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A recurrent solitary fibrous tumor of the thigh with malignant transformation: A case report



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ABSTRACT

INTRODUCTION: We describe an unusual case of a uniformly high-grade malignant solitary fibrous tumor (SFT) of the thigh with recurrence after wide resection in a 31-year-old man.

PRESENTATION OF CASE: Our current case showed a long-term benign course before the operation, although the subcutaneous tumor was larger than 10 cm at presentation. The SFT was diagnosed by needle biopsy, and wide resection was performed. Histological findings showed proliferation of capillaries surrounded by masses of spindle-shaped cells without any cytologic atypia, and the percentage of MIB-1-positive nuclei was 2.1%. However, a rapidly enlarging recurrent tumor was observed 11 months after the operation. A second wide resection for the recurrent tumor was performed. Histologically, the tumor cells uniformly displayed significant cytologic atypia and pleomorphism, and had 40–50 mitoses per 10 high-power fields. The proportion of MIB-1-positive nuclei was 48%. Consequently, the tumor was diagnosed as a SFT with malignant transformation.

DISCUSSION: The malignant transformation described in past studies showed high-grade areas within benign, low-grade, or intermediate-grade SFTs. Therefore, in contrast to our case, uniformly high-grade malignant histological findings at recurrence were not described.

CONCLUSION: Even if a tumor is non-malignant during the clinical course, as confirmed by tissue biopsy, the possibility of tumor progression to high-grade sarcoma at recurrence should be considered, and the treatment strategy should be determined carefully.

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1. Introduction

Solitary fibrous tumors (SFTs) are relatively unusual fibrous neoplasms first described as a distinctive tumors arising from the pleura [1]. SFTs have been reported at a wide range of anatomic sites as extrapleural SFTs, but reports of extrapleural SFTs at the extremities are rare [2]. SFTs at the extremities are frequently located deep within the tissue [3]. It is known that SFTs may show regions of high-grade atypia within the tumor.

Herein, we report about a patient with a recurrent SFT in the thigh with malignant transformation.

2. Presentation of case

A 31-year-old man presented with an 11-year history of a gradually enlarging soft-tissue mass in his right thigh. The patient reported no antecedent trauma and denied experiencing fevers, chills, or changes in weight. He never had hypoglycemia symptoms. Physical examination revealed a non-tender, 10- × 6.5- × 13-cm elastic soft, mobile mass overlying the lateral aspect of the right distal thigh (Fig. 1). There was a crust on the surface of the mass, and gradual bleeding was observed in the skin. Magnetic resonance imaging (MRI) showed low-to-intermediate signal intensity on T1-weighted images and heterogeneous intermediate-to-high signal intensity on T2-weighted images. Gadolinium-enhanced MRI showed heterogeneous high signal intensity (Fig. 2). Radiological examination showed that the tumor was located in the subcutaneous tissue. Needle biopsy was performed, and the lesion was subsequently diagnosed as a SFT. Resection of the tumor with 1 cm of skin around the tumor edges and fascia was performed. Histological examination of the tumor revealed the proliferation of capillaries surrounded by masses of spindle-shaped cells without any cytologic atypia (Fig. 3). Most parts of the tumor displayed high

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Fig. 1. Gross features of the tumor at presentation. A large mass can be observed on the lateral aspect of the right distal thigh.

cellularity, and the tumor cells had 1–4 mitoses per 10 high-power fields. On immunohistochemical analysis, the tumor was negative for cytokeratin, S-100 protein, alpha-smooth muscle actin, but was positive for CD34 and bcl-2. The percentage of MIB-1-positive nuclei was 2.1%. Consequently, the tumor was diagnosed as a SFT.

After the initial operation, the patient was regularly followed up using MRI. However, a rapidly enlarging recurrent tumor was observed 11 months after the operation (Fig. 4A). MRI of the tumor showed an 8- × 8-cm mass located in the subcutaneous tissue of the anterior surface of the right distal thigh (Fig. 4B). Needle biopsy of the recurrent tumor was performed, and histologic examination showed pleomorphic tumor cells with significant cytologic atypia, findings compatible with malignant SFTs. A second wide resection of the recurrent tumor and reconstruction using a free skin graft were performed. On histological examination, the tumor cells uniformly displayed significant cytologic atypia and pleomorphism, and had 40–50 mitoses per 10 high-power fields (Fig. 5). On immunohistochemical analysis, 48% of the nuclei stained positive for MIB-1. Consequently, the tumor was diagnosed as a malignant SFT with malignant transformation. Additional adjuvant radiotherapy (66 Gy/33 fractions) was performed, although a wide margin was obtained upon assessment of the resected specimen. There was no recurrence after surgery; however, lung metastases were detected at 8 months, and the patient died of disease 17 months after the second operation.

