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Case Report

Primary solitary fibrous tumor of the bronchus: a case report

Wenyong Huang[1], Xiao Xu[1], Jiali Hu[1], Gongwen Cheng[2], Ping Xie[3]

[1]Department of Pathology, The First Affiliated Hospital of Jiujiang University, Jiujiang, China; [2]Department of Cardiothoracic Surgery, The First Affiliated Hospital of Jiujiang University, Jiujiang, China; [3]Department of Pathology, Shanghai Pudong Hospital of Fudan University, Shanghai, China

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Abstract: Solitary fibrous tumors are ubiquitous tumors of fibroblastic type and may be found at any location, but primary solitary fibrous tumors of the bronchus were rarely reported. Here, we reported the case of a 46-year-old woman whose main stem bronchus was partly occluded by a 1.0×0.8×0.7 cm endobronchial mass. The mass was completely resected with lung preservation. The tumor was well circumscribed and consisted of a mixture of bland spindle cells and dense collagen bands. Immunohistochemical staining showed that tumor cells were positive for CD34, bcl-2 and vimentin, but negative for S-100, SMA and cytokeratin AE1/AE3. The tumor was diagnosed as a solitary fibrous tumor of the bronchus, which has been rarely reported to date.

Keywords: Solitary fibrous tumor, bronchus, resection, CD34

Introduction

Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm generally arising from the pleura, but also occurs in various anatomic sites including orbital cavities, nasal cavities, paranasal sinuses, meninges, salivary glands, thyroid gland, liver, pancreas, adrenal gland, spermatid cord, kidney, urinary bladder, prostate, testis, uterine cervix, spinal cord, skin, periosteum, bone and soft tissue [1-5]. The occurrence of SFT in an endobronchial location is a rarer finding; to our knowledge, only 3 cases of endobronchial SFTs have been described in the English literature to date [6-8]. In the present case, we report the clinical and pathological characteristics of bronchus SFT.

Case report

A 46-year-old woman presented with a bronchus mass slowly growing over 3 years. The patient showed no discomfort or local symptoms and had a cough and sputum after catching a cold two weeks ago. The plain computed tomography (CT) scan revealed well-circumscribed soft-tissue attenuation and enhanced CT scan noted strong enhancement and measured approximately 0.8 cm in diameter (**Figure 1A**, **1B**). Bronchoscopy revealed a 1.2×0.8 cm intramucosal mass in the main bronchus (**Figure 2A**). The patient underwent surgical resection in our hospital and the tumor was completely resected.

Macroscopically, the tumor was a 1.0×0.8×0.7 cm well circumscribed mass (**Figure 2B**) and revealed a tan-gray, lobulated cut surface without focal cystic changes. Besides, no necrosis and hemorrhage was noted.

Microscopically, the lesion was composed of a mixture of haphazard, storiform, fascicular spindle cells with rare mitoses (<1/10 high-power field) and dense collagen (**Figure 3A**). No evidence of tumor necrosis or lymphovascular invasion was observed in the lesion. Certain areas showed hemangiopericytoma-like patterns (**Figure 3B**). The lymph nodes and margins were negative.

On immunohistochemical studies, the tumor cells stained positive for CD34 (**Figure 3C**), Bcl-2 (**Figure 3E**), vimentin, and negative for S100, SMA, CD31 (**Figure 3D**), Calponin and cytokeratin AE1/AE3. The Ki67 proliferation index ranged from 10% to 20% (**Figure 3F**).

