Case Report

Intrapulmonary location of benign solitary fibrous tumor

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Abstract:

Intrapulmonary solitary fibrous tumors (SFTs) are sporadic mesenchymal neoplasms that typically arise from visceral or parietal pleura. While accounting for <5% of all pleural tumors, SFTs are known to occur in nearly all bodily organs, including nasopharynx, bladder, prostate, soft tissue of neck, buttocks, extremities, and abdominal wall. Such tumors have been previously designated localized fibrous mesothelioma or pleural fibroma. SFTs have no genetic basis and are unrelated to environmental factors such as tobacco smoking or asbestos exposure. Herein, we describe a 24-year-old woman whose clinical presentation mimicked atypical carcinoid tumor. A diagnosis of intrapulmonary SFT was achieved by surgical resection.

Keywords:

Lobectomy, lung cancer, solitary fibrous tumor, video-assisted thoracic surgery

Case Report

A 24-year-old woman presented to the outpatient clinic complaining of localized and intermittent left chest pain for the previous 10 months, accompanied by cough, dyspnea, fever, nausea, vomiting, diarrhea, facial flush, and weight loss. The patient was a nonsmoker and had no known allergies. By history, diagnostic arthroscopy of the left knee took place 5 months earlier. Our clinical examination was unremarkable.

Chest X-ray revealed a solitary opacity of the left lung [Figure 1], which on contrast-enhanced computed tomography (CT) occupied the left lower lobe and measured 4.0 cm × 3.3 cm. The mass was well-defined, rounded, heterogeneous, and predominantly hypodense, with some internal hyperdensity (perhaps enhanced soft tissue or hemorrhage). Segmental bronchi and pulmonary vessels were splayed as a result, but no definitive intrabronchial invasion was evident. A distal, linear atelectatic band devoid of fatty or calcific elements was also identified [Figure 2]. Although the left lower lobe airways and mucosa appeared normal during fiber-optic bronchoscopy, an obstructive tumor was found 2.5 cm from LC2.

Dual-incision hybrid video-assisted thoracoscopic left lower lobectomy was subsequently performed under general anesthesia. On inspecting the hemithorax, no pleural adhesions, effusion, or signs of seeding were detected. Along with the excised intraparenchymal lung mass, lymph nodes were sampled.

Gross examination revealed a firm, well-circumscribed gray-white tumor measuring 4.0 cm maximally. Numerous hemangiopericytoma-like blood vessels were present in the tissue sections [Figure 3] amid a haphazard array of cytologically bland spindled cells and densely hyalinized stroma [Figure 4]. There was no visible necrosis and mitoses were infrequent. The neoplastic cells showed diffuse

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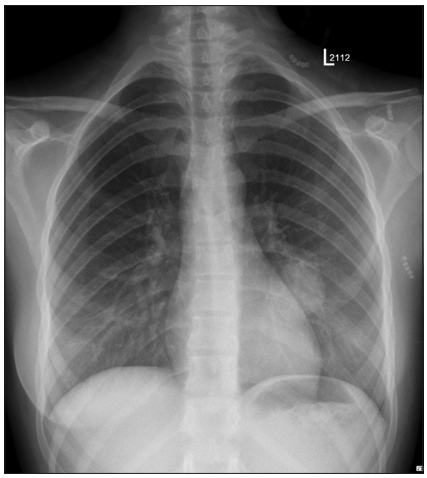

Figure 1: Chest X-ray showing a well-defined, rounded left lung opacity, measuring around 4 cm × 4 cm and devoid of calcification or air-fluid level

