

## Solitary Fibrous Tumor of the Thigh: A Patient Report

**Kensaku Yamaga, Takeshi Minamizaki\*, Mari Osaki\*, Kohei Shomori, Naoko Ishiguro†, Haruhiko Yoshida and Hisao Ito**

*Division of Organ Pathology, Department of Microbiology and Pathology, School of Medicine, Tottori University Faculty of Medicine, Yonago 683-8503, \*Department of Orthopedic Surgery, Yonago Medical Center, National Hospital Organization, Yonago 683-8518 and †Department of Pathobiological Science and Technology, School of Health Science, Tottori University Faculty of Medicine, Yonago 683-8503 Japan*

**We report a rare case of solitary fibrous tumor (SFT) within the thigh muscle. A 37-year-old woman noticed a painless mass in her left thigh, which had gradually enlarged. Computed tomography (CT) showed an intramuscular tumor and enhanced CT showed strong enhancement within the lesion. Magnetic resonance imaging revealed that the tumor measured 13.1 × 7.3 × 2.3 cm and was located within the sartorius muscle, with a clearly defined margin. Malignancy was suspected and the tumor was surgically resected. The tumor was characterized by a well-circumscribed lesion, and was composed of bland or short spindle cells, which were patternless, with scattered thick collagen bundles, and variably gaping thin- or hyalin-walled blood vessels. Immunohistochemically, the tumor cells were positive for vimentin, CD34, CD99 and focally, bcl-2. The tumor was subsequently diagnosed as an extrapleural SFT, which is classified into an intermediated (rarely metastasizing) category. Extrapleural SFT should be considered in the evaluation of soft tissue tumors in the extremities, although it may be rare.**

**Key words:** extremity; soft tissue tumor; solitary fibrous tumor

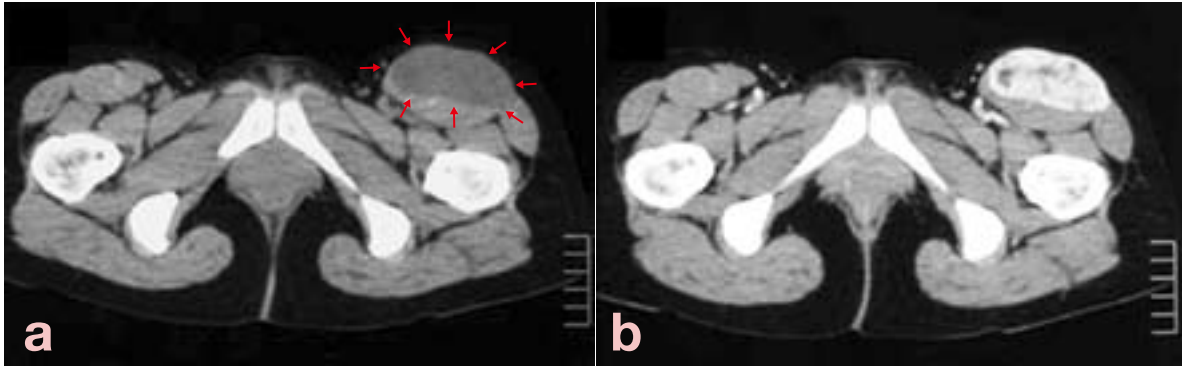
Solitary fibrous tumor (SFT) was first described by Klemperer and Robin in 1931 as a pleura-based lesion (Klemperer and Rabin, 1931). While SFT was known initially as a pleural lesion, it has been reported in various sites, including soft tissue of the extremities, head, neck, thoracic wall, mediastinum, pericardium and abdominal cavity (Guillou et al., 2002). We describe a 37-year-old woman who had an SFT of the deep soft tissue in her thigh. Extrapleural development of the tumor is relatively rare and the diagnosis was difficult prior to surgery.

