Clinical Case Report

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Malignant solitary fibrous tumor in retroperitoneum

A case report and literature review

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Abstract

Rationale: Solitary fibrous tumor (SFT) is a rare mesenchymal tumor occurs in various sites. Malignant SFT in retroperitoneum is extremely rare.

Patient concerns: We report a case of malignant retroperitoneal SFT in a 59-year-old man presented with right flank pain for 1 month.

Diagnoses, interventions and outcomes: A laparotomy and resection of the tumor were performed, the histopathologic and immunohistochemical findings were consistent with malignant retroperitoneal SFT. No adjuvant treatment was performed, and the patient had no signs of recurrence or metastasis at the 12 months follow-up.

Lessons: Complete surgical excision is the basic treatment principle for malignant retroperitoneal SFT. The histologic features and the Ki-67 label index are helpful for the diagnosis of malignant SFT.

Abbreviations: CT = computed tomography, SE = surgical excision, SFT = solitary fibrous tumor.

Keywords: literature review, malignant tumor, retroperitoneum, solitary fibrous tumor

1. Introduction

Solitary fibrous tumor (SFT) is a rare mesenchymal tumor occurs in various sites. SFTs usually develop in the pleura, and about 30% to 40% of SFTs arise in extra-pleural regions.[1] Retroperitoneal SFTs are a subgroup of extra-pleural SFTs, often present with nonspecific symptoms. They can present either benign or malignant characteristics at pathological examination.

Malignant SFT in retroperitoneum is extremely rare, with evidence of only 9 cases of malignant retroperitoneal SFTs published in literature.[2–7] In the present study, we report a case of malignant retroperitoneal SFT and review the clinical and pathological characteristics.

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Written informed consent was obtained from the patient for publication and any accompanying images.

YZ and XC have contributed equally to this work.

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2. Case report

A 59-year-old man presented with right flank pain for 1 month. The patient denied any other symptoms, as well as recent weight loss. The medical history was unremarkable. Physical examination showed no abnormal findings, except for a nonpainful mass at palpation. A computed tomography (CT) scan showed a well-circumscribed retroperitoneal tumor measuring $14 \times 13 \times 10$ cm with several circuitous vessels, compressing the right kidney (Fig. 1A and B). Based on these findings, a retroperitoneal tumor was diagnosed.

At laparotomy, a smooth-surfaced large tumor occupied the retroperitoneal space, compressing the right kidney. Most of the tumor was encapsulated and easily resected, thus right nephrectomy was avoided. The tumor was fed partially by the abdominal aorta and superior mesenteric artery. There were areas of hemorrhage and necrosis. Microscopic examination revealed that the tumor was composed of haphazard, interlacing fascicular spindle cells. There were hypercellular and hypocellular regions (Fig. 2A). The tumor consisted of a mixture of bland spindle cells and collagenous matrix with patternless pattern. In addition, moderate atypical mitoses (4 mitoses per 10 high power filed [HPF]) were found (Fig. 2B).

On immunohistochemical studies, the antibodies employed are listed in Table 1, together with their source and dilution. CD34 reactivity is regarded as the most prominent characteristic finding in the diagnosis of SFT.[8,9] Other positive immunoreactivities in SFT include CD99 and vimentin. In the present study, the tumor cells stained positive for CD34 (Fig. 2C), CD99 (Fig. 2D), vimentin (Fig. 2E), and STAT6 (Fig. 2F) and negative for S100, CD117, HMB45, EMA, SMA, and Dog-1. The proliferation rate Ki-67 was about 20% (Fig. 2G). Based on the histopathologic and immunohistochemical findings, the diagnosis of a malignant retroperitoneal SFT was made.

