Solitary fibrous pleural tumors are rare masses of mesenchymal origin that may be mistaken for mesothelioma. A positive staining of vimentin, negative staining of cytoplasmic keratin, and expression of the CD34 antigen can confirm the presence of a solitary fibrous pleural tumor. Although most tumors of this type are benign, they possess a malignant potential and thus should be excised. We report a case of a 63-year-old man who had an inconclusive biopsy of a lung lesion 15 years ago. Further testing after excision revealed a solitary fibrous pleural tumor. A brief discussion of the clinical presentation and incidence of these tumors is included.

Solitary fibrous tumors of the pleura were first described in 1931.1 One review of the literature reported that since then, only 800 cases of such tumors have been reported.2 Because solitary fibrous pleural tumors were first believed to originate from mesothelial cells, they were considered localized mesotheliomas. However, findings from immunohistochemical staining have revealed that these masses originate from mesenchymal tissue.3

Solitary fibrous pleural tumors typically stain positive for vimentin, a marker of mesenchymal cells, and negative for cytoplasmic keratins, which are found in mesothelioma.3-5 The CD34 antigen, which exists in normal and neoplastic endothelial cells, is present in nearly all occurrences of these tumors, whether they are benign or malignant.6 However, the antigen is widely expressed in mesenchymal neoplasms and is therefore not specific to these tumors. Nonetheless, CD34 expression is extremely useful because it is consistently absent in malignant mesothelioma.7

Vimentin, cytoplasmic keratin, and CD34 expression are therefore excellent markers for determining whether a mass is a solitary fibrous pleural tumor or mesothelioma. In the present report, we describe a man whose tumor, on excision, was determined to be a solitary fibrous pleural tumor.

Report of Case
A 63-year-old man presented to his family physician with increasing fatigue, dyspnea, and a mildly productive cough during the past 6 months. On physical examination, the patient was normotensive and afebrile with a regular heart rate and rhythm. His respirations were unlabored. He appeared healthy overall but was concerned with the progressive worsening of his symptoms.

The patient had a 45-year history of smoking cigarettes (1 pack/d) and denied any exposure to asbestos or other hazardous materials. Physical examination further revealed decreased breath sounds at the right lung base. No finger or toe clubbing or cyanosis was present. The patient’s medical history revealed that approximately 15 years earlier, a needle biopsy of a lung lesion guided by computed tomography (CT) suggested the presence of benign cystic cells. However, no additional follow-up testing was completed.

A chest radiograph showed an ill-defined mass of mediastinal opacity—or a consolidated infiltrate—on the right in the lower lobe of the lung. A CT scan of the chest revealed a smoothly margined, elliptical, abnormal mass in the retrocardiac portion of the right mid- to lower hemithorax approximately 6 cm in diameter with a density greater than that of simple fluid and with no substantial enhancement (Figure 1).

On thoracic surgical consultation, surgical excision was recommended. A posterior lateral thoracotomy of the right lung revealed a 6-cm mass attached to the lung pleura via a pedunculated stalk in the posterior segment of the lower lobe on that side of the lung. The mass appeared to be a solid tumor and was beefy red with a firm, fibrous texture. The tumor was resected from lung margins, and a frozen section showed marked fibrous changes and the appearance of a benign tumor.

Pathology reports revealed positive staining for vimentin, antiapoptotic protein Bcl-2, and antigens CD34 and CD99. The tumor stained negative for keratin markers, including hard keratin and cytokeratin; epithelial membrane markers; desmin; and smooth muscle actin. The tumor itself did not stain positive for factor VIII and had no evidence of adjacent
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structure infiltration, high mitotic activity, or substantial nuclear pleomorphism. As such, the tumor was determined to be a benign solitary fibrous pleural tumor of visceral origin. The lung expanded on the right side postoperatively, and the patient had an uneventful recovery. He was encouraged to stop cigarette smoking and to use incentive spirometry to minimize risk of postoperative atelectasis. The patient was discharged from the hospital and received physical therapy and supplemental oxygen through a home healthcare regimen. Postoperative osteopathic manipulative treatment, focusing on indirect muscle-energy and low-velocity, low-amplitude techniques in the direction of least resistance of the displaced right thoracic ribs 4 through 7, was added to augment the patient’s recovery. The patient returns regularly for follow-up with the thoracic surgeon (A.H.O.) to confirm that the tumor has not recurred.

