

## A CASE OF RETROPERITONEAL SOLITARY FIBROUS TUMOR

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Solitary fibrous tumor (SFT) of the retroperitoneal space is rare. We report a case of retroperitoneal tumor, diagnosed as SFT. A 69-year-old woman presented with right lower abdominal swelling, and was referred to our hospital with suspicion of right renal tumor. Abdominal ultrasound and computerized tomography (CT) showed a mass (about 15×14×10 cm) in the right abdomen. The tumor was thought to be right renal tumor, and right radical nephrectomy was performed. In the excised specimen the tumor was not connected to gastrointestinal tract, peritoneum, or right kidney. The histological and immunohistochemical examination of the specimen revealed SFT. The tumor has malignant potential with partially increased mitotic activity and cellularity in the histological examination. The patient is healthy and without evidence of recurrence or metastasis 26 months from surgery.

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**Key words** : Solitary fibrous tumor, Retroperitoneal tumor

### INTRODUCTION

Solitary fibrous tumor (SFT) was first reported by Klemperer and Rabin in 1931 as a tumor of pleura<sup>1)</sup>. The retroperitoneum is rare as the site of origin, and there are few case-reports<sup>2–4)</sup>. Clinical diagnosis is difficult because of rarity, and immunohistochemical examination is helpful for diagnosis. Most SFTs are benign, but some show malignant behavior. It is thought to be difficult to predict clinical outcome from histological findings. We describe a case of huge retroperitoneal SFT with histologically malignant transformation.

### CASE REPORT

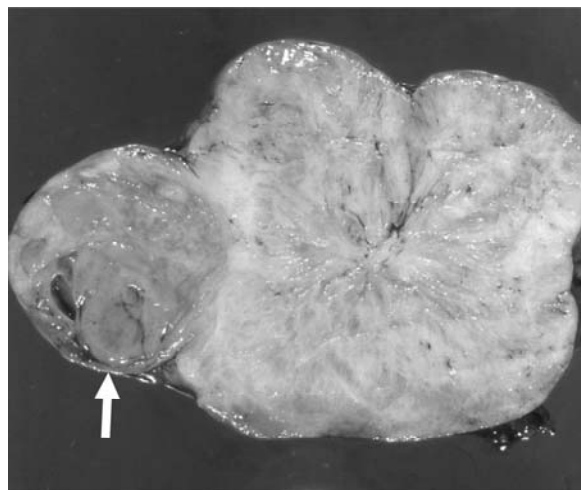
A 69-year-old woman presented with painless swelling in the right lower abdomen, and was referred to our hospital with suspicion of right renal tumor. Physical examination revealed a hard fixed mass occupying the right abdomen, but lymph node swelling was not palpable. Laboratory findings and immunosuppressive acidic protein (IAP) level were in normal range. Abdominal ultrasound showed a heterogeneous large tumor. Computerized tomography (CT) showed a huge mass (about 15×14×10 cm) with early enhancement in the right abdomen (Fig. 1). Neither lymph node swelling nor distant metastasis were detected by chest X-ray, CT or bone scintigraphy. The tumor was thought to be right renal tumor, and right radical nephrectomy was performed. In the excised specimen the tumor was not connected to gastrointestinal tract,



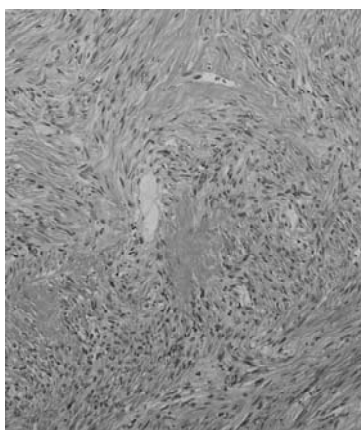
**Fig. 1.** Computerized tomography (CT) showed a huge mass (about 15×14×10 cm) with early enhancement in the right abdomen. The borderline between the tumor and right kidney is unclear.

peritoneum, or right kidney. The tumor was diagnosed as retroperitoneal tumor. The maximal diameter of the tumor was 14 cm, and the weight 1,192 g. The cut section of specimen showed mostly fibrous appearance, but partially myxoid appearance (Fig. 2). Histological examination of the specimen revealed that the tumor consisted of proliferation of spindle cells with patternless pattern and hemangiopericytoma-like appearance (Fig. 3A), the cellularity increased and the nuclear atypia were moderate. In the areas of fibrous appearance, the frequency of mitosis was from 0 to 2 per 10 high-power fields (hpf) and Ki-67 labeling index was 1–2%. On the other hand, mitotic activity in the areas of myxoid appearance was more than 4/10 hpf (Fig. 3B), and Ki-67 labeling index was 20%. The tumor cells were

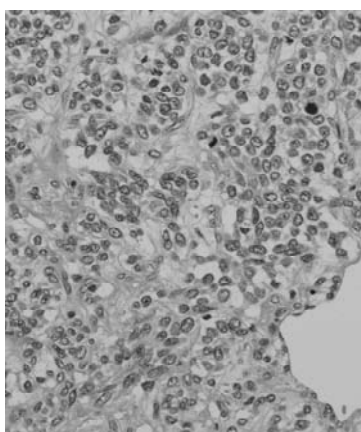
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**Fig. 2.** On cut surface of the specimen, the tumor shows myxoid appearance in part (arrow).



