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Mediastinal Solitary Fibrous Tumor Diagnosed by Endobronchial Ultrasound-Directed Biopsy

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 32
Final Diagnosis: Mediastinal solitary fibrous tumor
Symptoms: Cough • dyspnea • progressive dysphagia • worsening intermittent chest pain
Medication: —
Clinical Procedure: Flexible bronchoscopy • endobronchial ultrasound (EBUS)
Specialty: Pulmonology

Objective: Rare disease

Background: Solitary fibrous tumors of the middle mediastinal space are uncommon and often not discovered until symptoms secondary to compression of adjacent structures occur. Diagnosis requires surgical biopsy and histological tissue analysis. We describe the ECHO appearance of the solitary fibrous tumor and successful non-invasive EBUS diagnosis. This method of diagnosis allowed for surgical planning for resection and allowed us to exclude non-surgical diseases, such as small cell carcinoma.

Case Report: A 32-year-old man presented to his primary care physician with worsening intermittent chronic chest pain with recent progressive dysphagia, cough, and dyspnea. Physical examination and routine laboratory work-up were unrevealing. Chest radiograph and computed tomography (CT) of the chest revealed a middle mediastinal mass. Flexible bronchoscopy confirmed extrinsic compression of right and left bronchial trees. Endobronchial ultrasound (EBUS) was used to biopsy the mass and the diagnosis of solitary fibrous tumor was confirmed. The patient underwent successful tumor resection and was discharged home after an uneventful postoperative period.

Conclusions: Endobronchial ultrasound-directed tissue biopsy is an appropriate modality for suspected solitary fibrous tumors of the mediastinum. To our knowledge, this is only the second reported case of SFT diagnosed by EBUS-TBNA. Our case uniquely demonstrates the advantages of pre-surgical diagnosis of mediastinal masses with EBUS-TBNA when the diagnosis SFT is suggested on CT and US imaging.

MeSH Keywords: Biopsy, Fine-Needle • Bronchoscopes • Mediastinal Neoplasms • Solitary Fibrous Tumors • Ultrasonography

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/903680

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Background

Mediastinal solitary fibrous tumors (SFT) are uncommon, slowly growing neoplasms typically diagnosed via surgical biopsy. The presenting symptoms associated with SFTs are usually nonspecific, such as cough, dyspnea, and chest pain [1]. Symptoms can also relate to the tumor location and the compressive effects on surrounding structures. SFTs typically have low malignant potential and surgical resection can be curative in most cases [2]. SFTs are most commonly identified by cross-sectional imaging and have an appearance that is not reliably distinguishable from other soft tissue mediastinal tumors. In the absence of clearly pathognomonic imaging characteristics, confirmatory tissue diagnosis is required prior to resection.

The surgical approach for diagnosis can be challenging due to the close proximity of surrounding vascular structures. Endobronchial ultrasound bronchoscopy (EBUS) allows for real-time, minimally invasive fine-needle aspiration of mediastinal masses, and typically has high diagnostic yield. We report the ultrasound characteristics and successful diagnosis of a middle mediastinal solitary fibrous tumor using EBUS-guided needle aspiration. To our knowledge, this is only the second reported case of SFT diagnosed with EBUS-TBNA. The prior case report used EBUS in the diagnosis of recurrent SFTs [3]. Our case illustrates the potential of used EBUS in the primary diagnosis of mediastinal masses when the diagnosis of SFT is suggested by CT and US imaging.

Case Report

A previously healthy 32-year-old man presented to his primary care physician with worsening intermittent chest pain for 6 months. During the last month, he had developed progressive dysphagia, cough, and dyspnea. He denied constitutional symptoms. He had smoked cigarettes for the past 16 years. There was no family history of malignancy or cardiovascular disease.

He was referred to his local emergency department for evaluation. Physical examination and routine laboratory work-up were unrevealing. A chest radiograph showed indeterminate mediastinal opacity. A CT of the chest revealed a large heterogeneous middle mediastinal mass measuring 15×17×18 cm, without associated lymphadenopathy (Figure 1). There was anterior displacement of the heart and right pulmonary artery, with compression of the esophagus and adjacent main bronchi.

