

An unexpected diagnosis: Solitary fibrous tumor of the pleura revealed during appendicitis workup

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Background

Solitary Fibrous Tumor of the Pleura (SFTP) is a rare, slow-growing mesenchymal neoplasm that accounts for approximately 5% of all pleural tumors, affecting both men and women [1]. The estimated incidence is 2.8 cases per 100,000 hospitalized patients, with only 800 reported cases documented between 1931 and 2002 [2]. However, the number estimated to be increased. While solitary fibrous tumors can occur in various body parts, they are most commonly found in body cavities such as the pleura, peritoneum, and meninges. Among these, the pleura is the most frequent site, with over 80% of cases originating from the visceral pleura and about 20% from the parietal pleura [3]. Although SFTP is usually benign, 10-30% of cases exhibit malignant potential, characterized by aggressive local behavior and, in some instances, metastasis [1]. This case report highlights a unique case of a giant solitary fibrous tumor of the pleura incidentally identified during the workup for appendicitis and its successful management through surgical resection.

Case presentation

A 33-year-old female with a past medical history of alcohol use disorder, and a non-smoker, presented to the emergency department with right lower abdominal pain for the past two days. She described the pain as acute in onset, progressive in

nature, dull-aching, 9/10 in severity, and radiating to the umbilicus, with no aggravating factors. The pain was associated with one episode of subjective low-grade fever and chills. She also complained of a productive cough with yellowish sputum, occasionally blood-tinged, for the past month, for which she had been prescribed antibiotics. However, her symptoms persisted, and she did not seek further medical intervention. Additionally, she reported exertional shortness of breath and described her exercise tolerance being reduced to seven blocks due to dyspnea over the past month. She denied any history of nausea, vomiting, chest pain, dizziness, palpitations, diarrhea, constipation, weight loss, or other symptoms.

Initial vital signs revealed tachycardia at 102 beats per minute (bpm), blood pressure of 145/85 mm Hg, oxygen saturation of 97% on room air, respiratory rate of 16 breaths per minute, and a temperature of 99.7°F. On physical examination, she was alert and oriented but in mild distress due to abdominal pain, with no cyanosis or peripheral edema. Palpation of the abdomen revealed firmness with rebound tenderness in the right lower quadrant and periumbilical area. Auscultation showed air entry present bilaterally with reduced vesicular breath sounds in the right lower lung zone. Cardiac examination revealed normal heart sounds without any added murmurs, gallops, or friction rubs. Slightly increased bowel sounds were also noted.

Initial laboratory tests, including a Complete Blood Count (CBC), revealed leukocytosis with a white blood cell count of 12,000/ μ L and neutrophil predominance. Hemoglobin was 13.6 g/dL, and platelets were 354,000/ μ L. Kidney function and electrolytes were within normal limits. However, liver function tests identified mild transaminitis, with Alanine Transaminase (ALT) of 280 U/L, Aspartate Transaminase (AST) of 218 U/L, and alkaline phosphatase of 193 U/L.

Imaging studies, including a Computed Tomography (CT) scan of the abdomen and pelvis, revealed early features of acute appendicitis along with a mass in the right lower thoracic cavity. A chest X-ray showed a moderately elevated right hemidiaphragm with right intrahilar/basilar atelectasis or consolidation. A subsequent CT chest scan revealed a 15 cm heterogeneous mass in the right lower thoracic cavity, displacing the Inferior Vena Cava (IVC), with findings concerning for malignancy. A CT angiography of the chest suggested a large enhancing heterogeneous mass with regions of decreased enhancement, suggestive of necrosis, extending from the right hemidiaphragm into the right intrahilar region. These findings were concerning for primary lung/pleural carcinoma or bronchoalveolar carcinoma. Consequently, the patient was admitted with a diagnosis of acute appendicitis, and further evaluation of the chest mass was initiated.

Surgery consultation was sought, and conservative management with antibiotics was recommended for acute appendicitis. Pulmonary medicine consultation was requested, and after discussions with thoracic surgery and radiology, a CT-guided biopsy of the lung mass was performed. Pathological analysis of the biopsy revealed a solitary fibrous tumor of pleura, with immunohistochemical stains positive for STAT-6 and CD34 and negative for CAM5.2, supporting the diagnosis. A Mucicarmine special stain was negative for mucin.

