Solitary fibrous tumor of right anterior mediastinum: a case report

Hsiou-Chun Lin¹, Yung-Cheng Wang¹, Hsio-yun Chang¹,∗

ABSTRACT

Solitary fibrous tumors were previously referred to as localized mesotheliomas or pleural fibromas but were later defined as a submesothelial mesenchymal rather than a mesothelial neoplasm. These tumors are rare neoplasms that most frequently occur in the pleura but can be encountered in many other parts of the body.

We describe a case of a 54-year-old man with dyspnea on exertion who underwent surgical resection of a solitary fibrous tumor from the right anterior mediastinum. Computed tomography and angiography imaging studies were used in presurgical survey and tumor embolization. We provided a valuable experience for the image features of solitary fibrous tumors were rarely discussed.

Keywords: solitary fibrous tumor, mediastinum

INTRODUCTION

Solitary fibrous tumors (SFTs) are rare spindle cell neoplasms that occur commonly in the sixth and seventh decades of life with an even frequency in both sexes[1-10]. In 1931, SFTs were first described as localized mesotheloma by Klemperer and Rabin[2, 7-12], but these tumors were later proven to be of mesenchymal rather than mesothelial cell origin[2, 7-13]. Unlike mesotheliomas, SFTs are not associated with exposure to asbestos fiber or tobacco smoking and usually show good prognosis[2, 4, 7]. About 80% of SFTs arise from visceral pleura, accounting for 4% of all pleural neoplasms[1, 3-9, 11-15]. The extrapleural locations of SFTs include other serosal membranes, such as the peritoneum and pericardium, and nonserosal sites, such as the pulmonary parenchyma, mediastinum, nose, and paranasal sinuses[6-10, 12, 13, 15].

Most SFT cases are initially asymptomatic[1, 2, 4, 7-9, 13], and preoperative recognition of SFTs by radiologic imaging and core needle biopsy is often inconclusive[1-7, 9, 13, 14]. The treatment of choice is complete en bloc surgical excision combined with clinical follow up[1-3, 5-7]; the prognosis is usually good but is related to tumor size and location, histologic features, obtaining a
clear surgical margin, and metastasis[1, 2].

SFTs are rare, but mediastinum-originated SFTs are even rarer. Their diagnosis depends mainly on histomorphological findings and immunohistochemical staining. The image features are usually nonspecific and rarely mentioned. In this paper, we present a valuable imaging experience for this rare tumor, with specific computed tomography (CT) and angiographic features of the tumor pedicle from the mediastinum and right internal mammary artery supply. Our findings support the diagnosis of mediastinum-originated tumor.

CASE REPORT

The patient was a 54-year-old man who had experienced dyspnea on exertion for 6 months. In addition, a past history of hyperlipidemia and gastroesophageal reflux disease with associated regular medication was noted, but he denied a history of cigarette smoking and asbestos exposure. On physical examination, decreased breathing sounds were heard on the right side.

Chest X-ray showed a large opacity in the right middle and lower lung fields and right pleural effusion. CT revealed a large solid mass measuring 12.6 cm × 17.2 cm × 19.1 cm in the right hemithorax. The mass was heterogeneous, well circumscribed, enhanced, and with a parallel pedicle from the mediastinum.

CT-guided core needle biopsy revealed a spindle cell neoplasm, and thoracic aortography revealed a hypervascular tumor in the right lower thoracic area with arterial blood supplied by the right internal mammary artery. Transarterial embolization was performed, and the patient underwent a standard sternotomy and tumor resection. A tumor measuring 22.3 cm × 19.3 cm × 12.3 cm was found at the location of the anterior mediastinum with thymic tissue adhesion and right pleural cavity extension, and the right middle and lower lung collapsed because of tumor compression. The well-encapsulated, tan-colored, firm tumor weighed 2108 g. No definite focal aggressive invasion was noted.