3. Discussion

SFTs represent a spectrum of mesenchymal tumors encompassing tumors previously termed hemangiopericytomas, which are classified as having intermediate biological potential (rarely metastasizing) in the 2013 World Health Organization classification scheme [4]. A SFT was first described in 1931 by Klemperer and Rabin in a patient with a distinctive pleural lesion [1], and was

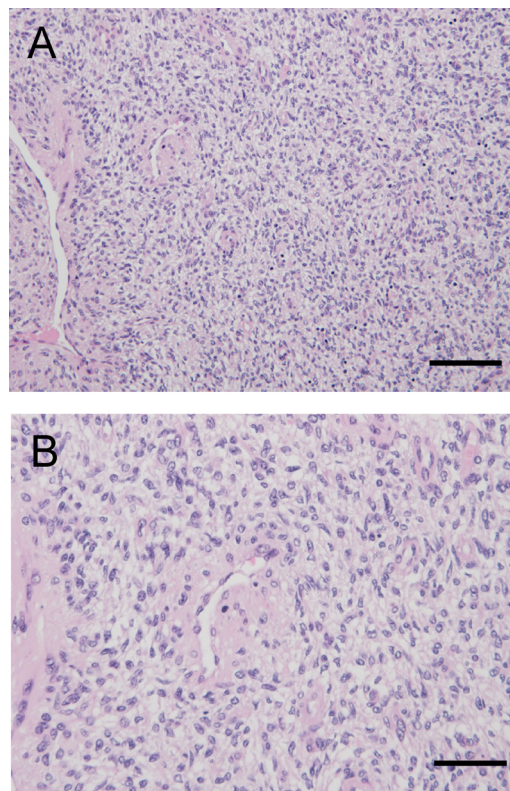


Fig. 3. Histological findings of the primary tumor (hematoxylin and eosin staining). Proliferating capillaries are surrounded by masses of spindle-shaped cells without any cytologic atypia. (A) Scale bar: 100 μ m. (B) Scale bar: 50 μ m.

considered to be exclusively located in the thoracic cavity similar to pleural fibrous tumors. However, SFTs have been recently reported at many extrapleural locations [4]. Nevertheless, SFTs of the extremities are rare. Gold et al. reported only two cases (2%) of SFTs arising from the extremities in 79 evaluated SFTs [2]. SFTs are generally slow-growing neoplasms that have a favorable prognosis, but approximately 10% of SFTs can show aggressive behavior with local recurrence and distant metastasis observed during the course of long-term follow up [5–7]. Therefore, wide resection is usually the preferred surgical treatment for SFTs. A positive surgical margin, the presence of a histologically malignant component, and a tumor size greater than 10 cm are reported risk factors predicting worse clinical outcome [4]. These clinically aggressive tumors with local recurrence and distant metastasis display histological characteristics such as hypercellularity, cellular atypia, tumor necrosis, >4 mitoses/10 high-power fields, and infiltrative margins, and are defined as malignant SFTs [8]. It has been reported that these findings were compatible with SFTs of the pleura. Specifically, a high

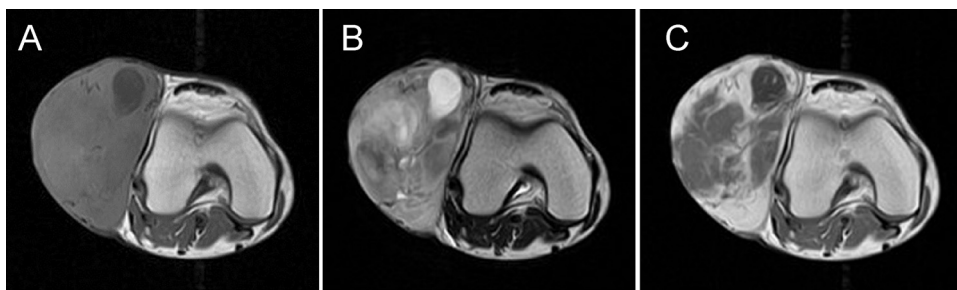


Fig. 2. Magnetic resonance imaging findings before initial surgery. (A) An axial T1-weighted image, (B) an axial T2-weighted image, (C) and an axial T1-weighted image after intravenous gadolinium enhancement. The tumor is located in the subcutaneous tissue on the lateral side of the distal thigh.

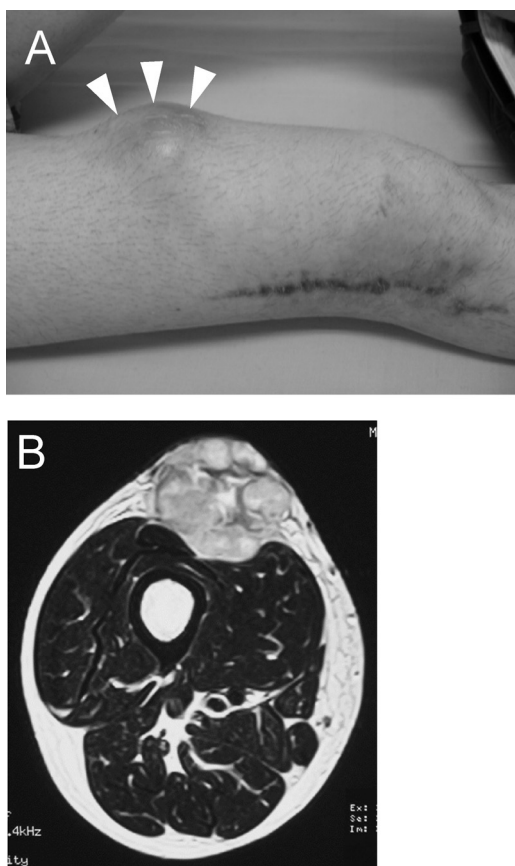


Fig. 4. (A) Gross findings of the recurrent tumor (arrowhead). (B) An axial T2-weighted image showing the recurrent tumor located in the subcutaneous tissue close to the location where initial surgery was performed.

