



CASE REPORT

# Groin synovial sarcoma with intraluminal femoral sheath involvement<sup>☆</sup>



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## KEYWORDS

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vascular  
reconstruction

**Summary** Synovial sarcomas are highly aggressive soft tissue malignant tumors that occur primarily in young adults. They arise from mesenchymal tissue and typically in or near tendons and tendon sheaths. They can develop at any site but the majority (80–95%) of synovial sarcomas are located in the extremities and mostly at the lower extremities. Groin synovial sarcoma is rare. About 10% of lower limb soft tissue sarcomas have major vessel involvement. In the past, amputation was the standard surgical procedure for lower limb soft tissue sarcoma with major vessels involvement. However, using modern surgical techniques, we can successfully perform a limb-preserving operation with a tumor-free margin. A 17-year-old girl presented with left inguinal synovial sarcoma with femoral artery and vein encasement. The patient received radical tumor resection with major vascular structures. Segmental femoral vessel defects were reconstructed with polytetrafluoroethylene grafts. After revascularization, we designed a pedicled anterolateral thigh fasciocutaneous flap for coverage of the wound and reconstructed vessels simultaneously. The circulation of the lower extremity was good and the girl recovered well after the surgery without morbidity. Synovial sarcoma would potentially be associated with local recurrence and distant metastasis. The presences of bone and neurovascular bundle invasion are adverse predictors of recurrence and mortality. Surgery remains the mainstay of treatment. In the advance of current surgical technique, limb-preserving surgery is possible even for tumor-involved major vessels.

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## 1. Introduction

Soft tissue sarcomas account for approximately 1% of malignant tumors. Most of them arise from the extremities, followed by the trunk wall, retroperitoneum, and head and neck.<sup>1</sup> Synovial sarcoma is an aggressive subtype<sup>2</sup> that occurs primarily in young adults.<sup>3,4</sup> The term “synovial” is derived from its morphologic origin and its histological resemblance, and is easily misinterpreted to mean that the tumor comes from synovium.<sup>5</sup> However, it is a mesenchymal tumor.<sup>2</sup>

The majority of the tumors appear in the extremities, with two-thirds being located in the lower extremities.<sup>2,5</sup> The most common site is within 10 cm of the knee. Extra-articular is dominant and true intra-articular tumor is very rare.<sup>2</sup> It presents as a slow-growing, frequently painful tumor.<sup>2,3</sup> The current standard treatment is wide resection of the tumor, followed by irradiation with or without chemotherapy.<sup>5</sup> Initial adequate radical ablation surgery is still the most important tumor-free prognosis factor.

When soft tissue sarcomas in the extremities involve major vessels, it may become a challenge to achieve a clear margin. In the past, amputation was the mainstay to obtain a cancer-free margin. Here, we report a case of groin synovial sarcoma that encased the femoral vasculature. We cooperated with vascular surgeons for a tumor wide excision and vascular segmental resection followed by vascular reconstruction and pedicled anterolateral thigh flap reconstruction for vessel and soft tissue defect.

## 2. Case report

A 17-year-old girl was in a healthy condition previously. She presented to our outpatient clinic due to a small palpable mass in the left groin area noticed for 2 months, which enlarged gradually. She denied recent trauma to the affected extremity. Physical examination revealed an elastic, firm, and fixed tumor, which was 10 cm in diameter. Its palpation provoked minimal discomfort without numbness or tenderness. Her lower extremity range of motion and muscular strength were normal. Magnetic resonance imaging also revealed a heterogenous tumor both on T1WI and T2WI images at the groin area with contrast enhancement. It also revealed septation and central necrosis. Femoral vessels were wrapped by the tumor (Fig. 1). An incisional biopsy was then performed and the final histologic diagnosis was synovial sarcoma. Further oncological surveillance to evaluate distant metastasis, such as bone scan, chest computed tomography, and chest X-ray all ruled out metastasis. A tumor wide excision was then planned and possible vascular resection will be needed. Due to the indispensability of the femoral artery for the lower extremity, further vascular reconstruction was necessary.