(A)



(B)

**Fig. 3.** Histological findings (H & E) of the specimen show that the tumor consists of proliferation of spindle cells with hemangiopericytoma-like appearance in the fibrous appearance (A). On the other hand, the cellularity and mitotic activity increase prominently in the myxoid appearance (B).

immunohistochemically positive for CD34 and bcl-2 protein, negative for  $\alpha$ -smooth muscle actin (SMA), S-100 protein, and c-kit. Thus, the tumor was diagnosed

as SFT. At the 26-month follow-up examination, the patient was healthy and without evidence of recurrence or metastasis.

## DISCUSSION

Klemperer and Rabin first reported the solitary fibrous tumor (SFT) as a pleura-based lesion in 1931<sup>1)</sup>. SFTs have been found in the pleura, but they have been reported in other sites including meninges, orbit, peritoneum, liver, kidney, retroperitoneum and soft tissue. The SFT occurring in the retroperitoneum is rare, and only some cases have been reported<sup>2-4)</sup>. The present case was not connected to the gastrointestinal tract, peritoneum, or right kidney, and the tumor was thought to be retroperitoneal tumor.

Nakatani et al.<sup>4)</sup> reviewed 25 patients with retroperitoneal SFT. There were 11 male and 13 female patients aged 17 to 82 years (mean 49 years), with tumor sizes from 2 to 26 cm (mean 10.6 cm). In the retroperitoneum, most of the small SFTs were asymptomatic, and some tumors were large (> 10 cm). Some large SFTs had local recurrence or metastasis. Our patient had a large tumor occupying the right abdomen. It was necessary to follow it carefully.

The pathological features of SFT were "patternless pattern" characterized by a haphazard, storiform arrangement of short spindle or ovoid cells, and "hemangiopericytoma-like appearance" with prominent vascularity by thin-walled vessels. Immunohistochemically, expression of CD34 have been reported<sup>5)</sup>. The majority of SFT have also been reported to be positive for bcl-2<sup>6,7)</sup>. Immunohistochemical examination such as CD34 and bcl-2 has been found to be helpful for the diagnosis of SFT. Our patient had both patterns and coexpressed CD34 and bcl-2.

England et al.<sup>8)</sup> described high cellularity, high mitotic activity (more than four mitotic figures per 10 high-power fields), pleomorphism, and necrosis as criteria of malignancy. Immunohistochemical marker such as Ki-67 is thought to be useful for differential diagnosis between benign and malignant SFT. Ueda et al.<sup>9)</sup> reported that the low Ki-67 labeling index groups in their 34 patients with soft tissue sarcomas showed more favorable prognosis than the high labeling index groups ( $p < 0.05$ ). Sun et al.<sup>10)</sup> reported a mean Ki-67 labeling index of 1.9% ( $1.9 \pm 0.43\%$ ) for benign SFT and 6.11% ( $6.11 \pm 1.05\%$ ) for malignant SFT ( $p < 0.05$ ). Hasegawa et al. described that the numbers of MIB-1-positive nuclei were low in all histologically benign cases, ranging from 0.5 to 10% (mean 1.8%). Therefore, Ki-67 labeling index of 20% was thought to be immunohistochemically malignant.

It was unique to have two different appearances in the tumor. Most of this tumor was included in the criteria of benign histologically, while there was a myxoid area in some part, where mitotic activity and Ki-67 labeling index increased prominently. It was thought that

malignant transformation might occur. The patient was healthy without evidence of recurrence or metastasis 26 months after surgery, but careful long-term follow-up is mandatory.

### ACKNOWLEDGMENT

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## 和文抄録

## 後腹膜 Solitary fibrous tumor の 1 例

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後腹膜 solitary fibrous tumor (SFT) は稀な疾患である。症例は69歳, 女性。右下腹部の腫瘍を主訴に近医を受診, 右腎腫瘍が疑われ当科紹介となった。腹部超音波および CT 検査にて右腹部に 15×14×10 cm 程の充実性腫瘍を認めた。右腎腫瘍と考えられ, 右根治的腎摘除術を施行した。腎と一塊に腫瘍を摘出した

が, 摘出標本にて腎との連続性はなく, 後腹膜腫瘍と判断された。病理組織検査にて solitary fibrous tumor と診断された。術後26カ月経過し, 現在再発の徴候なく外来通院中である。

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