mitotic count is the best indicator of poor outcome [4]. It was found that bFGF and Ki67 labeling indexes are diagnostically relevant for the evaluation of malignant SFTs, and these proteins are thought to be potentially useful markers for prognosis [9]. Nielsen et al. suggested that SFTs at the extremities have histologically malignant potential, and they recommended wide local resection of the tumor and long-term follow up after surgery [10]. However, Hasegawa et al. have suggested that clinically aggressive tumors with local recurrence and distant metastasis had mostly low (<10%) MIB-1 labeling indexes, and cases defined as malignant SFTs did not always show aggressive courses within limited follow up periods [7]. Our current case had a long-term clinical course before the operation even though the tumor was larger than 10 cm at presentation. Histological evaluation of the resected tumor specimen did not display the characteristics of malignant SFTs. However, the tumor recurred within 1 year, and the recurrent tumor had homogeneously high-grade malignant features such as a high mitotic index, atypia, and a high Ki-67 labeling index.

Two types of malignant transformation have been reported; the first is the malignant or high-grade transformation from benign, low-grade, or intermediate-grade SFTs, and the second is the de novo occurrence of malignant SFTs [11]. However, the malignant transformation described in these studies showed high-grade areas within benign, low-grade, or intermediate-grade SFTs. Therefore, in contrast to our case, uniformly high-grade malignant histological findings at recurrence were not described. Furthermore, Ki67 expression was up to 6% in the previously described cases of malignant transformation, whereas the Ki67 expression levels were more than 40% in our case, which was only previously seen in a de novo occurrence of a malignant SFT [11]. Because the current case

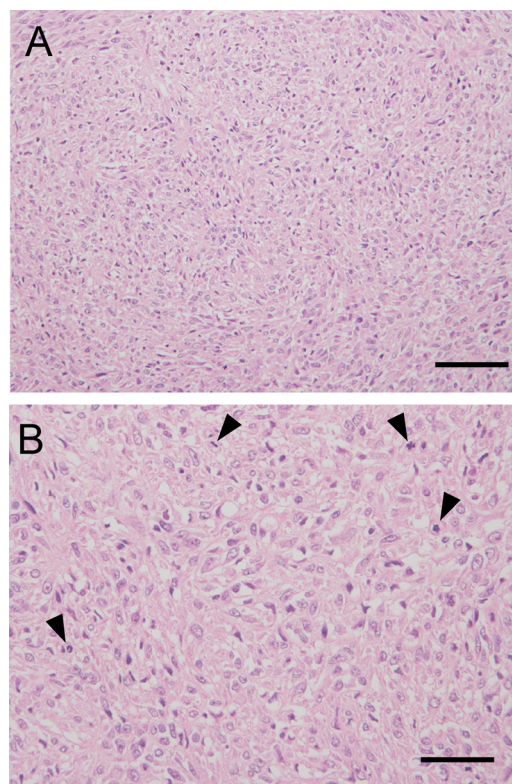


Fig. 5. Histological findings of the recurrent tumor (hematoxylin and eosin staining). Significant cytologic atypia and pleomorphism can be observed. (A) Scale bar: 100 μ m. (B) The tumor has a high mitotic activity (arrowhead). Scale bar: 50 μ m.

showed a slow growing tumor that did not display areas of a malignant SFT but that had recurred within a short period with uniform malignant transformation, it suggests that even if SFTs have benign characteristics, we should be careful about possible progression to a high-grade malignant tumor at recurrence. It has been reported that the pathological grade does not necessarily correlate with the clinical outcome in SFTs [8,12,13]. These discrepancies are also seen in extrapleural SFTs of the extremities.

4. Conclusion

Even if a tumor is non-malignant during the clinical course and if this finding is confirmed using tissue biopsy, we should consider the possibility of tumor progression to high-grade sarcoma at recurrence and determine the treatment strategy carefully.

Conflict of interest

The authors declare that they have no conflict of interest.

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This study received no funds from anybody.

Ethical approval

This study was approved by the ethics committee of Shinshu university hospital (No. 3311).

Consent

Written informed consent was obtained from the parent of the patient for publication of this case report and accompanying images.

Author contribution

Case report writing, data collection, and discussion writing were done by Yasuo Yoshimura and Kenji Sano. Discussion writing was carried by Ken'ichi Isobe, Kaoru Aoki, Munehisa Kito, and Hiroyuki Kato.

Guarantor

Yasuo Yoshimura.

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