An oncological resection with a cutaneous peripheral safe margin of 2 cm was designed (Fig. 2). Deep plane as far as the rectus femoris muscle fascia was included, sent for frozen section intraoperatively, and revealed to be free of tumor cells. The tumor encased the femoral artery and veins at the arterial bifurcation site, and the femoral nerve was spared (Fig. 3). The involved and estimated resection

vascular length measured 11 cm. The vascular surgeon decided to use a synthetic graft with polytetrafluoroethylene (PTFE) for vascular reconstruction. A 6 mm PTFE graft was first used to make a Y-shaped graft in the back table for arterial reconstruction. We did tumor *en bloc* resection with femoral artery and vein segmental resection. The lower extremity became cold and poorly circulated, so the vascular surgeon started revascularization. The Y-shaped PTFE graft is anastomosis from the femoral artery to the deep femoral artery and superficial artery. Another 8 mm graft was used for vein reconstruction from the proximal femoral vein to distal femoral vein (Fig. 4). No intraoperative heparin was administered. After revascularization, the lower extremity became warm and well circulated. An ipsilateral pedicled anterolateral thigh flap was then designed for resultant inguinal soft tissue defect reconstruction. The pedicled anterolateral thigh flap with skin paddle sized 15 cm × 9 cm with two musculocutaneous perforators, based on the lateral circumflex branch of femoral artery, was harvested (Fig. 4A–C). Partial tensor fascial lata was also included. The flap was rotated through the subcutaneous tunnel to reach the inguinal defect (Fig. 4D). A fascial lata was used for inguinal deep fascia reconstruction and repaired with 1-0 Surgilon (Covidien). The flap donor site was covered with a split-thickness skin graft harvested from the medial thigh.

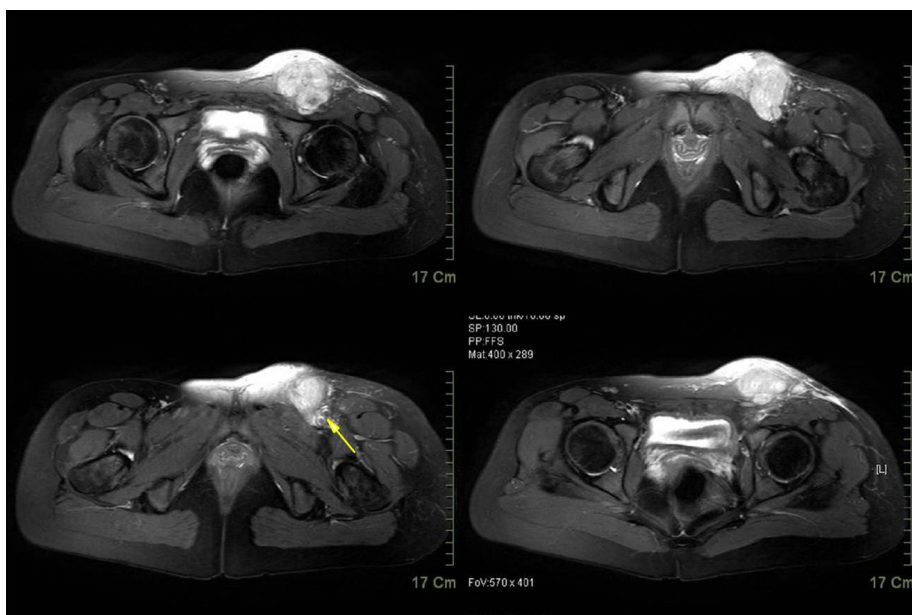
Neither anticoagulant agent nor antiplatelet therapy were given postoperatively. The circulation of the flap and lower extremity were good. The patient was discharged uneventfully.

### 2.1. Outcome

The biopsy specimen grossly revealed a tumor cell intramural invasion (Fig. 5). The final pathology report confirmed synovial sarcoma, biphasic type, with free margins and local regional lymph nodes were negative of malignancy. Pathological pictures revealed spindle-shaped neoplastic cells and focal epithelial components with glandular formation. Vascular invasion and emboli were also noted. Immunohistochemically, both the spindle and epithelial components were positive for epithelial membrane antigen (Fig. 6). Adjuvant radiotherapy and chemotherapy were suggested, but the patient and her parents refused radiotherapy. Therefore, chemotherapy with epirubicin and cisplatin were started 1 month after the operation. Six courses of chemotherapy were applied until abandoned at the patient's wish. During follow-up for 14 months, neither lower limb swelling, synthetic graft thrombosis, nor wound infection were detected (Fig. 7).

