Resection Of Giant Malignant Solitary Fibrous Pleural Tumor After Interventional Embolization: A Case Report And Literature Review

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Research Article

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Abstract

Background: Solitary fibrous tumor of the pleura (SFTP) is rare mesenchymal tumors that arise at a variety of sites and typically originate from the pleura. Most patients of SFTP are usually asymptomatic unless the tumor is large. Only about 20% of SFTP cases are malignant. There were few reports about the imaging diagnosis and interventional treatment of SFTP. We here in report a case of giant SFTP, which exhibited malignant behavior and underwent a successful resection after embolization of the main supply artery of the tumor.

Case presentation: We report a clinical case of giant SFTP in a 66-year-old Chinese female patient complaining of chest tightness and cough for more than two months. Ten years ago, the patient had a chest CT scan at the local hospital for a cough. The CT scan revealed a mass on the right thoracic,which was misdiagnosed as a pulmonary abscess by a CT-guided biopsy. So the patient was not received the correct and complete treatment at that time. She was hospitalized again because of CT showed a significant enlargement of the right thoracic mass, which caused obvious symptoms of discomfort. The pathological results of CT-guided biopsy in our hospital confirmed SFTP. Considering the large size of the tumor and rich blood supply, part of the main blood vessels were treated with embolization firstly before surgical resection. Then, a large tumor with a size of about 23cm×16cm×15cm was successfully removed by thoracic surgery. The diagnosis of malignant SFTP was confirmed by surgical pathology and immunohistochemistry.

Conclusion: The imaging findings of SFTP are not characteristic, especially when the tumor is large, the diagnosis is difficult, and the final diagnosis still depends on histological and immunohistochemical examination. This two-stage surgical treatment, which involves first embolization of the main supplying artery of the large tumor and then complete surgical resection of the tumor, is effective and safe for SFTP. Whether needle biopsy or vascular embolization, intervention plays a crucial role in the diagnosis and treatment of patients with SFTP.

Background

Solitary fibrous tumor of the pleura (SFTP) is rare mesenchymal tumors that arise at a variety of sites and typically originate from the pleura. Most patients of SFTP are usually asymptomatic unless the tumor is large. Only about 20% of SFTP cases are malignant. The imaging manifestations of giant SFTP are complex and varied, lacking characteristics. Imaging diagnosis and surgical resection of SFTP are challenging. There were few reports about the imaging diagnosis and interventional treatment of SFTP. In this paper, we report a rare case with giant SFTP, which exhibited malignant behavior and underwent a successful resection after embolization of the main supply artery of the tumor.

Case Presentation

A 66-year-old Chinese female had repeated chest tightness and cough for more than two months. CT examination of her chest showed a large mass in the right thorax. Ten years ago, the patient had a chest CT scan for a cough, and a mass in the right thorax was found. A CT-guided biopsy of the lesion was performed and misdiagnosed as a pulmonary abscess at the local hospital. The patient did not receive complete treatment at that time. She was hospitalized in July 2017 because of CT showed the mass in her right thorax was larger than before, causing obvious discomfort symptoms. She had no history of occupational exposure to silica, beryllium or asbestos. Laboratory examination had no specificity. Chest CT examination revealed a significant soft tissue mass at the bottom of the right lung, about 20cm×13cm×12cm in size, with a clear border. Also, the mass was significantly larger than before, with multiple surrounding nodules(Fig. 1). Chest magnetic resonance imaging (MRI) examination showed that the mass presented low signal intensity on T1WI, slightly high signal intensity on T2WI, and low signal intensity on internal strip. Contrast-enhanced CT and MR demonstrated noticeable heterogeneous enhancement of the lesion, and the necrotic area was not enhanced. The coronal view showed that the maximum diameter of the mass was located in the chest cavity, the lung tissue was compressed, and the mass was clearly separated from the lower liver tissue(Fig. 2–3). There was no evidence of chest wall or mediastinal involvement or significant mediastinal displacement.

The imaging appearance of a large chest mass was not sufficient to determine the origin and nature of the tumor, so a biopsy was performed. The CT-guided biopsy demonstrated that the tumor of spindle cells with local degeneration and necrosis, along with immunohistochemical detection, was considered to be SFTP.