The postoperative period was uneventful. The patient experienced no postoperative complications and was discharged 7 days

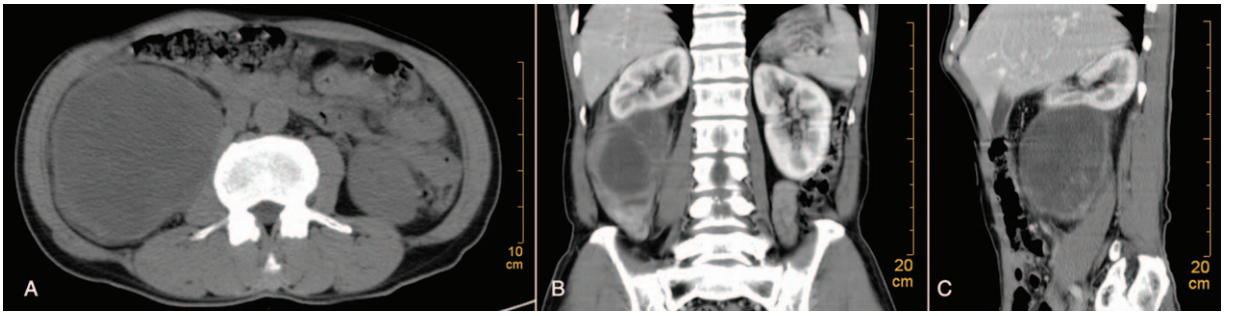

Figure 1. (A) Noncontrast abdominal CT showed a well-circumscribed mass in retroperitoneum. (B, C) Contrast abdominal CT showed a slightly enhanced mass, compressing the right kidney. (B) Coronal sections and (C) sagittal sections. CT = computed tomography.

