healing, smooth muscle loss, and fibrosis [1,2]. An article reported that the most common vasculopathy in NF-1 patients is isolated renal artery disease, followed by mesenteric artery stenosis/aneurysm and aortic dissection. AVM does not occur as frequently, but most cases arise in the cervical vertebral artery and lumbar vertebral artery, and an occurrence in the trunk, extremities, and face is very rare [3]. Systemic manifestations from this vascular pathology, such as hypovolemic shock, are rare, but bleeding is difficult to control once it has started.

NF-1 vasculopathy conditions such as aneurysm or pseudoneurysm are generally treated with transcatheter embolization of the feeding artery, which is usually limited to one or two major vessels. However, AVM has multiple tortuous vessels that feed the mass, which makes vascular intervention difficult. Moreover, in the case that the mass is superficially located and the feeding vessel is too small to perform selective embolization, a surgical approach is preferred.

Although the pathologic diagnosis was not AVM in our case, the massive bleeding nature and clinical diagnosis from the preoperative evaluation included a differential diagnosis of AVM. Based on the finding that the inguinal portion and the pelvic portion of the mass were septated, a bimodal approach combining selective embolization of the pelvic mass and surgical excision of the inguinal portion of the mass was successfully performed.

Here, we have reported successful treatment of a severe case of hypervascular neurofibroma with intractable bleeding, which was successfully controlled with selective embolization and surgical removal followed by reconstruction with a free flap. Moreover, because a hypervascular neurofibroma has clinical features similar to those of an AVM, our bimodal approach could be a good option for the treatment of a huge AVM that is caused as a complication of neurofibromatosis.

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Giant Extrapleural Solitary Fibrous Tumor of the Thigh

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A solitary fibrous tumor (SFT) is an uncommon tumor that arises from primitive fibroblast-like cells in the connective tissue [1]. It characteristically
shows a mixture of fibrous tissue, cellular components, and highly vascularized areas consisting of numerous, closely packed small to medium-sized blood vessels [2,3]. They usually affect adults between the fourth and the seventh decades of life (median, 50 years). Histological findings including immunohistochemistry are required for the diagnosis of SFT. Although SFTs are generally benign, well-circumscribed soft tissue tumors, 10%–15% of SFTs will recur and/or metastasize [4]. SFTs are usually located in the pleura or other serosal surfaces. Despite the fact that they are seldom located in extrapleural soft tissue, this tumor has been reported in a variety of extraserosal sites. The known extrapleural locations of SFT are as follows: the lumbar extradural space, intrameningeal space, cervical spine, deep soft tissue of the neck, orbital space, pelvic space, retroperitoneal space, vagina, thyroid gland, mammary gland, prostate, nasal mucosa, liver, renal pelvis, and extremities. The orbits and the soft tissues of the extremities are the most commonly reported extrapleural sites [1]. SFTs of extremities, particularly in the thighs, are known to have malignant potential [4]. To treat benign SFT, resection with an intact tumor capsule is required for full recovery of the patient. Reviewing the literature, we found no confirmed reasons for a wide resection [4,5]. Here, we report a case of an extrapleural SFT that occurred in the inner thigh area.

A 76-year-old female presented at our department with a left inner thigh soft tissue mass. Physical examination revealed a huge mass measuring 15 cm × 13 cm (Fig. 1). Preoperative magnetic resonance imaging (MRI) revealed a large (about 12.4 cm × 10 cm) lobulated soft tissue mass in the subcutaneous fat layer (red arrow). It shows a predominantly hyperdense mass with heterogeneous regions of low signal intensity.

Fig. 1.
A 76-year-old woman presented with a huge, protruding, firm, and fixed mass measuring 15 cm × 13 cm on her left inner thigh.

Fig. 2.
Coronal contrast-enhanced T2-weighted magnetic resonance imaging scan shows a large (about 12.4 cm × 10 cm) lobulated soft tissue mass in the subcutaneous fat layer (red arrow). It shows a predominantly hyperdense mass with heterogeneous regions of low signal intensity.

