A Patient with Giant Presacral Solitary Fibrous Tumor: A Case Report and Literature Review

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Abstract

The paper reports a rare case of large isolated presacral fibroma successfully treated by open surgery. A 43-year-old man was admitted with abdominal distention and difficulty urinating. Pelvic enhanced CT suggested a large cystic solid mass in the lower abdomen and pelvic cavity, up to L3 level and down to the pelvic floor, about 153mm×118mm×212mm in size, with a clear boundary and a close relationship with the leading edge of the spine. The enhanced CT showed uneven enhancement. Since the clinical symptoms and signs of SFT are not specific, imaging examinations such as CT and MRI are the main diagnostic methods of SFT, and the final diagnosis depends on postoperative pathological results. In this case, the postoperative histopathological and immunohistochemical results were consistent with isolated fibroma, and the patient was followed up regularly
within 5 years after surgery. The patient recovered well with no obvious surgical complications, and no tumor recurrence was observed on pelvic MRI.  

**Keywords:** Solitary fibrous tumor; Therapy; Diagnosis; Case report

## Introduction

Solitary Fibrous Tumor (SFT) is a rare spindle cell tumor of mesenchymal origin accounting for less than 2% of all soft tissue tumors [1]. SFT is mostly benign, with slow growth, clear boundaries, and no specific clinical symptoms. They are related to local compression caused by the gradual increase in tumors. Approximately 10% ~ 15% of clinical cases of SFTs show invasive biological behavior, especially tumors located in the mediastinum, abdomen, pelvis, and retroperitoneum, which grow rapidly, infiltrate around the surrounding area and metastasize distantly [2]. Specific advances in immunohistochemistry and molecular diagnosis have identified CD34 as the most consistent marker in SFT. At present, there is no literature report that adjuvant therapy, such as radiotherapy and chemotherapy, has an obvious curative effect on this disease, and surgical therapy is the main means of SFT treatment, which focuses on obtaining a tumor-negative margin. This paper reports a male patient with a large pelvic SFT close to the front of the sacrum. Combined with the literature, the clinical features, imaging findings, treatment, and prognosis of pelvic SFT are reviewed.

## Case Presentation

A 43-year-old male, inadvertently found a lower abdominal tumor in September 2015. The tumor was hard and fixed without abdominal distension and abdominal pain. Later, the tumor gradually increased, and abdominal distension, constipation and dysuria began to appear in February 2016. In March 2016, he was hospitalized because "lower abdominal tumor was found 6 months ago, abdominal distention and dysuria for 1 month". Physical examination showed that the lower abdomen was uplifted, and a large round tumor with a size of approximately 15cm×16cm could be touched, which was hard, fixed, and had poor mobility. The pathological results of tumor puncture showed that the tumor tissue had no necrosis and clear nuclear division; Immunohistochemistry showed that tumor cells CD34 diffuse enhancement (+), Dog-1 (-), CD117 (-), S-100 (-), Actin (-), Desmin (-) and Ki-67 accounted for about 1% (+). Combined with the morphology and immunohistochemical results, the lesion was considered to be a (pelvic) solitary fibroma (Figure 1).
Figure 1: Pathological biopsy in an external hospital considered a (pelvic) solitary fibrous tumor.

Extrapelvic enhanced Magnetic Resonance Imaging (MRI): T1 weighted image (T1WI) showed equal or slightly low signal, necrotic area showed low signal, T2WI showed obvious mixed signal, an enhanced scan showed obvious uneven enhancement, irregular liquefied necrotic area showed no enhancement, tumor cells and vascular aggregation area were significantly enhanced (Figure 2). Enhanced CT showed a huge, cystic solid mass in the lower abdomen and pelvic cavity, which was up to L3 level and down to the pelvic floor, approximately 153 mm×118 mm×212 mm in size, clear boundary, closely related to the anterior edge of the spine. The enhanced scan showed uneven enhancement. Considering the high possibility of neurogenic tumors, it is also necessary to exclude mesenchymal malignant tumors (Figure 3).