Because of the large size and abundant blood supply of the tumor, the patient was referred to our interventional therapy department for the preoperative embolization of some major blood vessels. After angiographic confirmation, hepatic artery and right phrenic artery branches involved in blood supply to the tumor were embolized(Fig. 4). A right posterolateral thoracotomy through the sixth intercostal spaces was performed for the resection of the tumor. When entering the pleura, we found that the encapsulated circumscribed massive tumor blocked the view. We chose the ninth intercostal incision spaces to put in the thoracoscopy found the pedicle of the tumor was located at the top of the diaphragm and located in the right inferior hemithorax was closely related to the lower lobe of the right lung. Finally, a large tumor about 23cm×16cm×15cm in size was wholly and successfully removed by thoracic surgery. The tumor weighed 2250g and appeared smooth surfaced and well-circumscribed on macroscopic examination(Figure 5). Besides, there were two nodules on the parietal pleura near the thoracic spine, which were soft and smooth. Histologically, the tumor cells were abundant, proliferative, and cells were heterotypic (nuclear <4/10HPF). In addition, the tumor cells degeneration and necrosis and an invasive growth pattern on the edge of the tumor were seen. The immunohistochemical evaluation demonstrated positive staining for CD34, vimentin and Bcl2, while pan-cytokeratin (PCK), smooth muscle actin (SMA), S-100, CD163, CD99, CD68, desmin, calretinin (CR) and human melanoma black-45 (HMB-45) were negative. The tumor was pathologically diagnosed as SFTP. The right lung expanded completely and pulmonary function recovered to the normal level after removal of the giant SFTP. The patient had no postoperative complications and no recurrence occurred after surgery in the follow-up period up to date.

Discussion

Solitary fibrous tumor (SFT) is a rare spindle cell tumor originating from dendritic stromal cells expressing CD34 antigen. Both SFT and hemangiopericytoma (HPC) showed 12q13 inversion, NAB-2 and STAT-6 gene fusion. The two were combined as SFT/HPC in 2016 WHO central nervous system tumor classification[1, 2]. SFTP is a rare neoplasm accounting for less than 5% of primary pleural tumors. It derives from the submesothelial mesenchymal layer and usually appears to arise from visceral pleura, and rarely from the parietal pleura. Whereas benign SFTP are often small pedunculated tumors, most malignant SFTP may reach more than 10cm in diameter[2, 3]. However, when the tumor diameter is greater than 15 cm or when the tumor occupies more than 40% of the hemithorax, it can be defined as giant SFTP [4]. The case we described had tumor growth of more than ten years, consistent with the benign tumor growth pattern. Finally, the tumor was surgically removed. The maximum diameter of the tumor was 23 cm, which could be regarded as a giant SFTP. Almost 80% of SFTPs are benign, and more than 50% of their course is asymptomatic. It is generally believed that whether SFTP causes clinical symptoms may be related to tumor size, and most SFTP patients are asymptomatic when the tumor size is small. When SFTP tumor size is large enough to cause compression of adjacent structures and lung tissue, patients may have chest pain, chest tightness, cough or dyspnea and other symptoms[5, 6]. It is known that some SFTP patients may develop so-called "paraneoplastic syndrome," including refractory hypoglycemia, digital clubbing, and pulmonary hypertrophic osteoarthropathy[7]. In our case, the patient had repeated chest tightness and cough for more than two months, which may be a typical clinical manifestation for SFTP.

Thoracic CT is the standard radiological modality for investigating patients with SFTP. It is a useful diagnostic method, which can clearly identify the location and size of the lesion and help surgeons to assess the possibility of resecting the SFTP. These tumors are usually large, well-defined, lobulated, solid and vascular masses, often with a prominent feeding vessel. The enhancement pattern can vary depending on cellularity, vascularity and density of the collagenous or fibrous stroma. Central hypoenhancing or non-enhancing areas may be seen in the tumour, which represents necrosis or cystic change. It often reveals the proliferation of fibrous tissues as well as tumor and adjacent tissue details. Giant SFTPs are usually more likely to cause myxoid or cystic degeneration, hemorrhage, or necrosis. Thus, patchy inhomogeneous enhancement is common in gigantic SFTP with enhancement scanning. This imaging characteristic is often called ‘map sign’. The larger the tumor, the more inhomogeneous the enhancement. Calcification is rare and can be seen in large benign or malignant tumors [8, 9]. Tumors can demonstrate remarkable heterogeneity, with variable degrees of enhancement, necrosis or hemorrhage, and these appearances are not found to differentiate between benign and malignant lesions accurately. SFTPs larger than 10 cm in diameter are usually more likely to be malignant[9]. However, it is difficult to determine the origin of the tumor when the SFTP tumor is giant and occupies the chest cavity. MRI multi-plane scans may be helpful for localization. MRI can provide more information in aiding distinction between benign and malignant lesions, with heterogeneous signal intensity and heterogeneous contrast uptake found to correlate with malignancy. If there is any suggestion of mediastinal invasion, MRI can be

useful for surgical planning[10]. Besides, some investigators have suggested that CT-guided aspiration biopsy is not advisable as a reliable tool due to its low diagnostic sensitivity[11]. In our case, imaging features of SFTP are consistent with previous literature reports, and there is no significant specificity. However, the patient had a CT-guided biopsy of the lesion10 years ago in the local hospital, and the result may have been that the necrotic tissue was obtained by puncture, which was misdiagnosed as a pulmonary abscess. This false-negative result of puncture is what we need to pay attention to.