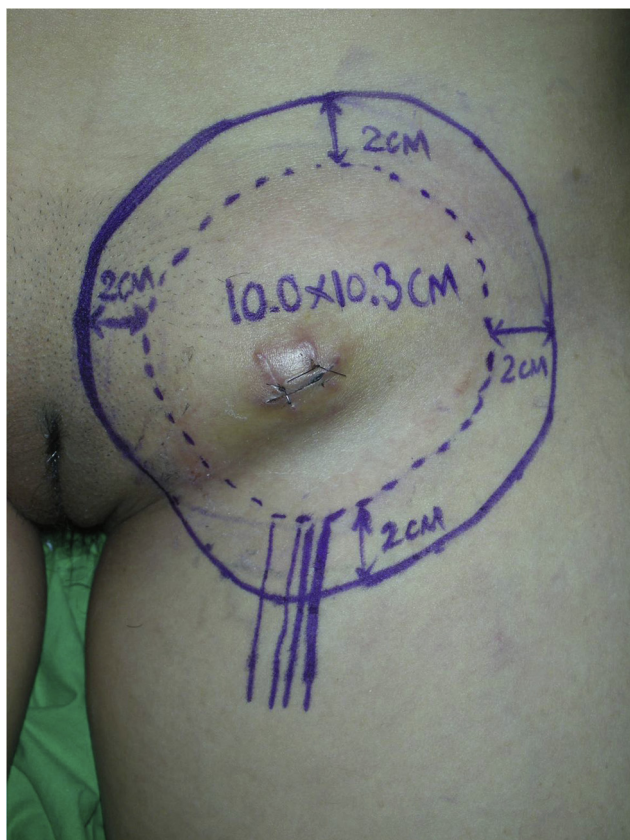
## 3. Discussion

Synovial sarcomas are malignant soft tissue sarcomas comprising approximately 6–9% of all adult soft tissue sarcomas. There is a wide age range at presentation, but young adults are affected most often.<sup>6</sup> There is no sex predilection. It can develop in any anatomical site; however, the extremities are the most common sites and the majority occur in the lower extremities. It mostly develops



**Figure 1** Magnetic resonance imaging revealed a heterogeneous tumor, 50 mm × 48 mm, located in the groin region characterized by septation, central necrosis, and encased femoral neurovascular bundle with femoral vessel intraluminal invasion (arrow).

at para-articular regions with little risk of intra-articular involvement.<sup>7</sup> Synovial sarcomas have been shown to develop nodal metastases more frequently than other soft tissue sarcomas, with an incidence of 10–12%.

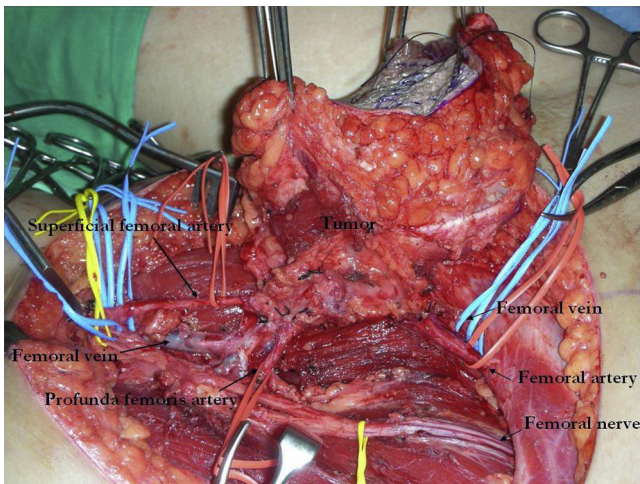


**Figure 2** Perioperative picture shows an elevated, firm tumor over the groin area. Oncological resection with a 2-cm safe margin was then designed for a tumor wide excision.

Despite its name, synovial sarcoma is not derived from synovial tissue. The name is derived from its microscopic resemblance to synovium. It is considered a high-grade sarcoma and is composed of two morphologically distinct cell types: spindle cells and epithelioid cells. These cells are in various proportions leading to classification into three subtypes: biphasic, monophasic, and poorly differentiated.<sup>3</sup> Synovial sarcomas consistently stain positive for cytokeratin and epithelial membrane antigen. These markers are strongly positive for epithelial components and patchy staining can help distinguish from carcinomas. Bcl-2 is positive in synovial sarcoma and was thought to be unique at first, but other nerve sheath tumors also presented Bcl-2 positive. S100 and CD99 are also positive in synovial sarcoma but also would be confused with Ewing's sarcoma or malignant peripheral nerve sheath tumor. Other antibodies, actins (muscle-specific actin,  $\alpha$ -smooth muscle actin), desmin, CD34, and CD31 are negative in synovial sarcoma.<sup>3,6</sup> In our case, the spindle and epithelial components were positive for cytokeratin and epithelial membrane antigen, and negative for desmin. Those characteristics are compatible with synovial sarcoma.