Figure 2: T1WI showed equal or slightly low signal, necrotic area showed low signal (a); T2WI showed obvious mixed signal (b); T1 enhanced scan showed obvious uneven enhancement, irregular liquefied necrotic area showed no enhancement, tumor cells and vascular aggregation area were significantly enhanced (c).
Figure 3: Plain pelvic enhanced CT scanning: showed a large cystic, solid mass in the lower abdomen and pelvic cavity, which was up to the L3 level, and down to the pelvic floor, approximately 153×118×212 mm in size, with a clear boundary and closely related to the anterior edge of the spine. (a, b). The enhanced scan showed uneven enhancement (c).

Combined with the patient’s condition, solitary fibroma tumor had the possibility of metastasis, and malignant tumors from mesenchymal sources are not excluded. If conservative treatment is continued, the progressive expansion of the tumor will lead to more obvious compression symptoms. Large tumors also have the risk of rupture and infection, which will also seriously threaten the lives of patients. Considering the above factors, surgical resection will be the best treatment at present. In view of the above considerations, experts from imaging department, vascular intervention department, gastroenterology department, anesthesiology department and urology department conducted Multidisciplinary Treatment (MDT) to further evaluate the operation risk and make sufficient preoperative preparations. According to the discussion of MDT, angiography and bilateral internal iliac artery balloon embolization were performed before operation to reduce the risk of intraoperative bleeding. Cystoscopy was performed before tumor resection, and double-J ureteral tube was planned to be placed, but the ureter was severely squeezed by the tumor, resulting in the failure of ureteral catheter placement. Therefore, the large tumor in the pelvic cavity should be carefully removed to avoid damaging the ureter as much as possible. During the operation, the tumor was tough, the surface blood vessels were dilated, the surface of the tumor envelope tissue was smooth, and the size of the tumor was about 16cm × 12cm × 20cm. The tumor compresses and adheres to the left ureter and expands. The bladder and part of the colon are pushed to the right side of the abdomen. Finally, the tumor ruptured and was resected in blocks (Figure 4).
Figure 4: a) the tumor capsule is smooth, and the surface blood vessels are dilated; b) the tumor adheres closely to the surrounding tissues; c) free protection of the compressed ureter; d) the tumor is broken, and the tumor is removed in blocks.

The intraoperative bleeding was approximately 11,000ml. The pathological results of postoperative tumor specimens showed that the tumor was composed of spindle cells with thick collagen fiber bundles. Immunohistochemical examination showed that CD34(++++), BCL-2(+++), Vimentin (+++), and Ki-67 were less than 1% positive, SMA(-), S-100(-), CD31(-), and CK(-). The pathological diagnosis was a pelvic SFT (Figure 5).

Figure 5: H&E staining (100 × magnification) showed small branching vessels, spindle tumor cells are arranged in bundles, and cell atypia is not obvious (a); Immunohistochemistry (100 × magnification) showed diffuse
The patient recovered well after the operation, with normal urination and defecation function and no operation-related complications. After five years of follow-up, no tumor recurrence was found on the pelvic MRI (Figure 6).

![MRI Images](image_url)