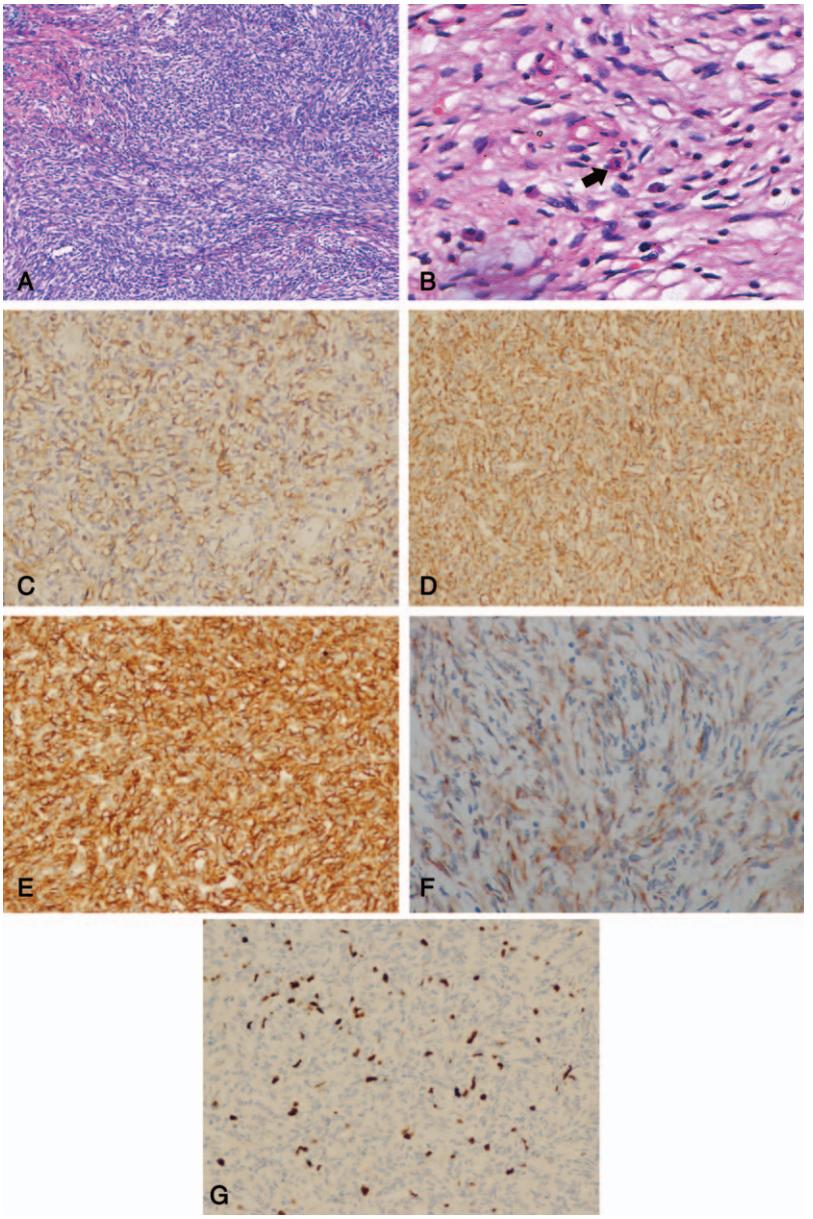

Figure 2. (A) Hematoxylin and eosin stain showed increased cellularity. The tumor consisted of spindle-shaped cells with patternless pattern (magnification ×200). (B) Nuclear mitoses were seen (arrow) (magnification ×400). (C) Immunohistochemical staining for CD34 was positive (magnification ×200). (D) Immunohistochemical staining for CD99 was positive (magnification ×200). (E) Immunohistochemical staining for vimentin was positive (magnification ×200). (F) Immunohistochemical staining for STAT6 was positive (magnification ×400). (G) The Ki-67 index was about 20%.

Table 1

Antibodies employed for immunohistochemistry.

Antibody	Source	Dilution
CD34	ZSGB-BIO, China	1:200
CD99	ZSGB-BIO, China	1:100
Vimentin	ZSGB-BIO, China	1:100
STAT6	Maxim biotech, China	1:100
S100	ZSGB-BIO, China	1:100
CD147	ZSGB-BIO, China	1:100
HMB45	Maxim biotech, China	1:100
EMA	Maxim biotech, China	1:100
SMA	ZSGB-BIO, China	1:50
Dog-1	Maxim biotech, China	1:100
Ki-67	ZSGB-BIO, China	1:200

after the surgery. No adjuvant treatment was performed, and the patient has no signs of recurrence or metastasis at the 12 months follow-up.

3. Discussion

SFT, first reported in 1931 by Klemperer and Rabin,[10] is a spindle cell neoplasm which occurs most often in the pleura. A wide variety of extra-pleural sites also have been noted, including orbit, nasal cavity, salivary glands, upper respiratory tract, thyroid, peritoneum, retroperitoneum and pelvis, genitourinary system, and soft tissue.[11,12] It is reported that 30% to 40% of SFTs are located at extra-pleural regions.[1] Retroperitoneal SFT is rare, <100 cases have been reported in the literature.[13]

Histomorphologically, SFT typically exhibits a patternless pattern characterized by a haphazard, storiform arrangement of spindle cells and a hemangiopericytoma-like appearance with prominent vascularity. Most SFTs are benign, approximately 10% to 20% of the tumors are with malignant behavior. According to the microscopic features, the pathologic criteria for malignancy are as follows: high cellularity, high mitotic activity (more than 4 mitoses per 10 HPF), pleomorphism, necrosis, and hemorrhagic changes.[14] To the best of our knowledge, only 9 cases of malignant SFT in retroperitoneum have been reported in the English-language literature (summarized in Table 2).

As SFT is originated from spindle cell neoplasm, the differential diagnoses include other spindle cell tumors such as leiomyoma, inflammatory myofibroblastic tumor, angiomyolipoma, and gastrointestinal stromal tumor. Immunohistochemical studies are helpful in confirming the diagnosis and differential diagnosis of SFT. CD34, reported to be diffusely and strongly positive in many cases of SFTs, is regarded as a positive marker.[8,9] In addition, the positive findings for Bcl-2, vimentin, and CD99 support the diagnosis of SFT. On the contrary, SFT generally shows negative expression of S100, cytokeratin, EMA, SMA, CD117, CD31, and Desmin. Recently, several studies have found that NAB2-STAT6 gene fusions occurred in the vast majority of SFTs.[15,16] In the study by Urabe et al,[17] they found that STAT6 was diffusely positive in a case of SFT. However, the role of IHC for STAT6 in SFTs remains uncertain. Therefore, in the present study, the immunohistochemical staining for STAT6 was performed. Interestingly, we found that the tumor cells stained positive for STAT6. All these findings indicate that STAT6 may be a potential positive marker in SFTs.

Ki-67, a proliferation-associated antigen, is expressed in active phases of the cell cycle including G1, S, G2, and mitosis. Sun et al[18] performed immunohistochemical staining for Ki-67 in 24 cases of benign (14 patients) and malignant (10 patients) SFT. Their results showed that the mean Ki-67 labeling index is 1.9% for benign SFTs and 6.11% for malignant SFTs ($P < 0.05$). It suggested that Ki-67 could be diagnostically relevant to the evaluation of malignant SFT. In our case, the Ki-67 index was about 20%, showing the tumor was capable of high degree of proliferation as a malignant tumor.