The patient underwent flexible bronchoscopy, which confirmed extrinsic compression of right and left bronchial trees. Endobronchial ultrasound (EBUS) revealed a large, heterogeneous, cystic mediastinal mass, which was sampled using a 22G needle (Figure 2). Gross examination of the excised specimen revealed a large tan-red to yellow lesion that was multicystic and partially necrotic,

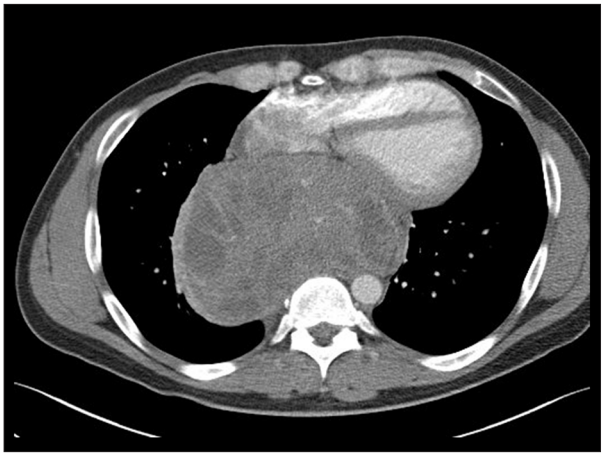

Figure 1. Computed tomography of the chest revealed a large heterogeneous middle mediastinal mass measuring 15×17×18 cm without associated lymphadenopathy and anteriorly displaced heart.

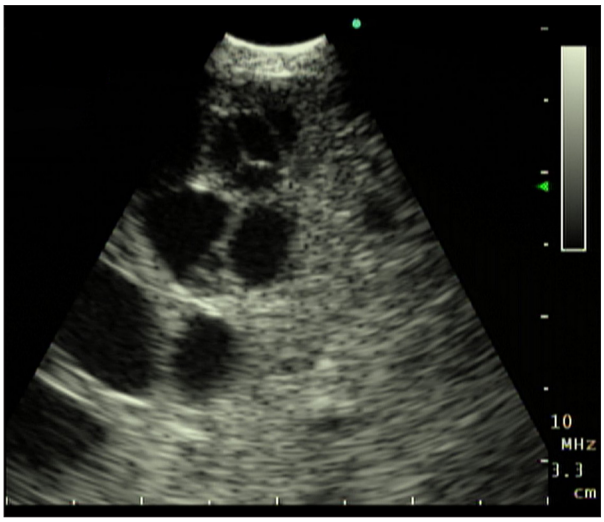

Figure 2. Endobronchial ultrasound (EBUS) revealed a large, heterogeneous, cystic mediastinal mass.

weighing 910 grams, and measuring 17.5×15.0×8.0 cm (Figure 3). Histologically, the lesion was confirmed to be a solitary fibrous tumor with closely packed spindle cells with mitotic figures (Figure 4). Immunostaining of cells revealed staining positive for CD34 and negative for S-100 and keratin, consistent with SFT. The tumor was classified as high risk based on size, patient age, and number of mitotic figures per high-power field. The patient underwent successful tumor resection. Postoperatively, the patient recovered well and was discharged home.

Discussion

Solitary fibrous tumors (SFT) are a rare heterogeneous group of benign and malignant neoplasms of mesenchymal origin.

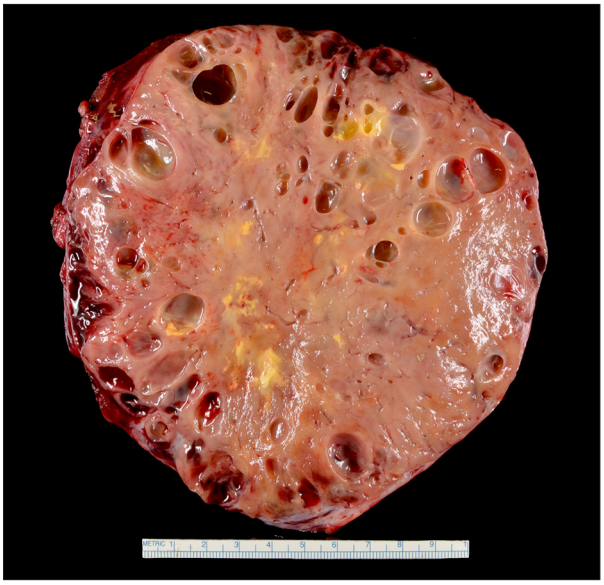

Figure 3. Excised specimen of a large multicystic, tan-red to yellow lesion that was partially necrotic, weighing 910 grams, and measuring 17.5×15.0×8.0 cm.