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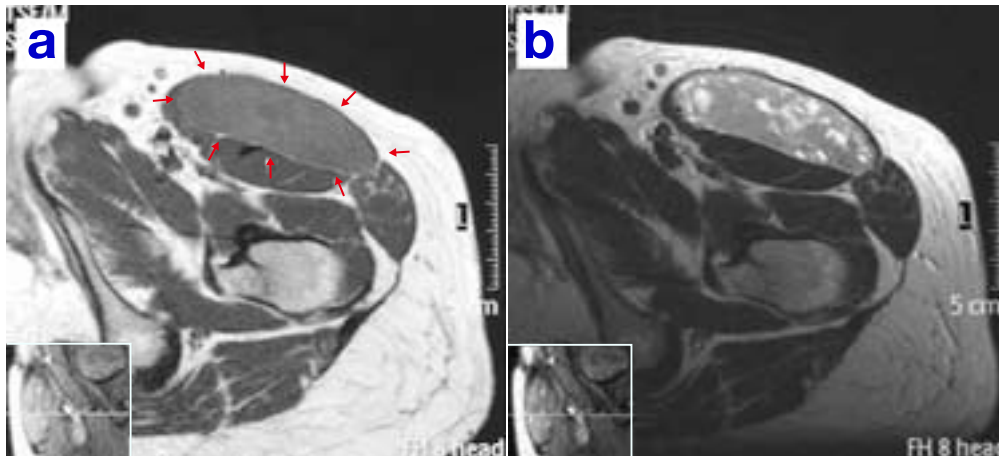
Abbreviations: CK, cytokeratin; CT, computed tomography; EMA, epithelial membrane antigen; HPF, high-power field; MRI, magnetic resonance imaging; MPNST, malignant peripheral nerve sheath tumor; SFT, solitary fibrous tumor; SMA, smooth muscle actin

### Patient Report

A 37-year-old woman noticed a painless mass in her left thigh a year ago. Six months later, she visited a nearby hospital because the mass had begun to gradually enlarge. A soft tissue tumor was noted in her left thigh. She was referred to our hospital for evaluation and therapy. Physical examination showed that the tumor was located in the deep soft tissue of the anterior thigh, 10 cm in diameter, elastic/hard, unmovable and hemispherical. No abnormality was observed in the skin. Computed tomography (CT) showed an intramuscular mass in her left thigh, which was well margined, and enhanced CT showed strong enhancement within the lesion (Fig. 1). Magnetic



**Fig. 1.** a: Plain CT showed an inter-muscle mass which was well-circumscribed.  
b: Enhanced CT showed strong enhancement in the tumor.



**Fig. 2.** The mass was located within the sartorius muscle.  
a: T1-weighted imaging exhibited low signal intensity of the mass with scattered lower signal intensity.  
b: T2-weighted imaging exhibited high signal intensity of the mass with scattered higher signal intensity.

resonance imaging (MRI) revealed that the tumor measured  $13.1 \times 7.3 \times 2.3$  cm and was located within the sartorius muscle, with a clearly defined hemispheric margin. T1-weighted imaging exhibited low signal intensity of the tumor with scattered lower signal intensity (Fig. 2a). In contrast, T2-weighted imaging exhibited high signal intensity of the tumor with scattered higher signal intensity (Fig. 2b). Tl scintigraphy demonstrated marked uptake in both early and delayed images. Blood glucose level was normal.

Based on the clinical findings and imaging examination described above, we diagnosed the mass as a malignant tumor of the soft tissue, such as synovial sarcoma, alveolar soft tissue sarcoma and malignant peripheral nerve sheath tumor (MPNST). Core needle biopsy was performed for

histopathological diagnosis. Although the tumor was diagnosed as a spindle cell tumor and suspected of low-grade malignancy, histological type could not be diagnosed. Thereafter, modified wide resection was performed.

### ***Pathological findings (operative specimen)***

#### ***Gross findings***

The tumor was capsulated and well circumscribed. On sectioning, it had a whitish and firm appearance with partial hemorrhage and no necrosis (Fig. 3).