**Figure 6:** After five years of follow-up, no obvious abnormality was found in pelvic MRI.

**Discussion**

Solitary Fibrous Tumor (SFT) was first found to originate from the pleura and was first reported by Klemperer et al. [3] Later, it was found that SFTs can also be widely distributed in human connective tissue, of which 30% of SFTs occur in extrapleural parts, such as the liver, peritoneum, thyroid, cerebellum, and vagina [4]. Most SFTs are benign, malignant and metastatic with low incidence. However, when the tumor diameter is greater than 50mm, and is accompanied by nuclear pleomorphism and tissue necrosis, malignant transformation should be assumed. In addition, a tumor diameter greater than 10cm and incomplete resection of tumor tissue are also high-risk factors for local recurrence and metastasis [5]. A clinical study of 110 cases of pleural and extrapleural SFT by Decco et al. [6] found that the metastasis rate of SFT 5 and 10 years was approximately 26% and 45%, the overall survival rate was approximately 89% and 73%, and its development and prognosis could not be predicted. SFT growth is generally slow, and the incidence rate of males and females is the same, but the incidence rate of middle-aged patients is higher than younger patients [7]. Because most of them are not accompanied by pain, they are often difficult to detect. Its clinical manifestation is an isolated soft tissue mass, quasi circular, or irregular, expansive growth, clear boundary, and envelope- or envelope-like structure on the surface of the tumor. Although the site of SFT can be anywhere in other parts of the body, the volume of SFT in
the pelvic and abdominal cavities is often large, and the maximum diameter can reach more than 10 cm [8]. Patients often have abdominal discomfort, sacrococcygeal pain, or defecation disorder due to tumor compression and the pushing of surrounding organs, and a few patients have accessory tumor syndrome. The most common symptoms are hypoglycemia of nonislet cells, hypertrophic osteoarthropathy, or clubbing fingers [9]. In this case, the diameter of the tumor was more than 20 cm. Abdominal distension, constipation, and dysuria occurred were caused by compression of the intestine and bladder, but there was no paraneoplastic syndrome. Because the clinical symptoms of SFT are not specific, imaging examination is still the main diagnostic method of SFT. On color Doppler ultrasound, SFT often shows a clear boundary, no calcification, and a uniform hypoechoic or uneven tissue mass. Although SFT cannot be accurately diagnosed, it has important clinical significance for initial diagnosis, differential diagnosis, and guiding tissue puncture biopsy [10]; CT examination can directly reveal the size, shape, essence, and location of tumors. Tumors of different properties also have different CT manifestations, but they do not have specificity for the diagnosis of SFT. On a plain CT scan, SFT showed a well-defined, equal, or slightly low-density mass with varying degrees of necrosis, cystic changes, or bleeding. After enhancement, characteristic "map like" enhancement can be seen in most arterial phases, and continuous enhancement in the portal phase and delayed phase shows "fast forward and slow out" [11]. Compared with CT, MRI has a better resolution for soft tissue tumors. SFT showed equal or slightly low signal on T1WI and mixed strip high signal in equal or slightly low signal on T2WI. Low signal intensity areas were observed in larger tumors, corresponding to the flow gap of blood vessels around the prominent lesions [12]. However, imaging examination lacks specificity and sensitivity in the diagnosis of tumor nature and origin. In this case, a well-defined cystic, solid mass was found on the preoperative CT, and the enhancement scan showed uneven enhancement; MRI showed equal or slightly low signal on T1WI, a low signal in the necrotic area, an obvious mixed signal on T2WI, and obvious uneven enhancement on the enhanced scan, which was in line with the imaging characteristics of SFT reported in the literature. The diagnosis of SFT depends on histopathological examination and immunohistochemical results. The pathogenesis of SFT is not completely clear. It is generally believed to be related to the generation of the nab2-stat6 fusion gene by paracentric inversion of chromosome 12q13 [13]. Microscopically, SFT tumor cells were spindle or oval, arranged in bundle, sheet and braid, and had a large number of collagen fibers. They were also arranged in bundle, mat pattern, and braid, with a large number of collagen fibers. Spindle cells were unevenly dense, with little cytoplasm, oval nuclei, and uniform size and chromatin distribution. SFT is a tumor derived from CD34-
positive dendritic stromal cells that can differentiate into fibroblasts. Therefore, CD34 is considered a specific marker of SFT, but CD34 is also expressed in many other tumors, such as neurofibroma, schwannoma, and cutaneous fibrosarcoma [14]. Ding ZY et al. [15] found that the expression of STAT6 in SFT had higher sensitivity and specificity than that of CD34, CD99 and Bcl-2. In addition, studies have shown that STAT6 protein in SFT is the most sensitive and specific marker for the diagnosis of conventional and malignant SFT, and CD34 combined with STAT6 is helpful to improve the specificity and sensitivity of SFT diagnosis [16]. In immunohistochemistry, SFT usually shows strong expression of CD34, BCL2, and CD99, but the specificity is poor [17]. CD34 positivity can be used as an important index for the diagnosis of SFT. When CD34 is negative, CD99 and Bcl-2 positivity are conducive to the diagnosis of SFT. Yuliang Sun et al. [18] studied the immunohistochemistry of 24 cases of benign and malignant SFTs in different parts and found that the expression of Ki-67 has a certain potential role in identifying benign and malignant SFTs. It is an important marker reflecting the proliferative activity of tumor cells and can help to understand the biological behavior of tumors and judge the prognosis. Therefore, immunohistochemistry often uses CD34 combined with vimentin, CD99, and Bcl-2 as the basis for the diagnosis of SFT, and the positive rate of Ki-67 reflects the degree of proliferation of SFT. Through these surveyed literatures, it was found that CD34 and BCL-2 are important markers for the pathological diagnosis of SFTs (Table 1). The postoperative pathological immunohistochemistry of this case showed CD34 (+++), bcl-2 (+++), and Vimentin (+++), while the expression of Ki-67 was low, indicating that the pathological diagnosis of this patient was in line with the diagnosis of benign SFT reported in the literature. It indicated that the proliferation of SFT is not high and has a low trend of malignant lesions, and the prognosis was good.