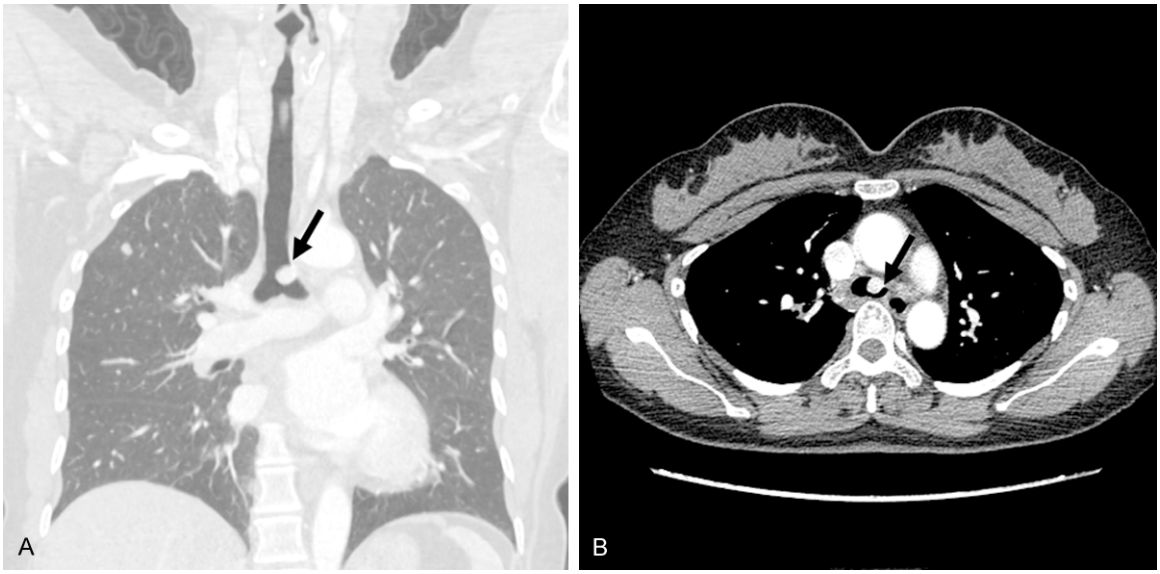

Figure 1. Computed tomography image demonstrates (A and B) a strong enhancement and measured approximately 0.8 cm in diameter in the main-stem bronchus (arrow).

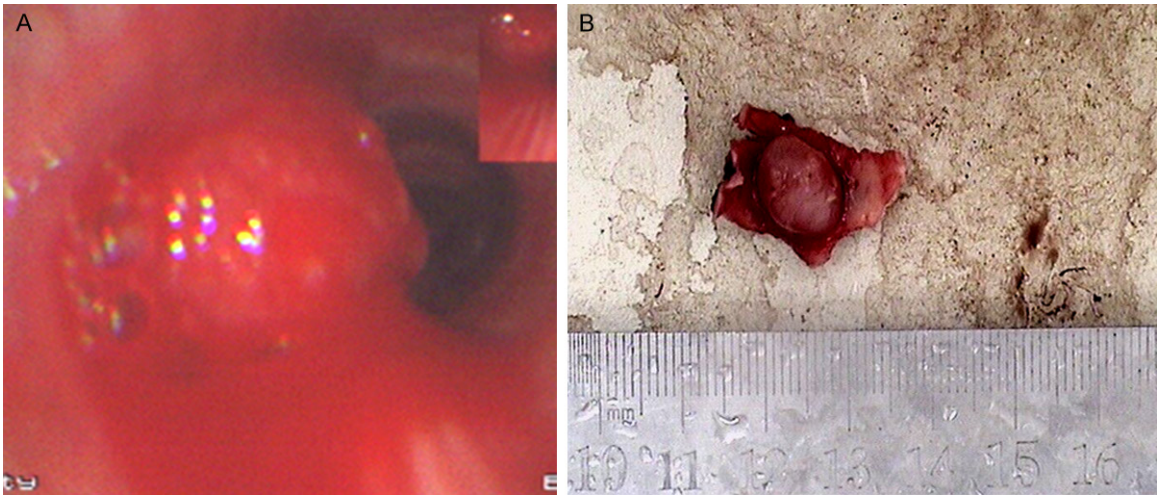

Figure 2. A. Bronchoscopy revealed a 1.2×0.8 cm intramucosal mass in the main bronchus. B. Gross specimen of the tumor was a 1.0×0.8×0.7 cm well circumscribed mass.

Based on the histopathology and immunohistochemical findings, the diagnosis of a bronchus SFT was made. A five-month postoperative follow-up was conducted and the patient was asymptomatic with no evidence of tumor recurrence. Follow-up of the patient is ongoing.