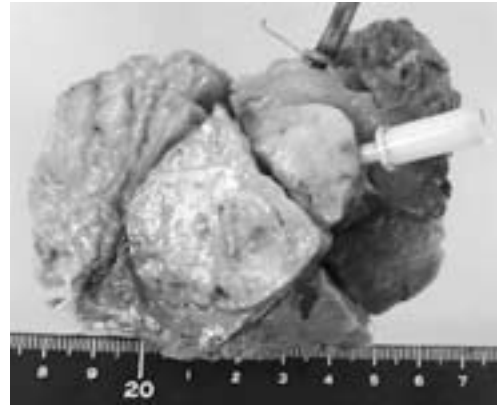
#### ***Histopathological examination***

The tumor was well circumscribed. It was composed of alternating cellular and less cellular

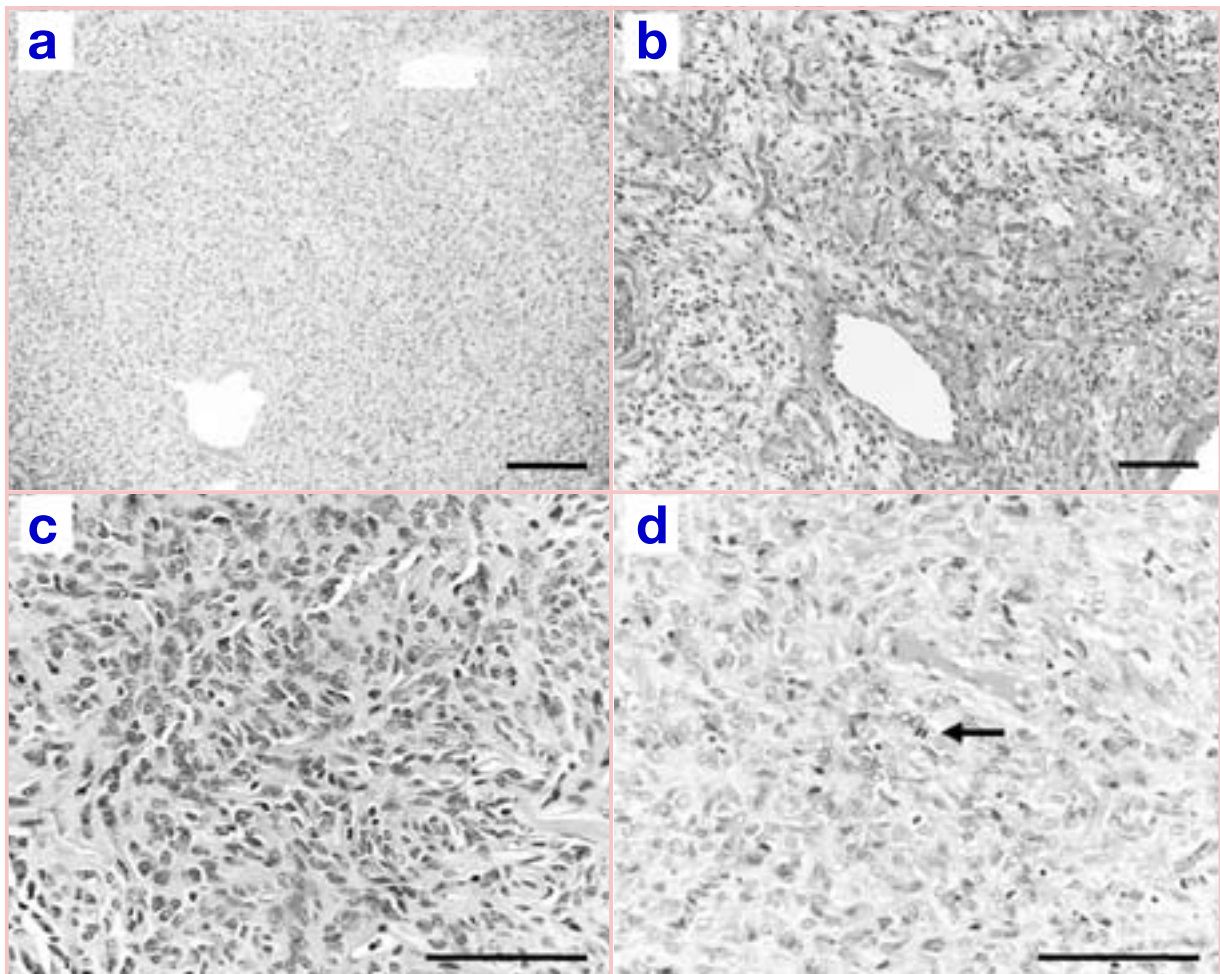
areas, and of cytologically bland spindle or short spindle cells arranged without an obvious pattern, with scattered thick collagen bundles, and variably gaping thin- or hyalin-walled blood vessels, which had a hemangiopericytoma-like appearance, and a fibromyxoid stroma (Fig. 4). Mitotic figures were less than 1/HPF. Based on the morphologic features, there was no convincing evidence of malignancy.