Some imaging features are considered as characteristic of SFTs. The findings, such as a well-circumscribed mass, T2 hypointensity on magnetic resonance imaging, avid and heterogeneous enhancement on CT and magnetic resonance images, all indicate the diagnosis of SFTs.[19] However, the imaging findings of SFTs are variable and not specific. Recently, several studies have reported SFTs mimic other different kinds of tumors. Bae et al[5] reported a case that the gastroscopic examination and abdominal CT presented as a gastric submucosal tumor. However, the surgical and histologic results revealed a retroperitoneal SFT. Similarly, Urabe et al[17] reported a case of omental SFT that presented as a gastrointestinal stromal tumor of the small intestine. In their study, the CT scan revealed that the feeding artery to the tumor might be the left gastroepiploic artery, and it was finally proved that the tumor was originated from the greater omentum. Thus, detecting the feeding artery may be helpful for the diagnosis and differential diagnosis of SFTs.

Surgical excision (SE) is the basic treatment principle for both benign and malignant SFTs. Recently, Rajeev et al[13] analyzed 51 patients of retroperitoneal SFTs from the National Cancer

Table 2

Summary of cases of malignant SFT in retroperitoneum.

Reference/year	Age	Sex	Max diameter	Treatment	Recurrence	Metastases	Outcome	Mitosis, per 10 HPF	Immunohistochemical positive
Vallat-Decouvelaere et al (1998)[2]	40	M	17	SE	Yes, 12 mo	Lung, 12 mo	AWD, 34 mo	4.5	CD34
	63	F	4.5	SE	Yes, 168 mo	No	NED, 180 mo	15	UA
	70	F	10	SE	No	No	NED, 10 mo	4.5	CD34
Takizawa et al (2008)[3]	UA	F	UA	SE	No	No	UA	10.2	CD34, CD99, Bcl-2
	UA	M	UA	SE	No	No	UA	10	CD34, CD99, Bcl-2
Ito et al (2008)[4]	48	F	5.5	SE	No	No	NED, 30 mo	UA	CD34, CD99, Bcl-2, vimentin, p53
Bae et al (2011)[5]	59	M	22	SE	No	No	NED, 36 mo	10	CD34, CD99, Bcl-2
Baldi et al (2013)[6]	41	M	UA	SE	Yes, 23 y	UA	DOD, 24 y	UA	UA
Yoh et al (2014)[7]	43	M	17	SE + AC	No	No	NED, 12 mo	3	CD34, CD99, Bcl-2, vimentin, Ki-67 (37%)
Present	59	M	14	SE	No	No	NED, 12 mo	4	CD34, CD99, vimentin, STAT6, Ki-67 (20%)

AC = adjuvant chemotherapy, AWD = alive with disease, DOD = dead of diseases, HPF = high power filed, NED = no evidence of disease, SE = surgical excision, SFT = solitary fibrous tumor, UA = unavailable.

Database and 24 patients from systematic review of published literature. Their results revealed that complete SE was a feasible and reasonable first line of therapy for retroperitoneal SFTs with minimal perioperative morbidity and mortality and overall median survival above 4 years. Postoperative radiotherapy or chemotherapy was performed to reduce recurrence in several studies.[20,21] However, comparing to other retroperitoneal sarcomas, the recurrence rate was reported to be relatively lower in retroperitoneal SFTs.[22,23] Thus, the use of routine adjuvant radiation or chemotherapy in malignant retroperitoneal SFTs is controversial. In our case, the tumor was completely resected and no adjuvant treatment was performed. The patient has no signs of recurrence or metastasis at the 12 months follow-up.

In summary, we present a rare case of malignant SFT in retroperitoneum. The histologic features and the Ki-67 label index are helpful for the diagnosis of malignant SFT. Complete SE is the basic treatment principle. As no established standard systemic therapy, the use of routine adjuvant radiation or chemotherapy needs to further study. Careful clinical long-term follow-up is necessary.

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