SFTP is a primary tumor arising from CD34-positive dendritic mesenchymal cells and accounts for <5% of all pleural tumors. Immunohistochemically, SFTP is positive for vimentin, CD34, CD99, and Bcl2[12]. Thus tumors tend to grow into a huge mass before local compression symptoms develop, especially in patients without routine physical examinations.

Conclusion

We describes a rare giant SFTP that was successfully treated by surgical resection. In this current case, a thoracic CT scan was an essential diagnostic imaging method, which demonstrated the characteristic patterns of the tumor. Nevertheless, the confirmed diagnosis and differential diagnosis still depended on subsequent histological and immunohistochemical examinations. The first step is to embolize the main blood supply artery, and the second step is to remove it entirely surgically. Such a two-stage surgical treatment is highly effective and safe for SFTP[13]. Intervention plays a vital role in the diagnosis and treatment of this patient. A long-term follow-up of the patient after surgery is necessary for early detection of tumor recurrence.

Abbreviations

SFTP: Solitary fibrous tumor of the pleura; CT: Computed tomography; MRI: Magnetic resonance imaging; PCK: Pan-cytokeratin; SMA: Smooth muscle actin; CR: Calretinin ; HMB-45: Human melanoma black-45; DWI: Diffusion weighted imaging.

Declarations

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Authors' contributions

YAO K.L. and ZHU L.C. contributed equally to this work. YAO K.L. and ZHU L.C. designed the study and collected data. YAO K.L. analyzed data and wrote the case report. XIA R.M., YANG J.F., WANG L., HU W.B. contributed to the discussion of results and to the review of the manuscript. All authors have read and approved the final manuscript.

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Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request. KelinYao and Jianfeng Yang will make the data available to readers.

Ethics approval and consent to participate

The Ethics Committee of the Affiliated Hospital of Shaoxing University approved the study.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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Figures

Figure 1

Fig1A was the CT film of 2007, Fig1B was the CT film of 2017 at the axial lung window, the mass was significantly larger than before. Fig1C and Fig1D showed a large soft tissue mass at the bottom of the right chest, about 20cm×13cm×12cm in size, with a clear border, significantly enhancement at the axial mediastinal window.

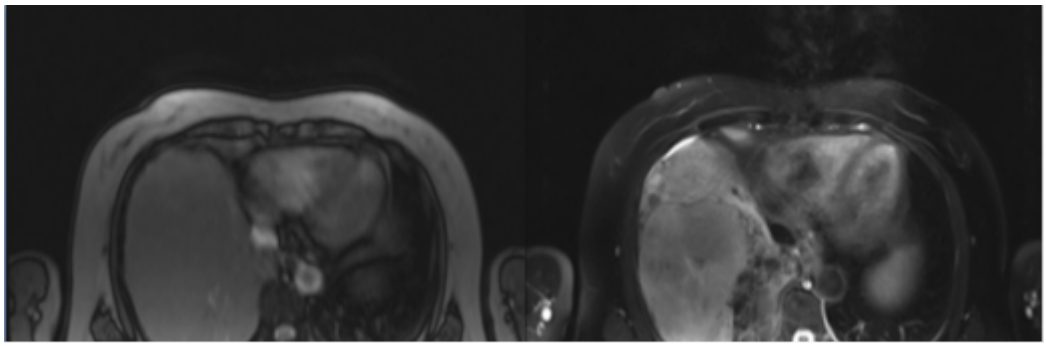

Figure 2

Chest MRI examination showed that the mass presented low signal intensity on T1WI（Fig 2A）, slightly high signal intensity on T2WI（Fig 2B）, and low signal intensity on internal strip. Contrast-enhanced MR demonstrated obvious heterogeneous enhancement of the lesion, and the necrotic area was not enhancement（Fig 2C、2D）

Figure 3

Fig 3A、3B、3C Coronal view of chest MRI examination showed that the maximum diameter of the mass was located in the chest cavity, the right lung lobe was compressed, and the mass has a distinct border with the underlying liver tissue. Fig 3D The mass showed significant high signal on DWI（Diffusion weighted imaging）.

Figure 4

Fig4A、4B Angiography confirmed that the hepatic artery was involved in the blood supply to the tumor and communicated with the right phrenic artery.Fig4C、4D Post-embolization angiography showed that the staining of blood vessels in this part of the tumor had almost disappeared.

Figure 5

5A: Surgical excision of the specimen; 5B: HE staining at 20 times magnification, the tumor cells were abundant, proliferative, and the tumor cells were heterotypic (nuclear <4/10HPF); 5C□5D: Immunohistochemistry showed positive expression of Bcl-2 and CD34 at 20 times magnification.

Supplementary Files

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