**Immunohistochemical examination**

The tumor cells were positive for vimentin, CD34, CD99, and focally for bcl-2, whereas alpha-smooth muscle actin (SMA), desmin, S-100 pro-



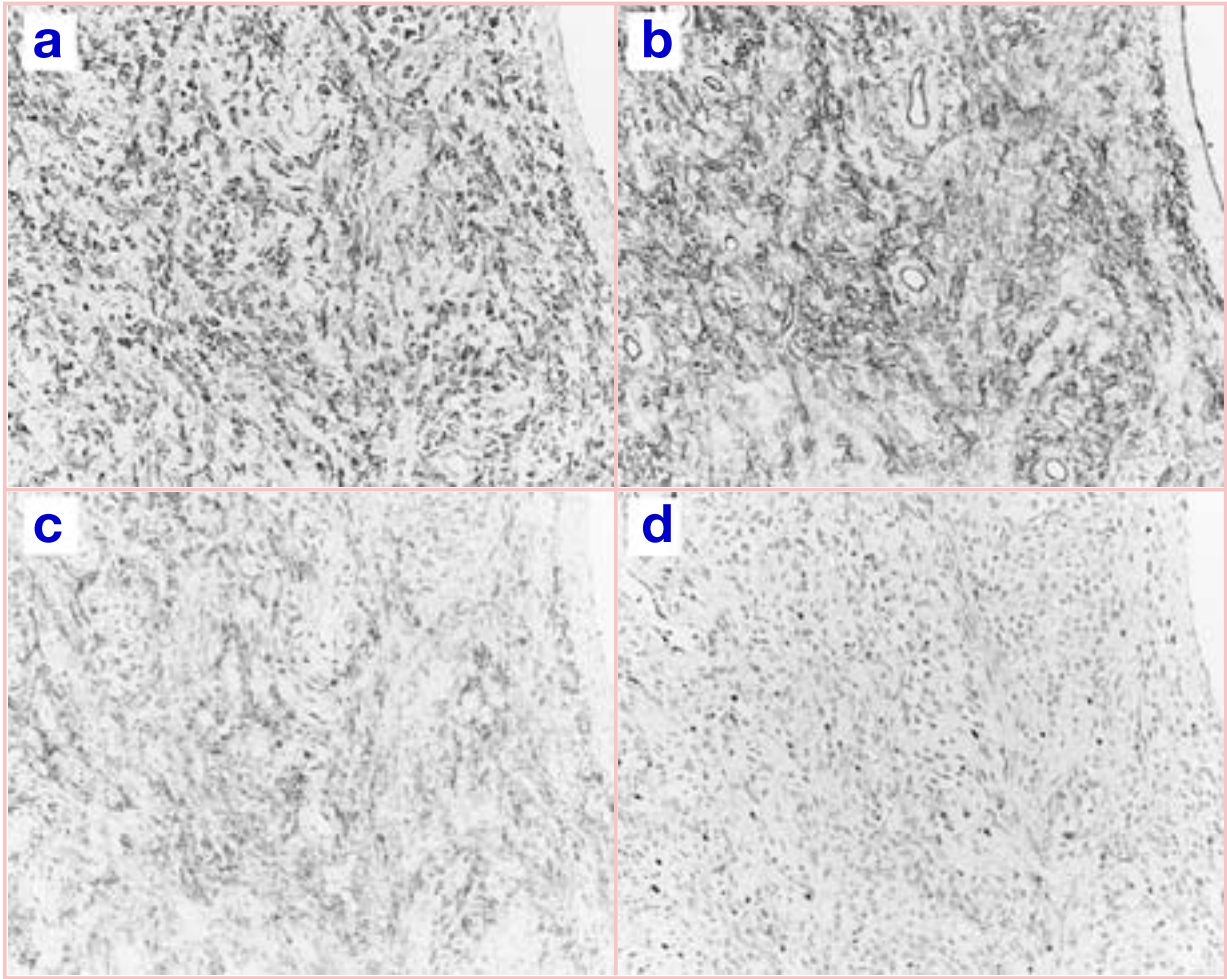
**Fig. 3.** The mass presented as well-circumscribed and had a whitish and firm appearance with focal hemorrhage.



