**A rare case of retroperitoneal solitary fibrous tumors and literature review**

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**ABSTRACT**

Solitary Fibrous Tumors (SFT) is a relatively rare occupying lesion, which often originated from the pleura and has been reported in a series of reports. However, there are few reports on retroperitoneal SFT. To better understand its biological behavior, clinical diagnosis and follow-up strategy, this case studied the manifestations, diagnosis and treatment of retroperitoneal SFT in a 57-year-old woman, and reviewed the relevant literature.

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**Introduction**

Solitary Fibrous Tumors (SFT) is a relatively rare space-occupying lesion, often originated from the pleura, pleural solitary fibroma has been reported in a series of reports. However, there are few reports on retroperitoneal SFT. So far, no more than 10 cases of retroperitoneal SFT have been reported. Therefore, there is no adequate and unified understanding of the biological behavior, clinical diagnosis and follow-up points of retroperitoneal SFT. Based on this, this case studied the diagnosis and treatment of retroperitoneal SFT in a 57-year-old woman, and reviewed the relevant literature.

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**Case presentation**

A 57 years old female patient came to department with the complaint of left peritoneal mass was found for 3 months. She had any uncomfortable symptom. She was admitted to the hospital with the diagnosis of “retroperitoneal mass”.

She had any uncomfortable symptom and any abnormal physical examination in the left abdominal wall. After in hospital, the patient, with good in consciousness and mentally, denied significant weight loss since the mass was found. The patient underwent CT examination found that left abdominal mass with the size of 5.5 × 7.0 cm (Fig. 1), the left kidney was compressive displacement, ultrasound study found that heterogeneous mass in front of the lower pole of the left kidney. No obvious abnormality was in tumor markers study. A resection of retroperitoneal lesions was conducted smoothly. During the procedure, the tumor is located in the left lateral extraperitoneal space, with the size of about 5.5 × 7.0 cm and abundant blood vessels on the tumor surface.

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**Discussion**

SFT is a rare space-occupying lesion. Klemperer and Rabin reported it for the first time in 1931, which originated from the interstitial subcutaneous connective tissue and often found in the thorax. It is common in the lung parenchyma, mediastinum and pericardium. Then, extrathoracic solitary fibrous tumors have also been reported, such as peritoneum, retroperitoneum, pelvic cavity, meninges, orbit, upper respiratory tract, salivary gland, thyroid, liver, bladder, cervix, spinal cord, periosteum and soft tissue. Madhuvrata and his colleagues reported a case of pelvic SFT and it is also the first comprehensive literature summary.\(^1\)

The pathological microscopic features have been reported as a bland-looking short spindle or polygonal cells with a “patternless” growth, haphazard arrangement, alternating hypercellular and hypocellular sclerotic foci, keloid-like stromal hyalinization and prominent branching vasculature. The immunohistochemical study showed that CD34 and CD99 were positive, which is considered as a positive tumor marker for SFT.\(^2\) They are not specific for SFT, but the expression of CD34 combined with the lack of expression of tumorigenic targets in other immune groups can exclude many kinds of other soft tissue tumors. SFT could also express BCL2, vimentin and desmin in rare cases. Among the...
immunohistochemical study, STAT6, signal transducers and activators of tranion 6, is one of six members in the family, among which STAT6 is not only involved in the activation of il-4 signaling pathway and regulation of immune system, but also related to cell proliferation and apoptosis. In SFT, STAT6 is an ideal diagnostic indicator with 98–100% and 98–100% in sensitivity and specificity, respectively. The biological behavior of extrathoracic SFT is difficult to predict, sometimes atypical SFT has the possibility of recurrence and metastasis. Although the typical SFT has no mitotic capacity, and mild to moderate nuclear atypia, the microscopic features of malignant SFT are: high cell density, pleomorphism, increased mitotic activity (mitosis >4/10HPF). Mitotic >4/10HPF is an indicator used to identify benign and malignant tumors of extrathoracic thoracic cavity. The clinical manifestations of retroperitoneal SFT are often found by chance in physical examination, without clinical symptoms, and can also be palpable abdominal mass sometimes, in one case which has lower abdominal pain and urinary tract compression. The differential diagnosis of retroperitoneal SFT should be retroperitoneal lesions, including

Fig. 1. A retroperitoneal lesion in the CT scan.

Fig. 2. The gross appearance of the retroperitoneal SFT.

Fig. 3. Immunohistochemistry study of the retroperitoneal SFT. top-left, CD 34; top-right, CD 99; bottom-left, BCL2; bottom-right, STAT6.
neurofibroma, schwannoma, vascular leiomyoma, benign and malignant fibrous, histiocytoma, spindle cell lipoma, synovial sarcoma, hemangiopericytoma, dermatofibroma, protuberans, fibromatosis collagenous fibroma and fibrosarcoma. The biological behavior of the thoracic SFT is difficult to predict, and there may be local infiltration, recurrence and even distant metastasis. In the case of SFT in the retroperitoneum, Madhuvrata P concluded that one case recurred 6 months after the operation, and the other 6 cases showed no recurrence during postoperative follow-up.

Surgical resection has been the main treatment. Tumor sizes ranged from 5 to 26 cm, with clear boundaries and pseudomembrane formation on the smooth surface, the SFT tumor is tough texture, easy stripping from surrounding tissues and organs, some cases showed lobulated and multinodular in tumor section. For patients with large tumor that cannot be completely removed, radiotherapy might be effective. Although SFT is a benign tumor, even in the absence of atypical tissue features, there is still the possibility of recurrence and metastasis after complete resection. Aggressive behavior is associated with tumor size, hypercytosis, pleomorphism, and large amounts of mitosis, not always necrosis and bleeding. Therefore, even after complete surgical resection, rigorous long-term follow-up is necessary.

Conclusion

SFT is a rare space-occupying lesion, often originated from the thorax. Nevertheless, lesions located in retroperitoneum should be considered as SFT under certain circumstances, and they could be confirmed by STAT6, CD99, BCL2 and CD34 in immunohistochemistry study. Even its benign character, SFT might have the possibility of recurrence and metastasis after complete resection and a rigorous long-term follow-up is necessary.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2019.101027.

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