In molecular genetics, most synovial sarcomas (90%) demonstrate a specific translocation between chromosome X and chromosome 18 (p11.2; q11.2). This includes the SYT gene on chromosome 18 and one of several homologous genes (*SSX1*, *SSX2*, and *SSX4*) on the X chromosome. This leads to the expression of the unique SYT/SSX fusion protein in all cases of synovial sarcoma, which suggests a crucial role of the fusion protein in the etiology of this tumor.<sup>3,6</sup>

Clinically, the patient typically presents with a slow-growing mass that may grow over weeks to months. Deeper lesions, which are difficult to detect, may become very large. The groin area is a rare anatomical site for the tumor. In a study reported by St Jude Hospital in 2009, among 300 nonrhabdomyosarcoma soft tissue sarcoma young patients,



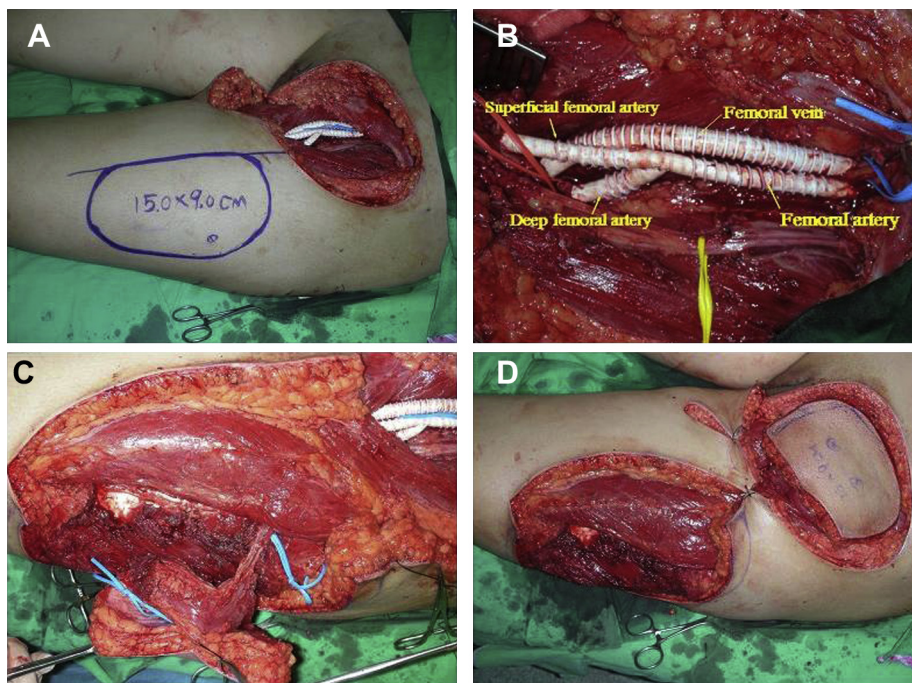
**Figure 3** Intraoperative picture showing the tumor-encased femoral artery and vein at the arterial bifurcation region and to such an extent that dissection to free them was impossible. The femoral nerve was spared.

only six were diagnosed with groin synovial sarcoma.<sup>8</sup> Image studies can appear as multilobular, septated masses that show heterogenous signal intensity on T1WI and T2WI on magnetic resonance imaging. Heterogenous contrast enhancement is caused by central necrosis, calcifications, and hemorrhage within the tumor.<sup>1</sup> Three components are classically described: cellular with an