Pathology confirmed the diagnosis of a benign SFT of the mediastinum with thymus adhesion, although thymic tissue near the tumor was not invaded by tumor cells. Focal necrosis, myxoid areas, and scattered hemorrhagic spots were also found. Microscopically, the spindle cell tumor revealed alternative hypercellular and hypocellular areas, as well as a patternless growth pattern embedded in a collagenous background. Mitotic figures were absent and immunohistochemistry staining was positive for CD34, Bcl-2, and CD99. Therefore, the criteria for malignancy were not achieved.

After surgery, the patient recovered without complication. After discharge, he continued follow up with regular chest plain film and low-dose lung CT. No tumor recurrence was noted until now.

DISCUSSION

SFTs are uncommon neoplasms that are usually associated with pleural and serosal surfaces[1, 7-9], although their occurrence has been noted throughout the body[6-9]. However, their occurrence in extrapleural sites, such as deep soft tissue, the mediastinum, peritoneum, head and neck, orbit, bladder, kidney, and liver, is extremely rare[6, 7, 9, 10]. SFTs, which were previously recognized as being of mesothelial origin, exhibit immunohistochemical features showing submesothelial mesenchymal differentiation[1, 6, 7, 10].
Over 50% of SFT cases are asymptomatic and discovered incidentally[1, 2, 4, 7-9, 13]. SFTs grow slowly over time; thus, the patient is usually unaware of the tumor’s existence until it has become very large and occupies the entire hemithorax, giving rise to extensive opacities and causing respiratory problems including cough, dyspnea, and chest pain[1, 2, 4, 6-10, 13]. Paraneoplastic syndromes, such as weakness, nocturnal sweating, chills, weight loss, digital clubbing, hypertrophic osteoarthropathy, and hypoglycemia, are less common[1, 2, 4, 6-10]. Hypertrophic osteoarthropathy (Pierre-Marie-Bamberg syndrome) is noted in 20% of cases[2, 7, 10]. Hypoglycemia is caused by the production of insulin-like growth factor II and has been reported in 2%-4% of cases[2, 7, 10]. Serous pleural effusion has been observed in fewer than 10% of all cases[2, 7-10].

The imaging modalities of choice for SFTs are chest radiography and CT scanning, but findings are usually nonspecific and not ideal for use in preoperative diagnosis[2, 5, 7]. Furthermore, differentiation between malignant and benign forms is often not possible through CT or magnetic resonance imaging (MRI)[5], and the effectiveness of determining the features of SFTs using images depends on tumor size and morphology. Evaluating the relationship between the lesion and adjacent structures is another important element[4].

The SFT is typically seen on chest X-ray as a well-defined, lobular, solitary nodule or mass, which may appear to be in the lung periphery, and typically abuts a pleural surface or is located within a fissure[4, 9]. Meanwhile, small tumors arising from the parietal pleura classically form obtuse angles with the chest wall, whereas a large or pedunculated SFT may form an acute angle[10]. Furthermore, pedunculated tumors may show mobility or a change in shape[1, 2, 4, 8-10]. However, the larger the SFT, the more firm its attachment to the adjacent structures through adhesions, which makes the mass less mobile[4].

CT findings are strictly dependent on tumor size[13]. Attenuation usually depends on the collagen content, which presents as hypodense or hyperdense with respect to muscle[8, 9]. Small SFTs are usually homogeneous, well-circumscribed lesions, whereas large SFTs are heterogeneous with necrosis, hemorrhage, or calcification, and cystic or myxoid degeneration[1, 4, 7-10, 13]. Intravenous contrast administration may demonstrate moderate to strong enhancement because of the high vascularity of the tumor[8, 9, 13]. About 100% of malignant and 60% of benign SFTs exhibit heterogeneous enhancement[8] with an intralesional geographic pattern[8, 9, 13]. Intralesional calcifications (punctate, linear, or coarse) are more easily observed in large lesions; moreover, they are associated with areas of necrosis and are described in 7%-20% of cases[13]. Local invasion and lymphadenopathy are rare[9]. CT visualization of a pedicle is rarely reported[9].