Comment
Mesenchymal cells in areolar tissue next to the pleura encourage the growth of solitary fibrous pleural tumors. More than half of all patients with such tumors are asymptomatic, and 80% of such masses are benign. Extrinsic compression of the solitary fibrous pleural tumor on lung parenchyma may cause patients to seek medical care with the common symptoms of cough, dyspnea, and chest pain. These complaints, along with hypertrophic pulmonary osteoarthropathy, are found in more than 33% of symptomatic patients. An alternate patient presentation may be tumor-associated hypoglycemia, which is caused when the tumor produces insulin-like growth factors. This particular symptomatology is seen in the paraneoplastic phenomenon referred to as Doege-Potter syndrome.

Eighty percent of solitary fibrous pleural tumors are of visceral origin. The other 20% originate in the parietal pleura. Only one case of a solitary fibrous pleural tumor reported in the medical literature has suggested a genetic component. However, it is possible that this familial case may have resulted from exposure to a common environmental agent or a germ-line mutation that was genetically transmitted.

Histologic evaluation usually discloses cellular areas with intermittent hyalinized or necrotic areas, while electron microscopy reveals both fibroblasts and mesothelial cells. Findings from CT scans depend largely on the location, size, and histologic features of the tumor. However, such lesions typically are well defined with clear margins and smooth contours. In addition, they usually have homogenous attenuation and are at right or obtuse angles with the pleura. Larger lesions have been noted to have more lobulated contours and “geographic” patterns. Larger lesions are also most often at acute angles with the pleura.

The size of the tumors can vary greatly—between 1 cm and 36 cm (mean, 6 cm) in diameter. Many large tumors are pedunculated on pleural-based pedicles with hypertrophic vasculature. Numerous thin-walled vessels may be present in larger tumors. Although no single histologic feature provides a definitive prognosis, the presence of a pedicle supporting a well-circumscribed tumor without invading the surrounding structures of the lung, mediastinum, or chest wall is an indicator of a good prognosis.

One review of the literature concluded that complete surgical resection was the preferred therapy for both benign and malignant solitary fibrous pleural tumors. It also stated that though pedunculated tumors are effectively removed using wedge resection, sessile lesions require that a larger mass of lung parenchyma be excised to reduce the likelihood of local tumor recurrence. However, local recurrence is not as worrisome in benign lesions, which have an 8% chance of recurrence, compared with malignant lesions, which have a 63% recurrence rate even after complete resection. Although the majority of recurrent tumors are fatal within 2 years, recovery rate is estimated at 88% to 92% for all patients with solitary fibrous pleural tumors.

In a prospective study, 18 patients were treated with complete resection of tumors with a mean size of 10 cm, receiving follow-up at approximately 6 months. Histologic features were benign in 16 patients and malignant in the remaining 2. The authors determined that the recovery rates were 100% at 1 year, 93% at 3 years, and 80% at 5 years. Tumor recurrence was associated with malignant histology, parietal...
pleura origin, and a lower expression of progesterone receptors. One disquieting case documented the recurrence of a solitary fibrous pleural tumor four times within 10 years, ending finally with a malignant transformation.

Biopsy before excision, however, is controversial. Scarsbrook and colleagues recounted an alarming case in which a solitary fibrous pleural tumor recurred after an ultrasound-guided transthoracic biopsy. Although there have been no other similar reports in the literature, it seems prudent to avoid unnecessary biopsy to prevent this potential complication. As the study recommends, biopsy should only be done if disease management will be substantially affected by the results or if surgical intervention is contraindicated and a diagnosis would alter treatment.

Conclusion
Physicians must obtain the necessary thoracic imaging and surgical consultation to ensure the proper identification of the minimally symptomatic solitary fibrous pleural tumor. The various outcomes associated with these tumors continue to advocate for their therapeutic surgical excision. However, current clinical evidence confirms that long-term clinical follow-up is essential in all patients with the inexplicably impetuous solitary fibrous pleural tumor.

References

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