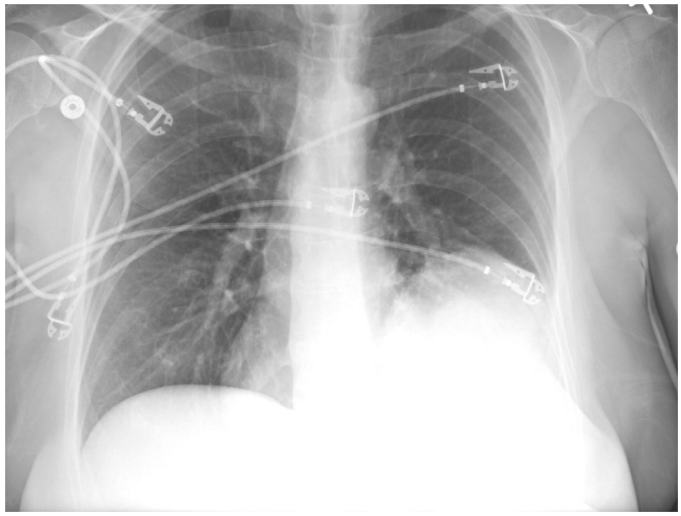

Fig. 1 – A 62-year-old woman with chest pain. Anteroposterior chest X-ray demonstrates mass inseparable from left hemidiaphragm and cardiac border (arrow). No evidence of adenopathy is seen.

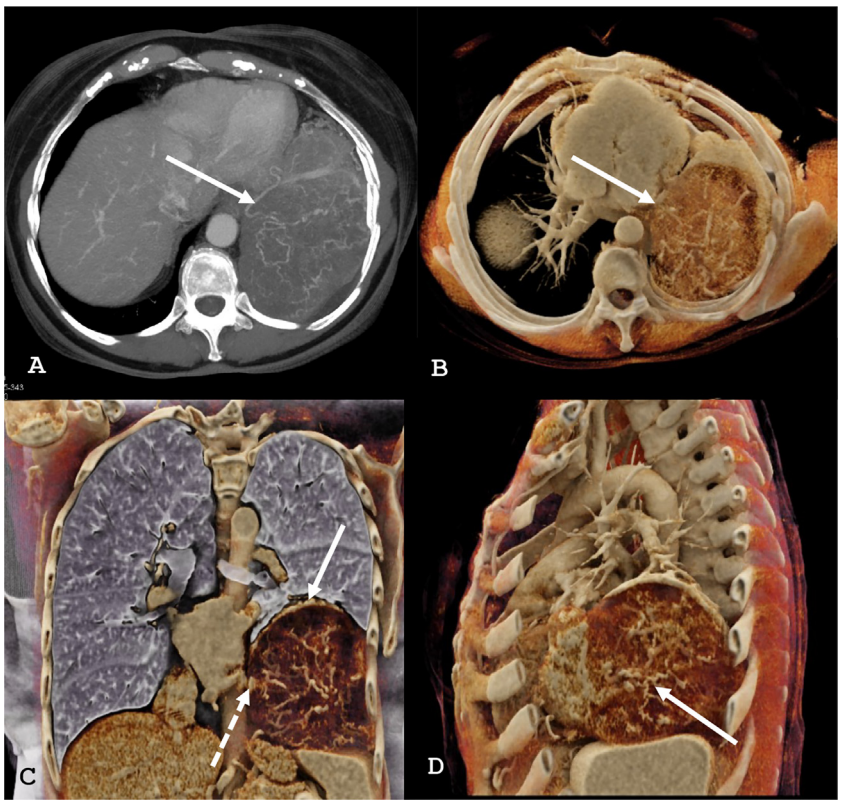

Fig. 2 – A 62-year-old woman with chest pain. (A) Contrast enhanced axial arterial phase image demonstrates a heterogeneously hypodense 10.8cm mass lesion in the lower lobe of the left lung (arrow). (B) Axial cinematic rendered IV-contrast enhanced arterial phase image improves visualization of hypervascular lung mass (arrow). Cinematic rendered IV-contrast enhanced arterial phase image in the coronal (C) plane demonstrates feeding vessels arising from the subdiaphragmatic aorta (dotted arrow) and mass effect resulting in inversion of the left hemi diaphragm (arrow). Cinematic rendered IV-contrast enhanced arterial phase image in the sagittal (D) plane redemonstrates mass and highlights its impressive internal neovascularity (arrow).

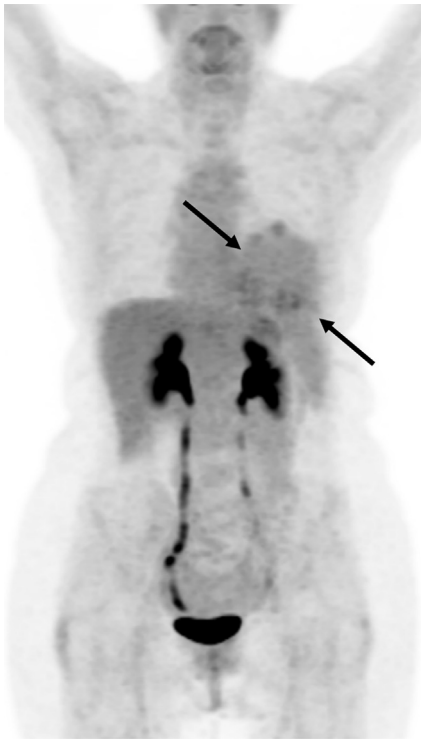

Fig. 3 – A 62-year-old woman with chest pain. Coronal FDG positron emission tomography images acquired following administration of intravenous FDG tracer demonstrates large left lower lung mass with mild to moderate heterogeneous FDG avidity (arrow).