Discussion

Although most SFT arise in the pleura, an endobronchial location is a rare occurrence. To our knowledge, endobronchial SFT has been documented in 3 prior reports [6-8]. We summarize the prior reported cases of SFT features. In our review, there are 4 cases in total including our case. The clinical and pathological features of all cases are detailed in **Table 1**. All patients are adults, with the median age of 64 and the male: female ratio is 3:1. The symptoms occurred in 50.00% (2/4) of patients. After surgical resection, all 4 patients got good effects.

The tumor usually forms a well circumscribed mass. The cut surface of the tumor is usually

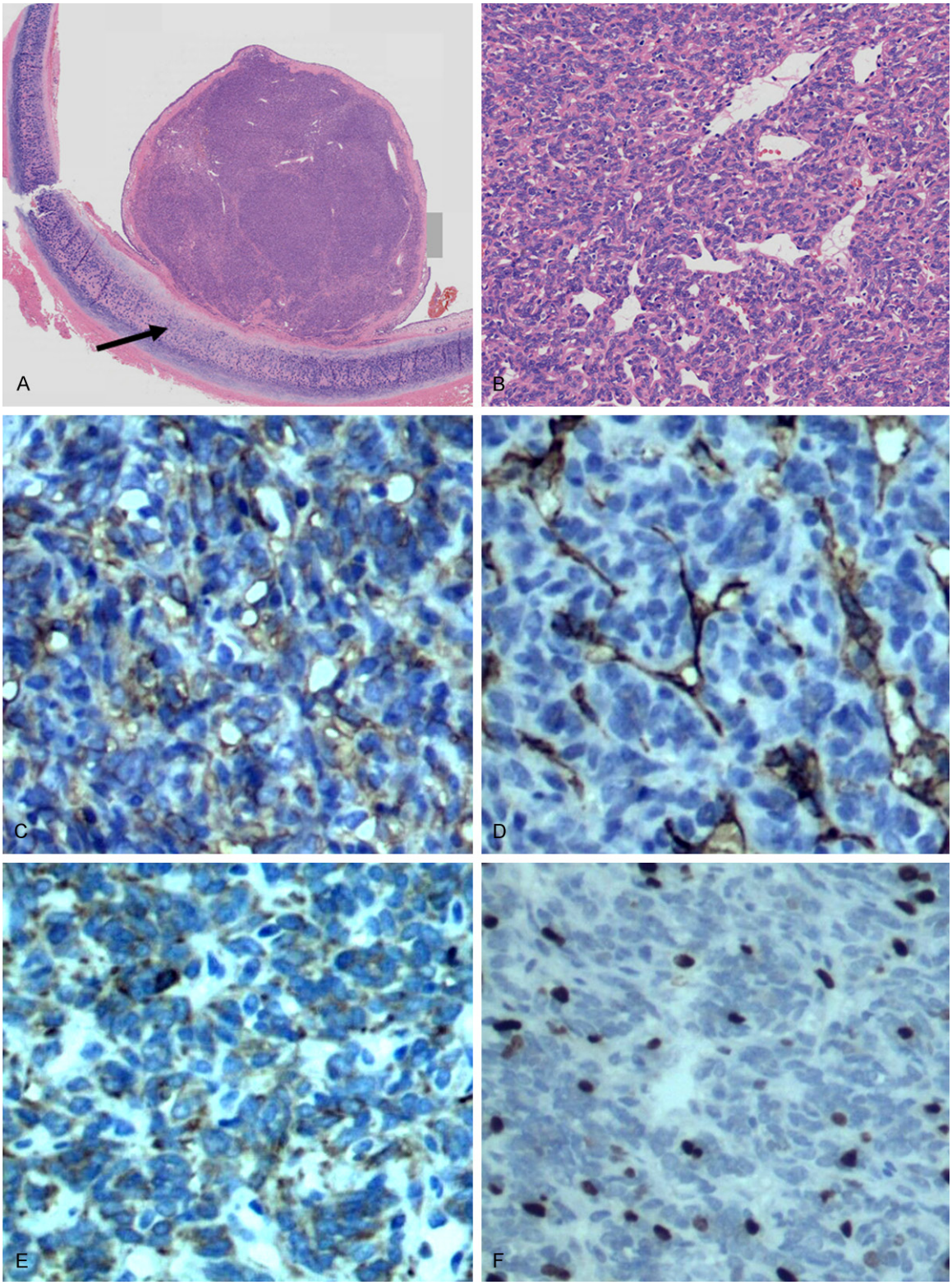

Figure 3. (A) The tumour covered tracheal epithelium and was composed of a mixture of spindle cells and dense collagen. The arrow indicates tracheal cartilage. (B) Hemangiopericytoma-like patterns presented in the tumour. Immunohistochemical staining is positive for CD34 (C) and bcl-2 (E), negative for CD31 (D). (F) The Ki67 proliferation index ranged from 10% to 20%.

Table 1. Clinical features of the endobronchial SFT

Case no	Age sex	Presentation	Size	Treatment	Pathological Features
1	74/M	Asymptomatic	4 cm	Enucleation of the lesion	UA
2	67/W	pneumonias	2.1 cm	Enucleation of the lesion	CD34+, vimentin+
3	69/W	Atelectasis	UA	Enucleation of the lesion	CD34+, bcl-2+, CD99+ ,vimentin+
4	47/W	Asymptomatic	1 cm	Enucleation of the lesion	CD34+, bcl-2+, vimentin+

UA, unavailable; R, right; L, left; M, man; W, women.

solid and the capsule or lobulation can easily be observed. Myxoid and haemorrhagic changes are infrequent [9]. Tumor necrosis and infiltrative margins are mostly observed in locally aggressive or malignant tumors [10, 11]. The 4 cases of endobronchial SFTs in our review are well circumscribed in gross specimen. On section, they have a lobulated firm appearance and no infiltrative margins and necrosis in the tumors.

The tumor is composed of fusiform or ovoid spindle cells and a various amounts of thick, hyalinized or keloid-like collagen bands with patternless, storiform, or fascicular arrangements [12]. Heman-giopericytomatous pattern is frequently noted [13, 14].