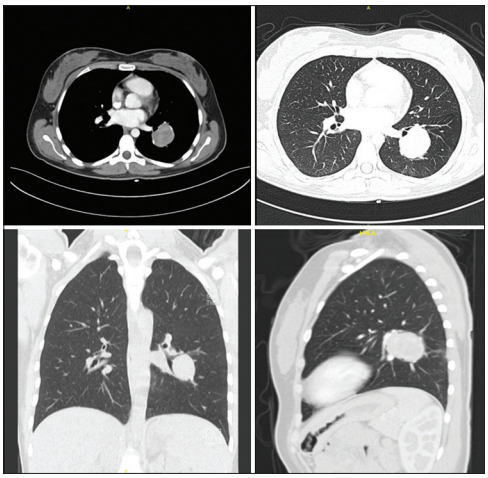

Figure 2: Contrast-enhanced CT of the chest showing a 4.0 cm × 3.3 cm welldefined, rounded, and heterogeneous mass of the left lower lobe. Note the internal hyperdensity and the absence of intrabronchial invasion

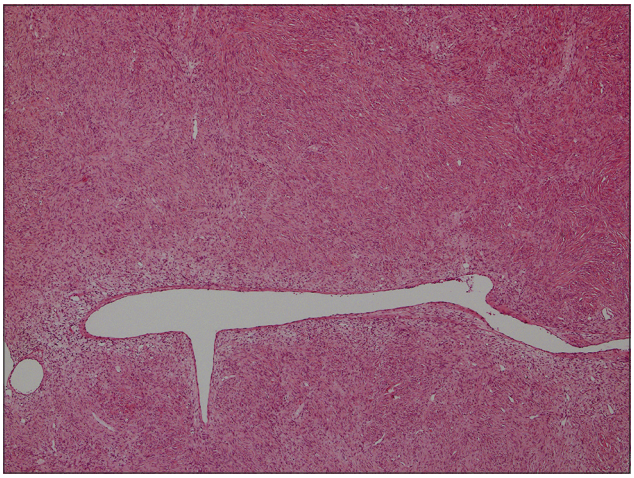

Figure 3: Low-power view of tumor marked by diffuse spindle cell proliferation in hyalinized background and hemangiopericytoma-like blood vessel (H and E, ×40)

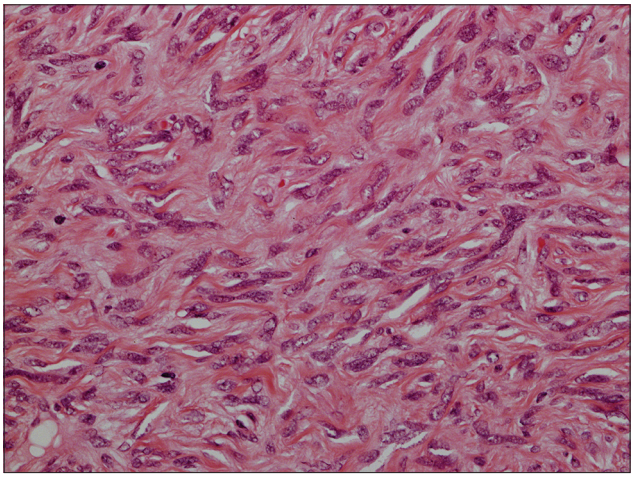

Figure 4: Tumor at high power composed of bland spindle cells with vesicular chromatin in hyalinized stroma (H and E, ×400)

immunoreactivity to CD34 and Bcl-2, whereas no reactivity to smooth muscle actin, S100 protein, or Wilms tumor-1 was apparent. These histologic features are characteristic of solitary fibrous tumor (SFT), confirming the diagnosis and ruling out other potential lesions (i.e., leiomyoma, peripheral nerve sheath tumors, and mesothelial neoplasms). The patient's immediate postoperative course was uneventful, enabling discharge on the postoperative day 5.

Discussion

SFTs are rare mesenchymal growths, with an incidence of 1.4 per million and a fairly low (13%–37%) reported rate of malignancy.[1,2] SFTs of the pleura arise from noncommitted mesenchymal cells of areolar tissue subjacent to mesothelial-lined pleura, usually visceral pleura,[3] that may occasionally present as intrapulmonary lesions (5%–13%).[1,4-6] In such instances, the radiographic features mimic those of a primary pulmonary tumor, and advanced age (i.e. sixth or seventh decade) is common, with no predilection for sex.[3,4,7]