Subsequently, oncology was involved in the case and advised further testing and referral to cardiothoracic surgery for possible resection of the mass. Further imaging, including Positron Emission Tomography (PET-CT), demonstrated a mildly hypermetabolic large mass in the right lower lobe of the lung, highly suspicious for low-grade malignancy, with no other hypermetabolic activity noted. An MRI of the brain revealed no abnormalities. An echocardiogram showed a left ventricular ejection fraction of 65%, with no evidence of diastolic dysfunction or valvular abnormalities. Pulmonary function tests revealed reduced FEV1 and FVC, with a preserved FEV1/FVC ratio and normal Diffusing capacity of the Lungs for Carbon monoxide (DLCO).

The patient subsequently underwent a right thoracotomy for resection of the right pleural mass, along with cryoablation of the intercostal nerves. She tolerated the procedure well and was discharged with instructions for close follow-up in the outpatient clinic.

Discussion

Solitary Fibrous Tumors of the Pleura (SFTP) are rare, slow growing neoplasm often present asymptotically, posing a significant diagnostic challenge. When symptomatic, they manifest with non-specific complaints such as cough, dyspnea, or chest discomfort, which may easily be misattributed to other common conditions until the tumor reaches a detectable size on imaging [4]. In rare cases, SFTP may present with paraneoplastic syndromes, including pulmonary hypertrophic osteoarthropathy, digital clubbing, or hypoglycemia caused by the se-

cretion of insulin-like growth factors [5].

Imaging plays a pivotal role in the evaluation of pleural lesions, with chest CT being the modality of choice for detailed characterization of the tumor, its size, and its relationship to surrounding structures [6]. While chest X-rays may reveal a well-defined thoracic mass, CT scans provide superior resolution, guiding further management. MRI, though of limited utility, can help delineate soft tissue involvement, while CT angiography is crucial for assessing vascular supply, particularly when planning biopsies for highly vascular lesions to mitigate the risk of bleeding. PET-CT may be employed to evaluate the tumor's hypermetabolic activity, though it cannot reliably differentiate benign from malignant SFTP due to potential false-positive results [7,8].

Definitive diagnosis of SFTP relies on histopathological and immunohistochemical analysis. The hallmark markers include STAT6 and CD34 positivity, which are seen in approximately 98% of cases, while EMA and S100 are typically negative [9,10]. Histological features indicative of malignancy includes dense cellularity, nuclear hyperchromasia, pleomorphism, increased mitotic activity (>4 mitotic figures per 10 high-power fields), necrosis, and hemorrhage, which were absent in our case indicating benign pathology [11].

Surgical resection with negative margins remains the cornerstone of treatment for SFTP, regardless of its benign or malignant nature. For benign tumors, the 5-year survival rate approaches 100% [12,13]. Despite this favorable prognosis, recurrence remains a concern, with most recurrences occurring within two years of resection. As such, close postoperative surveillance is essential, involving biannual chest X-rays and CT scans for the first two years, followed by annual CT imaging thereafter [14]. In cases of recurrence, radical surgical resection is advised [15].

In our case, the patient initially presented with abdominal pain related to acute appendicitis, and during the evaluation, an incidental pleural mass was identified on abdominal CT. Subsequent imaging with a chest CT confirmed the presence of a thoracic lesion, and PET-CT revealed mild hypermetabolic activity, suggesting a lower malignant potential. Immunohistochemical analysis showed positivity for STAT6 and CD34, with negativity for CAM5.2, supporting the diagnosis. Histopathological findings were consistent with a benign pathology. The patient underwent complete surgical resection of the mass, and the initial postoperative follow-up demonstrated no signs of recurrence. To mitigate the risk of relapse, the patient is being closely monitored with biannual CT scans as per current guidelines.

Conclusion

This case of an incidental solitary fibrous tumor of the pleura emphasizes the importance of thorough history-taking, attention to subtle symptoms, detailed physical examination, and a broad differential diagnosis. Multidisciplinary involvement is crucial for accurate diagnosis. Complete en bloc surgical resection ensures favorable outcomes, and vigilant follow-up with serial imaging is essential to monitor for recurrence.

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