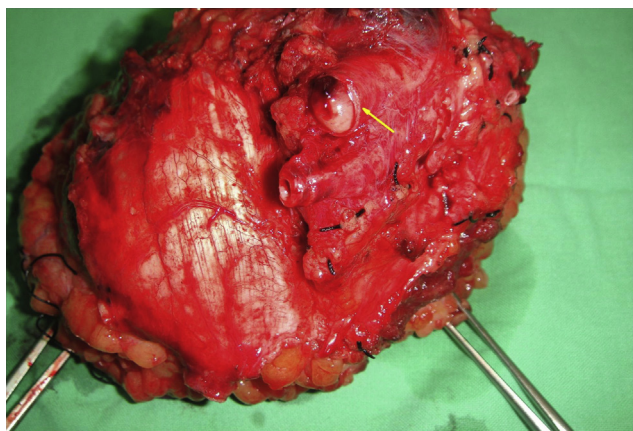
intermediate signal; necrotic or hemorrhage with a high signal; fibrous or calcified with a hypointensity, and this is known as the “triple sign”.<sup>2,7,9</sup> Slow growth of the tumor can result in superficial pressure cortical erosion, periosteal reaction, and osteoporosis.<sup>2</sup>

The primary treatment of synovial sarcoma is surgery, comprising wide excision with negative margins. It should be carried out through normal adjacent tissue planes. Adjuvant radiotherapy is considered helpful for local recurrence control. Synovial sarcoma demonstrates moderate chemosensitivity and approximately 50% response rate due to doxorubicin-based regimens.<sup>10</sup>

Synovial sarcomas involving major vessels, such as the axillary artery and femoral artery were thought unresectable in the past.<sup>11</sup> In a review of the literature, only 5–10% of soft tissue sarcoma infiltrated or encased vascular structures.<sup>12</sup> Historically, amputation of limbs was considered the only resolution to obtain negative surgical margins. With the advancement of operative technique and available grafts, limb preservation with tumor and vascular *en bloc* resection and reconstruction with saphenous vein graft, femoral venous grafts, or synthetic grafts can be achieved.<sup>12</sup> The first description of sarcoma resection in the extremity with arterial replacement was reported by Fortner et al<sup>13</sup> in 1977. There were several studies showing that limb salvage combined with vascular reconstruction offers rates of local control and overall survival that are comparable to amputation.<sup>14</sup> Estimation of resection length is based on a preoperative image study, which is helpful for identifying the appropriate graft to choose.<sup>15</sup> In a large



**Figure 4** (A) Ipsilateral pedicled anterolateral thigh flap with skin paddle 15 cm × 9 cm designed for soft tissue defect coverage. (B) The vascular surgeon used a synthetic polytetrafluoroethylene graft for vascular reconstruction and made a Y-shaped graft in the back table for arterial reconstruction. Another straight graft was used for femoral vein reconstruction. (C) Two musculocutaneous perforators, based on revascularized lateral circumflex branch of the deep femoral artery were included. (D) The flap was rotated through the rectus femoris muscle to the inguinal region.



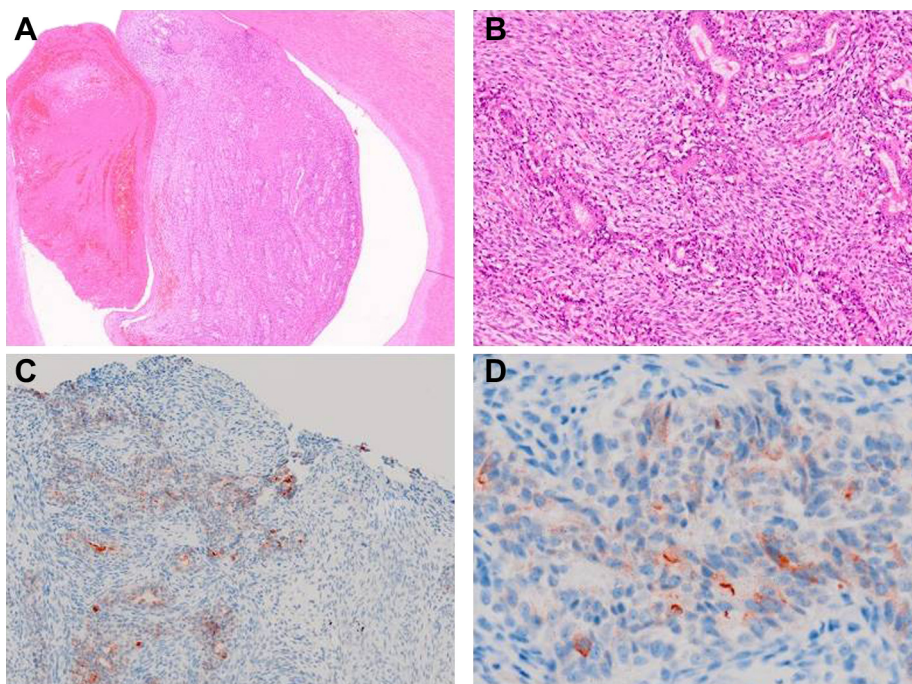
**Figure 5** The biopsy specimen reveals tumor thrombi with intraluminal invasion (arrow).