In MRI, SFTs are described as having a low or intermediate signal intensity on both T1- and T2-weighted images and on proton density-weighted images, revealing the characteristic fibrous nature of the tumors[1, 2, 8-10]. SFTs appear as vascular tumors with vigorously heterogeneous enhancement, and flow voids reflect the vascular nature (10% of cases)[8, 9]. MRI is highly sensitive in excluding local invasion of the diaphragm and chest wall[9]. In some studies, fluorodeoxyglucose positron emission tomography was useful in identifying SFTs with aggressive features and metastasis and in differentiating benign from high uptake malignant mesothelioma, although associated da-
ta are limited[1, 5, 7, 8]. Angiography is useful in determining the vascular supply to the lesion; determining blood supply from the inferior phrenic, intercostal, or internal mammary arteries may be a helpful clue in evaluating the extrapulmonary origin of the tumor[9]. Ultrasonography is useful for localizing a tumor in the lower hemithorax and assessing its relationship with the diaphragm and extrapleural locations such as the neck, abdomen, pelvis, and extremities[4, 8, 9]. SFTs are typically hypoechoic, but they are occasionally heterogeneous and related to myxoid degeneration[8].

SFTs can be difficult to distinguish from tumors of the mediastinum, lung parenchyma, and chest wall, particularly when they are large enough to occupy the hemithorax[1, 9]. Pleural SFTs are the most common[1, 3-9, 11-15]; they are usually peripheral and smooth with well-demarcated soft tissue density and are often pedunculated and mobile with a fibrovascular stalk[1, 4, 8-10]. Intrapulmonary SFTs are much less common[8], but they typically arise from visceral pleura, interlobular septal connective tissue, or pulmonary parenchymal fibroblasts[4, 8, 9]. Pulmonary SFTs usually present as well-circumscribed round or ovoid soft tissue density nodules or masses, with early and strong enhancement[8]. They are usually benign and slow growing[8]. SFTs have also been found to originate in the pericardium, usually in the anterior and superior mediastinum[8, 15, 16]; about 64% of mediastinal SFTs are reported to have a more aggressive clinical course than pleural SFTs[8, 15, 16].

In the case of our patient, the tumor was large and occupied the right lower hemithorax at an acute angle, with a fixed position and no mobility. With hindsight, the pedicle from the mediastinum with displaced bronchus and angiographic feeding of the right internal mammillary artery could be considered possible clues to a mediastinum-originated tumor extending to the pleural cavity. Differential diagnosis of mediastinal SFTs is extensive as it includes the possibility of spindle cell thymoma, sarcomatoid carcinoma, mesothelioma, hemangiopericytoma, inflammatory myofibroblastic tumor, peripheral nerve sheath tumors, various sarcomas, lymphoma, and metastasis[6, 8, 10].

SFTs are a fibroblast-originated intermediate malignancy that present as spindle cell neoplasms within a background of collagen stroma[1, 6-8]. A “patternless pattern” and hemangiopericytoma-like vascular pattern are diagnostic features[1, 6, 8-10, 13, 14]. Immunohistochemical staining of positive CD34 and negative S-100 may confirm diagnosis[8, 9], as CD34 is a specific marker that differentiates SFTs from mesothelioma and most other pleural tumors[1, 7, 10, 13-15]. Morphological features suggestive of malignant behavior include hypercellularity, an infiltrative pattern, cytounuclear atypia, an increased mitotic figure (>4/10 HPF), large necrosis, or hemorrhage[1, 3, 7].

SFTs are classified as intermediate neoplasms in the WHO classification of soft tissue tumors, with a 12%-30% frequency of malignancy; even benign SFTs have an indeterminate malignant potential[1-3, 6-8, 14]. Therefore, wide and complete surgical tumor resection with a free margin should be performed, and long-term clinical follow up is ideal[1-3, 5-7, 9-11, 15]. To achieve a clear surgical margin, extended resection of the chest wall, parietal pleura, pericardium, diaphragm, or lobectomy is necessary[1-3, 7, 10, 11]. However, video-assisted thoracoscopic surgery may be feasible for use in selected cases, such as small-size tumors (<10 cm) or those that are pedunculated[1, 2, 5, 7, 8].
9-11], although the traditional open technique may have to be performed if margin clearance cannot be attained[1, 2, 7]. Large SFTs with major feeding vessels reportedly benefit from preoperative angioembolization, thereby reducing blood loss during surgery[7]. Following resection, there is little experience and no established rule for adjuvant chemotherapy and radiotherapy[1, 3, 7, 10].