Table 1: Characteristics of SFT patients in the surveyed literature.

<table>
<thead>
<tr>
<th>Author</th>
<th>Numbers (M/F)*</th>
<th>Surgical Year</th>
<th>Age(A)*</th>
<th>Symptoms</th>
<th>Tumor size(cm)</th>
<th>Pathological/Immunohistochemistry features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Klemperer P. et al. [3]</td>
<td>5(3/2)</td>
<td>1919-1926</td>
<td>26-53(43.8)</td>
<td>Pain in the chest and shadow in the right side of the chest</td>
<td>7x8x5 to 25x19x12</td>
<td>Mesenchymal structure and originate from the subpleural areolar tissue.</td>
</tr>
<tr>
<td>Author</td>
<td>Cases</td>
<td>Gender</td>
<td>Age</td>
<td>Location</td>
<td>Size</td>
<td>Description</td>
</tr>
<tr>
<td>-------------------</td>
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<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Demicco EG, et al. [6]</td>
<td>103(51/52)</td>
<td>N/A</td>
<td>19-81(52)</td>
<td>Tomors in abdomen, pleura, head, neck and trunk</td>
<td>1~40(10.3)</td>
<td>Most tumors were at least moderately cellular, with only 3 cases demonstrating uniformly low cellularity and 51 cases being highly cellular. Necrosis was present in 37 cases, marked nuclear pleomorphism in 12, and distinct, sharply demarcated poorly differentiated areas were seen in 3 cases. Mitotic figures ranged from 0 to 23/10 high-power fields.</td>
</tr>
<tr>
<td>Eltawil KM, et al. [9]</td>
<td>1</td>
<td>N/A</td>
<td>63</td>
<td>Periumbilical pain and nausea</td>
<td>11.4×6.8×6.6</td>
<td>Negative from CD31, ASMA, S100, AE1/AE3, calretinin, desmin and CD117, positive for vimentin, BCL-2 and CD34.</td>
</tr>
<tr>
<td>Badea R.et al. [10]</td>
<td>1</td>
<td>N/A</td>
<td>57</td>
<td>a large solitary mass of the left hemithorax</td>
<td>12×8</td>
<td>Reactive for vimentin and stained strongly and consistently for CD34 and bcl2.</td>
</tr>
<tr>
<td>Yuliang Sun et al. [18]</td>
<td>24(10/14)</td>
<td>N/A</td>
<td>29-89(56.2)</td>
<td>tumors occurred in the pleura, pelvic space, prostate, retroperitoneum, cerebellum, diaphragm, prostate, thigh, shoulder, abdominal wall and popliteal fossa.</td>
<td>1.8×2×3 to 10×15×33</td>
<td>Highly immunoreactive to antibodies for vimentin, CD34 and MIC2. Bcl2 was detected in 20 of the 24 patients.</td>
</tr>
</tbody>
</table>