**Fig. 4.** Histopathological findings. Hematoxylin and eosin stain. Bar = 100  $\mu$ m.

- a:** The tumor was composed of an alternating cellular area.
- b:** Thick collagen bundles and hyalin-walled blood vessels were scattered in fibromyxoid stroma.
- c:** Bland spindle or short spindle cells were arranged without an obvious pattern.
- d:** Mitotic figures (arrow) were rare.





**Fig. 5.** The tumor cells were positive for **a)** vimentin, **b)** CD34, **c)** CD99 and focally **d)** bcl-2.

tein, cytokeratin (CK) (AE1/AE3) and epithelial membrane antigen (EMA) were negative (Fig. 5 and Table 1). MIB-1 labeling index was about 2%.

#### ***Genetic diagnosis***

The fusion gene SYT-SSX1 or SYT-SSX2, which is specific in synovial sarcoma (Fisher et al., 2002), was not detected by reverse transcriptase-polymerase chain reaction.

#### ***Pathological diagnosis***

This case was finally diagnosed as SFT of the left thigh, based on clinical findings and histologically bland spindle or short spindle cells arranged without an obvious pattern, with scattered thick collagen bundles, thin- or hyalin-walled blood

vessels, hemangiopericytoma-like appearance and immunohistochemical positivity for CD34 and CD99.

#### **Discussion**

SFT is a ubiquitous mesenchymal tumor of possible fibroblastic type, showing a prominent haemangiopericytoma-like branching vascular pattern (Guillou et al., 2002). SFT was first described by Klemperer and Robin in 1931 as a pleura-based lesion (Klemperer and Rabin, 1931). After that, SFT began to be reported as extrapleural SFT in various locations. In the latest World Health Organization classification, extrapleural SFT is classified into the intermediate (rarely metastasizing) cate-