Approximately 90% of SFTs have a benign clinical outcome[1-4]. Favorable prognostic factors include a clear and complete surgical margin, benign histological appearance, a pedunculus, and a limited size[2, 3, 5, 7]. As mentioned, malignant factors are usually related to a large size (greater than 8 cm in diameter), necrosis or hemorrhage, associated pleural effusion, atypical location, invasion of adjacent structures, or metastasis at the time of diagnosis[2, 3, 7]. Recurrence after complete resection is possible in both benign and malignant SFTs[3, 8-10]. De Perrot and colleagues provided a classification for the recurrence rate of SFTs according to tumor characteristics and prognosis: (1) benign pedunculated: 2%, (2) benign sessile: 8%, (3) malignant pedunculated: 14%, and (4) malignant sessile: 63% with a 30% mortality rate and most deaths occurring within 24 months[7-11]. Therefore, careful long-term follow up is highly recommended for both benign and malignant tumors[1, 3, 9-11, 15]. Most recurrences, particularly of sessile malignant tumors, occur within 24 months of initial resection[7, 10]. CT scan is the choice of modality for monitoring recurrence; it should be conducted once every 6 months for the first 2 years and then annually[7, 10]. All patients with SFT require long-term follow up for 15-20 years because of the possibility of late recurrence[7, 9]. Once recurrence occurs, the initial course of treatment is to repeat surgical resection[7, 10].

**CONCLUSIONS**

SFTs are significant uncommon neoplasms, and those of a mediastinum origin are even rarer. Preoperative imaging survey is rarely reported, because surgical and pathological findings are used as the standard diagnostic tool. We now offer an imaging modality for this rare tumor. The specific CT and angiographic features of the tumor pedicle from the mediastinum and right internal mammary artery supply support the diagnosis of extrapulmonary mediastinum-originated tumor. The large tumor occupying the hemithorax with only few clinical symptoms also leads to the consideration of SFT.

**REFERENCES**

J Cardio-Thorac 32 2007; 32: 403-408


Figure 1. Chest X-ray showing large opacity in the right lower hemithorax with pleural effusion.

Figure 2. CT scan of the thorax shows a large, solid, well-circumscribed mass in the right hemithorax measuring 12.6 cm × 17.2 cm × 19.1 cm, with internal calcification (a). Enhanced pattern is heterogeneous (b). The tumor pedicle is from the anterior mediastinum (arrow) (c), (d), (e).
Figure 3. Angiogram of the thoracic aorta revealed the internal mammary artery to be the tumor feeding artery with vivid tumor stain (a)(b). Thus, preoperative transarterial embolization via the right internal mammary artery was performed (c)(d).

Figure 4. Solitary fibrous tumor measuring 22.3 cm × 19.3 cm × 12.3 cm and weighing 2108 g was found at the location of the anterior mediastinum, with thymic tissue adhesion and right pleural cavity extension (a)(b). Microscopically, the spindle cell tumor revealed alternative hypercellular and hypocellular areas, as well as a patternless growth pattern embedded in a collagenous background (c)(d). Mitotic figures were absent. Immunohistochemical stain was positive for CD34 and Bcl-2 but negative for S-100.
中文摘要

單一纖維瘤是罕見的腫瘤，多發生在胸膜，但也可以出現在身體的其他部位：包括頭頸部、縱隔腔、腹腔、深部軟組織。我們描述一位 54 歲男性因為呼吸困難就醫，經 X 光片，電腦斷層掃描，以及血管攝影診斷，手術切除來自右前縱隔腔的單一纖維瘤。

關鍵詞：單一纖維瘤，縱膈腔

*國泰綜合醫院 放射線科
投稿日期：2016年 06 月 01 日
接受日期：2016 年 10 月 12 日
通訊作者：張筱筠 電子信箱：chaos7422@yahoo.com.tw