*(M/F), Male/Female; (A), Average; N/A, Not Available.
SFT is a tumor that is mostly benign, but 10% ~ 20% are malignant or latent malignant [19]. Clinically, benign pelvic SFT is often differentiated from gastrointestinal stromal tumors and schwannoma. Gastrointestinal stromal tumors have diverse shapes, clear boundaries, uneven density. They are prone to cystic degeneration or necrosis. Plain CT scans show uneven enhancement, and abnormal vascular shadows are seen in the arterial phase. Schwannomas tend to occur in the retroperitoneum, which are mostly distributed along the nerve. The tumor boundary was clear, and the edge was smooth. It had a complete capsule, and was prone to cystic changes. CT scans mainly focus on cystic and solid changes. After dynamic enhancement, the solid part shows progressive enhancement, the cystic part does not strengthen, and the enhanced scan is characterized by mild enhancement, which is helpful for the differential diagnosis of them. Malignant pelvic SFT should be differentiated from undifferentiated pleomorphic sarcoma and leiomyosarcoma. Undifferentiated pleomorphic sarcoma, also known as malignant fibrous tissue lymphoma, mostly occurs in elderly men. CT scans show uneven density, equal to or lower than adjacent muscle tissue, uneven enhancement on enhanced scans, pleomorphic calcification at the tumor edge, and cord-like shadows around the tumor [20]. Leiomyosarcoma shows invasive growth, blurred boundaries, necrosis, and bleeding. It shows a large lobulated solid mass on CT, generally containing cystic space or necrosis. MRI mainly shows an equal signal on T1WI, with cystic and flake low signal inside. T2WI often shows slightly high or high signals, and enhanced scanning shows mild to obvious delayed enhancement [21]. At present, surgical intervention is still the main treatment for pelvic SFT in order to completely remove the tumor and negative margins and avoid damaging the surrounding tissues and organs or other important structures. Benign and noninvasive solitary fibrous tumors generally will not recur or metastasize after complete resection. The resection plane of SFT should be as close to the normal tissue as possible, and the resection range includes the tumor base and the normal tissue no less than 1 cm around the tumor to obtain a satisfactory result [22]. Therefore, SFT should be completely radical resection, and the surgical method should be formulated according to the location, size, and boundary of the tumor. Whether the tumor is completely removed also directly affects the prognosis. For SFTs with large volumes and difficult operations, intra-arterial embolization can be used to strive for surgical treatment [23]. SFT mostly shows benign biological behavior. It has been reported that approximately 10% ~ 23% of intrapleural and nearly 10% of extrathoracic SFTs have malignant biological behavior, showing local recurrence or distant metastasis [24]. Many studies have found that tumors with a diameter of >10cm have a higher metastasis rate, while invasive growth, abundant and dense tumor cells, common mitotic images (>4/10 high power field), cell atypia, nuclear
pleomorphism, and/or interstitial or vascular invasion, necrosis or bleeding are considered to be the malignant biological manifestations of SFT, and having one or more of the above histological features indicates the presence of malignant lesions [23]. Therefore, the histological conformation of SFT cannot completely and accurately predict its prognosis, and patients should be followed up regularly. Based on the analysis of 219 cases of multisite SFT, it is considered that the location and size of the tumor are important risk factors affecting the prognosis. There is a greater risk of distant recurrence and metastasis of thoracic, abdominal, and retroperitoneal SFTs, and the risk of postoperative recurrence is greater for SFTs with diameters >8cm [25]. Combined with this case, the maximum diameter of the tumor is more than 20cm, although the pathological and immunohistochemical results suggest benign SFT. However, the large tumor and severe adhesion with surrounding organs and tissues still has a certain tendency of recurrence and metastasis. Close follow-up is needed after the operation.

Conclusion
This case report is a supply previous report of solitary fibroma tumors. It is a rare tumor that can be found on B-ultrasound, CT, and MRI, but its diagnosis still depends on pathological examination. In this case, we found that CD34 and BCL-2 are important markers for the pathological diagnosis of SFT, and the expression of Ki-67 may play an important role in differentiating benign from malignant SFT. In our experience, it is important and necessary to seek MDT for complex SFT in order to prepare the work well before operation. Only by formulating the best individualized treatment plan in multiple dimensions can we obtain a satisfactory prognosis.

References


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