gory (Fletcher et al., 2002). SFTs usually develop in the pleura, but are rarely of extrapleural origin. Extrapleural SFTs may be found at any location in middle-aged adults between 20 and 70 years (median 50 years), with no sex predominance. Approximately 40% of extrapleural SFTs are located in the subcutaneous tissue, while others are located in the deep soft tissue of the extremities or extracompartmentally in the head and neck region (especially the orbit), thoracic wall, mediastinum, pericardium, retroperitoneum and abdominal cavity. Other described locations include the meninges, spinal cord, periosteum, as well as organs such as the salivary glands, lungs, thyroid, liver, gastrointestinal tract, adrenal glands, urinary bladder, prostate and spermatic cord and testes (Guillou et al., 2002). Fifteen cases of SFTs in the thigh have been reported (Suster et al., 1995; Hasegawa et al., 1996, 1999; Brunnemann et al., 1999; de Saint Aubain Somerhausen et al., 1999; Morimitsu et al., 2000; Nakamura et al., 2005; Anders et al., 2006; Hoshino et al., 2007; Inoue et al., 2007; Martorell et al., 2007). Interestingly, it has been reported that SFTs are associated with hypoglycemia due to secretion of insulin-like growth factors by the tumor in about 5% of cases (Fukasawa et al., 1998). MRI is not sufficient for final diagnosis. Therefore, pathological findings are very important (Anders et al., 2006).

Histopathologically, most SFTs present as well circumscribed, often partially encapsulated masses. Typical SFTs show a patternless architecture characterized by a combination of alternating hypo- and hypercellular areas, separated from each other by thick bands of hyalinized, somewhat keloidal collagen and branching haemangiopericytoma-like vessels. Myxoid changes, areas of fibrosis and interstitial mast cells are commonly observed. Mitotic figures are generally scarce. Immunohistochemically, tumor cells in SFTs are known to be immunoreactive for CD34 (90–95% of cases) and CD99 (70%) as confirmed in the present tumor. Approximately 20% to 35% of the tumors are also variably positive for EMA, bcl-2 and SMA (Guillou et al., 2002).

**Table 1. Results of immunohistochemistry**

Antibodies	Source	Dilution/ antigen retrieval	Results
Vimentin	Nichirei	PD / mw	+
CD34	Nichirei	PD / mw	+
CD99	Dako	1:50/ mw	+
Bcl-2	Dako	PD / mw	Focally +
EMA	Nichirei	PD / mw	–
CK (AE1/AE3)	Dako	1:50/ mw	–
S-100 protein	Nichirei	PD / mw	–
Desmin	Dako	1:50/ mw	–
Alpha-SMA	Dako	1:50/ mw	–
HHF-35	Dako	PD / mw	–

+, positive; –, negative; mw, microwave; PD, prediluted.

In the present case, clinical and histopathological findings at operation were typical, as described above. Differential histopathological diagnoses were monophasic synovial sarcoma and MPNST. CD34 is usually negative, CK and EMA are usually positive, and stromal collagen is usually wiry and scanty in monophasic synovial sarcomas (Fisher et al., 2002). The fusion gene SYT-SSX1 or SYT-SSX2 was not detected in the present tumor. We could also distinguish this case from MPNST by the following criteria: S-100 protein is usually positive, CD34 is usually negative, the cells are arranged in sweeping fascicles and palisading, and mitotic figures are readily encountered in MPNST (Woodruff et al., 2000). On the other hand, histopathological findings were atypical upon needle biopsy before surgery. The needle biopsy specimen was composed of a cellular area, but thick collagen bundles and hyalin-walled blood vessels were not conspicuous. However, if we had noted that CD34 was strongly positive and that there were a few collagen bundles in the needle biopsy specimen, we might have been able to diagnose SFT before surgery in the present case.

Morphologic distinction between benign and malignancy SFT is often difficult. Malignant SFT are usually hypercellular lesions, showing at least focally moderate to marked cytological atypia, tumor necrosis, numerous mitoses (> 4 mitoses/10

HPF) and/or infiltrative margins. Although there was no convincing evidence of malignancy in the present case, the biological behavior of the lesion is difficult to predict because rare cases show abrupt transition from conventional benign-appearing SFT to high grade sarcoma.

We have presented a case of SFT located in the thigh, which is relatively rare. A rare location of an uncommon lesion led to a confusing diagnosis. Although it may be rare, extrapleural SFT should be considered in the evaluation of soft tissue tumors in the extremities.

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*Received July 4, 2008; accepted August 4, 2008*

*Corresponding author: Kensaku Yamaga, MD*