The intrapulmonary origins of SFT may be attributed to (i) direct continuity between mesenchyme and subpleural pulmonary interlobular septa; (ii) parenchymal lung fibroblasts; or (iii) invaginations of visceral pleura.[5] Two main hypotheses have thus been proposed. The first is that subpleural mesenchyma in direct continuity with interlobular connective tissue septa or invaginations of visceral pleura may give rise to intrapulmonary fibromas. The second is that these tumors originate from facultative fibroblastic elements present in the submesothelial areas of normal pulmonary parenchyma. Reports indicate that these elements have ultrastructural and immunohistochemical features similar to those of subpleural connective tissue.[6] According to the literature, intrapulmonary SFTs are seldom

malignant (12.5%).[5] However, this figure reflects only a limited number of cases.

Clinically, a majority of patients with SFT (54%–67%) are symptomatic, presenting with cough, chest pain, dyspnea, fever, and weight loss. In asymptomatic patients, the tumors are generally discovered by routine chest radiographs.[7] Chest pain seems more common in tumors arising from parietal pleura, whereas atelectatic changes regularly reflect a sizeable central mass that compresses neighboring bronchi.[1,3] Still, the clinical and radiologic features of intrapulmonary SFT are nonspecific, hampering the ability to reach a presurgical diagnosis.[8] Excisional biopsy and histopathologic examination are usually required.[3] Fine-needle aspiration or bronchoscopic biopsy are often insufficient for reliable therapeutic guidance.[9]

In our patient, the CT findings (i.e., circumscribed, rounded, and heterogeneous, but largely hypodense parenchymal lesion) and the spectrum/chronicity of clinical symptoms (facial flushing, nausea, vomiting, abdominal pain, and diarrhea) were suggestive of carcinoid tumor. This was later disproven after surgery, but we have found no publications linking SFTs to a paraneoplastic syndrome of this sort. Paraneoplastic syndromes that have been associated with SFTs include Doege–Potter (recurrent hypoglycemia) and Pierre-Marie-Bamberger (hypertrophic pulmonary osteoarthropathy with clubbing of fingers, long bone periostitis, and arthritis) syndromes, which resolved following complete resection.[10,11]

The differential diagnosis of a patient with SFT is that of any mass lesion of the chest, ranging from lung cancer to various intrapleural sarcomas. Tumor location bears a close relation to the likely diagnosis, and immunohistochemistry is critical in differentiating SFTs from other neoplasms. These tumors stain positively for CD34, vimentin, and Bcl2, showing no immunoreactivity to cytokeratin, desmin, alpha-smooth muscle actin, or S100 protein.[12,13]

The mainstay of treating intrapulmonary SFT is complete resection (nonanatomic [wedge], anatomic [segmentectomy or lobectomy], or even sleeve lobectomy) through thoracotomy or video-assisted thoracic surgery (VATS), with 1–2 cm of tumor-free margins.[7,9] VATS is recommended in this setting for greater accuracy of resection margins in some large, broad-based tumors of the parietal pleura. Since contact metastasis or local recurrences at port sites have been reported,[9] avoiding contact between tumor and thoracoscopic sites is also strongly urged.

Outcomes have been favorable in a majority of patients with intrapulmonary SFTs, the location having no detrimental impact on prognosis. In most case reports, follow-up periods after surgery were limited (range, 3 months–13 years; median, 1 year).[13] However, Inoue *et al*. have documented a local recurrence of SFT 2 years after nonanatomic wedge resection of the left upper lobe, emphasizing the importance of longer-term postsurgical follow-up in even benign-appearing tumors.[14] There is no consensus on follow-up in the literature, but extended periodic monitoring is essential, starting with 6-month intervals for the first 2 years. Annual chest X-rays or CT scans are reasonable thereafter, given a known recurrence >20 years after initial resection.[4]

Conclusion

Intrapulmonary SFT is a rare variant of pleural SFT that often eludes presurgical diagnosis. In our experience, related paraneoplastic manifestations are paramount, serving as warning signs of SFT during diagnostic workups of thoracic lesions. Despite their frequently benign course, available data indicate that such tumors warrant close extended clinical follow-up.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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