mass prompting referral to our hospital for further workup and management. Upon referral, chest X-ray showed a mass inseparable from left hemidiaphragm and cardiac border (Fig. 1). Subsequent contrast enhanced CT demonstrated a 10.8 cm hypervascular mass lesion in the lower lobe of the left lung (Fig. 2). The mass exerted significant mass effect on the adjacent structures resulting in inversion of the left hemidiaphragm and left lower lobe atelectasis. Positron emission tomography demonstrated the mass to have mild to moderate heterogeneous FDG uptake, favoring a low-grade and/or non-malignant neoplasm (Fig. 3). A needle biopsy was performed on the mass and immunohistochemistry analysis of the specimen revealed tumor cells that were strongly and diffusely positive for STAT6; a finding most consistent with solitary fibrous tumor (SFT). The patient subsequently underwent a left thoracotomy with complete resection of the tumor. Intraoperative evaluation and histopathological analysis of the surgical specimen demonstrated a pleural origin of the SFT.

Discussion

Solitary fibrous tumors (SFT) are a spectrum of mesenchymal tumors presumed to be of fibroblastic differentiation. First described in 1931, these tumors were previously believed to exclusively involve the pleura, however it has since been established that these tumors may arise from virtually anywhere within the body [4,5]. The pleura, however, remains the most common site of origin for SFTs, accounting for approximately 30% of all cases [6]. Other commonly involved sites are the meninges and the abdominal cavity, accounting for 27% and 20% of cases, respectively [7]. SFTP have no sex predilection and typically arise in adults with a peak incidence in the 5th and 6th decade of life [8]. Overall SFTPs are rare tumors, with an annual incidence rate of 2.8 per 100,000, with SFTPs accounting for only 5% of all pleural tumors [1].

Clinically, pleural SFTs are usually asymptomatic and discovered as incidental findings [9]. Symptomatic pleural SFTs may present with localizing intrathoracic symptoms such as chest pain, as present in our case, as well as dyspnea and hemoptysis [10]. Nonspecific systemic symptoms such as fever, weight loss and fatigue may also be present in a smaller subset of patients [10]. Associated paraneoplastic syndromes resulting in symptomatic hypoglycemia (Doege-Potter syndrome) and hypertrophic pulmonary osteoarthropathy may also be accompanying manifestations in 5% and 20% of patients, respectively [11,12].

On CT imaging, pleural SFTs typically present as well-defined, predominantly hypervascular masses. On unenhanced images, these tumors can be hypodense or hyperdense to adjacent muscle depending on intratumoral collagen content [13]. Following intravenous contrast administration, SFTs display marked heterogenous enhancement [13]. The presence of nonenhancing areas may indicate necrosis, intratumoral hemorrhage or myxoid degeneration [14]. Up to 96% of SFTPs form at least 1 acute angle with adjacent pleura, with 65% forming only acute angles [10]. These lesions may therefore appear as a subpleural pulmonary mass and may be misdiagnosed as peripheral lung cancer [10,14]. However, rib erosion, pleural effusion and calcification, are characteristically absent, aiding in differentiation from more aggressive malignancies. Differential considerations for very well defined pleural SFTs consist of pleural lipoma and intercostal nerve schwannoma. For SFTs that are not well defined, differential diagnoses consist of peripheral lung cancer, intrapleural sarcoma, localized mesothelioma, solitary pleural metastasis and organized inflammation.

Pathological diagnosis of SFTP is established based on histological evaluation and immunohistochemistry (IHC) [15]. IHC markers CD34 and STAT6 strongly support the diagnosis of SFT; STAT6 was positive in our case [15]. In equivocal cases, testing for the NAB2-STAT6 fusion gene, which has recently emerged as the most specific marker of SFTs, can help confirm diagnosis [15].

While the majority of SFTPs are benign, malignant features may be detected at pathological exam in approximately 30% of patients with 8% of patients experiencing tumor recurrence following complete (R0) resection [2]. In cohorts of patients with malignant SFTPs, these recurrence rates have been demonstrated to be as high as 30% [16]. Five-year survival rates following resection range from 83% to 92% [2]. CT features have been demonstrated to be predictive of malignancy, with tumor size $\geq$10cm, hypervascularity and presence of a pleural effusion being the most significant factors indicating malignant histology [2,3]. Risk of metastases may also be predicted; at our institution, we use a modified 4-variable

risk stratification model to predict risk of metastatic disease in SFTs that includes patient age (<55, ≥55), tumor size (<5 cm, 5 cm to <10 cm, 10 cm to <15 cm, ≥15 cm), mitotic count (0, 1-3, ≥4), and presence of tumor necrosis ≥10% [17].

Surgical resection remains first line treatment of choice for SFTP regardless of histological characteristics [2]. Due to the highly vascular nature of these tumors, preoperative embolization may also be considered. Novel antiangiogenic agents have shown promise as first-line therapies, but research in this area is still ongoing [18]. Given the moderate risk of recurrence, long term surveillance is recommended. While further work is needed to establish guidelines governing surveillance methodology and duration, up to 23% of patients experienced first recurrence >5 years following resection [2]. Multidisciplinary discussions regarding long term surveillance may therefore be warranted on a case-by-case basis.

Patient consent

The patient reported in the manuscript signed the informed consent/authorization for the participation in the research, which includes the permission to use data collected in future research projects including presented case details and images used in this manuscript.

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