Immunohistochemical study is important for diagnosis and differential diagnosis. Virtually all cases of SFT stain with vimentin and the vast majority are positive for CD34, bcl-2 and CD99 [15]. On the contrary, SFT generally shows negative expression of S100, cytokeratin, SMA, CD117, CD31, and Desmin. These markers are useful for the differential diagnosis of SFT from other spindle cell tumors such as angiomyolipoma, leiomyoma, inflammatory myofibroblastic tumor and gastrointestinal stromal tumor. Given the histopathologic and immunohistochemical nature, the present case was consistent with the diagnosis of SFT.

Complete surgical resection with margin negativity is the mainstay of treatment. In a recent report by Harrison-Phipps and colleagues [16], the average median survival for all types of SFT in 84 patients was 24 years. However, It has been reported that 10 to 15% extrathoracic SFTs will recur or metastasize [17]. Certain histological features might indicate a malignant potential, such as more than four mitoses per 10 high power fields, hypercellularity, and larger tumor size [18]. After resection, adjuvant therapy is generally not recommended in histologically benign tumors or the pedunculated lesions that appear malignant [19]. Given that these tumors can have high recurrence rates, regular follow-up with radiographic studies were advised.

In conclusion, we presented a case of primary endobronchial SFT with characteristic clinicophathological features. To our best of knowledge, the occurrence of an endobronchial SFT is exceedingly rare. The characteristic findings of imaging studies, the histological features and immunohistochemical staining for CD34, bcl-2 and vimentin are helpful for the diagnosis of endobronchial SFT. For the time being, surgical resection is the gold standard management and follow-up is necessary.

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Wenyong Huang, Department of Pathology, The First Affiliated Hospital of JiuJiang University, 57 Xunyangdong Road, Jiujiang 332000, Jiangxi, China. Tel: +86-150-79270316; E-mail: wenyongh2009@yeah.net

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