Most SFT are benign, occur both at intrathoracic and extrathoracic locations, and are most commonly diagnosed between the ages of 50 and 70 [4,5]. Our patient was relatively young at the time of diagnosis. SFT are typically asymptomatic and presenting symptoms often occur secondary to compression of adjacent structures, as observed in our case. Rare paraneoplastic syndromes have been reported with large tumors. Hypertrophic pulmonary osteoarthropathy (Pierre-Marie-Bamberger syndrome) has been described in 10–20% of patients [6,7]. In rare cases, hypoglycemia can occur secondary to secretion of insulin-like growth factor-2 [7,8].

Imaging studies include plain radiography, CT, and MRI. MR angiography and Doppler ultrasonography have been proposed as diagnostic tools due to the highly vascular nature of these tumors [9]. Definitive diagnosis requires histological analysis, usually via an invasive surgical approach. Diagnosis with endobronchial ultrasound-directed transbronchial needle aspiration (TBNA) allows for both real-time visualization and biopsy. Our patient's imaging (CT and ultrasound) were consistent in revealing a large multicystic mass, and biopsy confirmed a diagnosis of SFT prior to surgical resection. This early diagnosis enabled additional time for surgical planning for resection and eliminated non-operative differential diagnoses prior to surgery. While EBUS-TBNA can be a valuable tool in the diagnosis of mediastinal tumors, tissue sampling occurs via an aspiration of cells rather than a core biopsy. The cytologic nature of this can be a limitation in terms of overall diagnostic yield.

Microscopically, SFT demonstrates closely packed spindle cells, prominent vascular channels, and zones of hypercellular, hypocellular, and fibrotic areas [6,9]. Immunostaining of SFT is not specific, but is usually positive for CD34, CD99, and B cell lymphoma 2 (Bcl-2), and negative for S100 and cytokeratins. Recent studies indicate that SFT has a NAB2-STAT6 fusion gene and nuclear STAT6 immunoreactivity, which may prove to be useful in diagnosis of SFT [4,10,11].

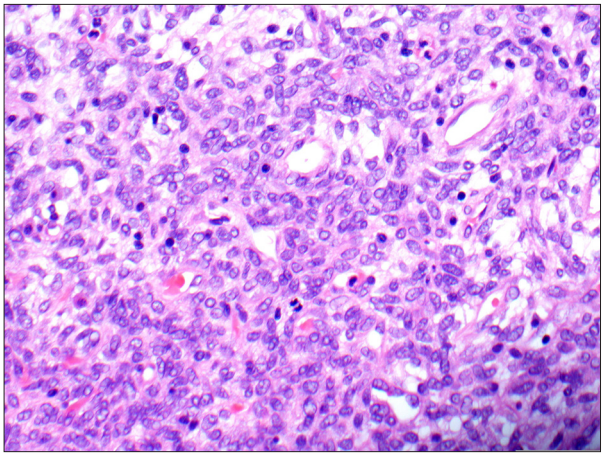

Figure 4. Histological examination of the lesion showing closely packed spindle cells with mitotic figures consistent with a solitary fibrous tumor.

Definitive treatment is surgical resection. Thirty percent of patients have a reoccurrence after surgical resection [12]. Continued long-term follow-up with serial cross-sectional imaging is recommended, as late reoccurrence has been reported (>10 years) [13]. Prognosis is calculated using a risk stratification model. Factors associated with an increased risk of metastasis and poor outcome include: larger tumor size (≥15 cm), increased mitotic figures (≥4/10 high-power fields), and advanced patient age (≥55 years). Five-year survival for patients undergoing complete resection is 89–100% [14].

Conclusions

Endobronchial ultrasound-directed tissue sampling is an appropriate modality for suspected solitary fibrous tumors of the mediastinum and can be used to obtain adequate diagnostic tissue specimens. To our knowledge, this is only the second reported case of SFT diagnosed by EBUS-TBNA. Our case demonstrates the advantages of pre-surgical diagnosis of mediastinal masses with EBUS-TBNA when the diagnosis of SFT is suggested by CT and US imaging.

Conflict of interest

None.

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