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A 76-year-old female presented at our department with a left inner thigh soft tissue mass. Physical examination revealed a huge mass measuring 15 cm × 13 cm (Fig. 1). Preoperative magnetic resonance imaging (MRI) revealed a large (about 12.4 cm × 10 cm) lobulated soft tissue mass with a diffuse hypointense signal on T2-weighted imaging in the subcutaneous fat layer with a bulging contour and peripheral heterogeneous enhancement, which was consistent with a hypervascular tumor (Fig. 2). Total body scan revealed no distant metastasis. The mass was located in the subfascial layer of the inner thigh, and surgical treatment was carried out. The whole tumor was totally excised and submitted for histopathological study. The excised specimen was about 15 cm × 13 cm in size, encapsulated, and well-circumscribed, with a firm, rubbery texture and tan-brown color (Fig. 3). The initial histopathological findings revealed an acellular spindle-cell tumor with nuclear pleomorphism and cellular atypia. They also revealed the proliferation of capillaries surrounded by masses of round or spindle-shaped cells. The cellularity varied considerably in different areas with a predominance of hypercellular areas. However, increased mitotic activity, nuclear pleomorphism, or any foci of coagulative necrosis were not observed in...
this case. Further, immunohistochemistry was performed for the differential diagnosis. It revealed that CD-34 was positive and S-100 protein was negative (Fig. 4). The final histopathological diagnosis was a benign extrapleural SFT. The patient did not receive further radiotherapy or chemotherapy. No recurrence was found 12 months after surgery.

An SFT is a rare neoplasm that derives from mesenchymal cells. The differential diagnosis of an SFT in an extremity includes neoplasms such as fibrosarcoma, fibrous histiocytoma, desmoid tumor, dermatofibrosarcoma protuberans, hemangiopericytoma, neurofibroma, and malignant peripheral nerve sheath tumor [1]. Because of the tumor’s rarity, it usually takes a long time to reach the diagnosis of SFT. Imaging studies like plain radiography and ultrasound are non-specific and not suitable for the differential diagnosis. On MRI, the diagnosis of SFT is suggested by a well-circumscribed mass with smooth margins, and a focal or diffuse hypointense signal on T2-weighted imaging due to the fibrous content in the tumor. An SFT also demonstrates strong focal or diffuse contrast enhancement due to the highly vascularized areas in the tumor [1]. Malignant SFTs are usually demonstrated as hemorrhage, cystic degeneration, and central necrosis on MRI. However, in our case, there was no such evidence suggestive of malignancy. Radiological findings alone cannot definitely determine whether the tumor is benign or malignant [3]. To confirm a diagnosis and to differentiate it from other soft tissue tumors, an immunohistochemical analysis is required. SFTs are a well-circumscribed tumors. Histologically, they consist of a proliferation of capillaries surrounded by masses of spindle-shaped cells. SFT cells are separated by thick bands of collagen, demonstrating foci of keloid-like hyalinization. Prominent vascularity showing a hemangiopericytoma-like vascular pattern and thick, hyalinized vessel walls are seen. Immunohistochemically, SFTs are negative for cytokeratin, S-100 protein, desmin, and alpha-smooth muscle actin, while positive for vimentin and CD34 [2,3]. In our case, immunohistochemical staining was positive for CD34 and negative for S-100 protein, which satisfied the diagnostic criteria for SFTs. Patients with a benign SFT are usually treated with complete surgical excision. The prognosis of this tumor is good and the local recurrence rate is very low in the case of benign SFTs. However, other studies have reported that SFTs in the extremities are more likely to be malignant [4]. Further, immunohistochemical patterns are used for therapeutic decision making. With mitotic activity, increased cellularity, necrotic areas, and nuclear pleomorphism, there is a possibility of malignant SFT. Thus, if there is evidence suggestive of malignant potential, a further wide resection, a long-term follow-up, and regular MRI are proposed. Otherwise,
simple excision with an intact tumor capsule is the optimal treatment of benign SFTs [4,5]. In our case, mitotic activity, nuclear pleomorphism, and central necrosis were not observed. Therefore, simple excision rather than wide excision was sufficient, and there was no evidence of recurrence over the 1 year follow-up period.

In conclusion, although these tumors are an uncommon entity, the possibility of SFTs should be kept in mind during the evaluation of any huge soft tissue mass occurring in the extremities, so that the physician may examine the appropriate differential markers, arrive at an accurate diagnosis, and administer appropriate treatment.

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