series presented by Schwarzbach et al<sup>11</sup> in 2005, in the iliofemoral and femoropopliteal region above the knee, they used predominantly synthetic grafts for arterial reconstruction. This can reduce operative time and preserve the greater saphenous vein (GSV). Long-term patency rates between the synthetic graft and autologous graft were identical. The venous reconstruction protocol is controversial. Some studies suggest that if ipsilateral GSV is preserved and patent, the remaining femoral vein stumps could be ligated. However, if the GSV is sclerotic or resected together, reconstruction should be performed. Recently, however, venous reconstruction is associated



**Figure 7** Eight months after the surgery, the patient recovered well and no lower limb swelling, graft thrombosis, or wound infection were found. She could ambulate without difficulty.

with better outcome, and less morbidity such as extremity swelling, seroma formation, and wound infection is seen. In Schwarzbach et al's<sup>11</sup> series, venous patency rate was 54.9% with PTFE graft reconstruction. Due to the rapid



**Figure 6** (A) Hematoxylin and eosin stain, 40 $\times$ : vascular invasion with tumor emboli. (B) Hematoxylin and eosin stain, 100 $\times$ : this section shows a sheet of spindle-shaped neoplastic cells and focal epithelial components with glandular formation. (C) Immunohistochemistry, 100 $\times$ : epithelial membrane antigen positive. (D) Immunohistochemistry, 400 $\times$ : both spindle and epithelial components are positive for epithelial membrane antigen.

formation of collateral drainage, occlusion of a venous bypass several weeks after surgery usually would not require a second revision operation. In our case, we performed both arterial and venous reconstruction with synthetic grafts due to the long segment of vascular and complicated soft tissue defect. Cautious administration of low-dose heparin intraoperatively is sufficient for occlusion prophylaxis. Further oral form anticoagulant or antiplatelet therapy was prescribed only for selected patients.<sup>16</sup> The added risk of complications (primarily bleeding) with vascular procedures performed on patients treated with aspirin is measurable.<sup>17,18</sup> In our case, no heparin injection or postoperative anticoagulant and antiplatelet therapy were given because the girl was young and medically healthy without such myocardial or cerebral risk factors. After 8 months, no sequelae of lower limb swelling, circulation compromised, or wound infection occurred. Adjuvant radiotherapy may have potential adverse effects on the reconstructed graft, such as occlusion or arterial blowout. A radiation dose exceeding 50 Gy can induce vessel wall abnormality. Nevertheless, there is still no published series about the effect of radiotherapy and extremity vascular reconstruction following a sarcoma surgery.

Approximately 25% of patients have metastases at presentation, frequently to the lung. Other common metastatic sites include bone and lymph nodes. In the literature, 5-year survival ranges from 25% to 60%. Multiple studies have identified prognostic factors and these predictive values remain controversial and constantly evolving.<sup>9,19</sup> Incidence of metastases at diagnosis was thought to have a strong correlation to poor prognosis. Tumor size greater than 5 cm has a significant correlation to survival.<sup>19</sup> In primary extremity synovial sarcomas that are larger than 5 cm, distant metastasis rate is over 50% and resultant 5-year tumor-related mortality is almost 40%. Tumor invasion of adjacent bone, nerve, and vascular structures are also adverse prognostic predictors. From the molecular genetics aspect, many studies also highlight a worse prognosis for patients with the SYT/SSX1 translocation than those with SYT/SSX2 translocations.<sup>4,20</sup>

In conclusion, synovial sarcoma is an uncommon high-grade soft tissue sarcoma. It is prevalent in young adults and occurs near joints. It has a high tendency for local recurrence and distant metastasis if major vessels are involved. First-time surgical procedure of radical wide excision with enough clear margins has significantly improved the prognosis. With the advance of current surgical techniques, limb-preserving surgery is feasible even when intramural invasion is diagnosed.

We herein presented a case with rare groin synovial sarcoma with intramural femoral vessels involvement; limb-preserving surgery, including complex vascular and soft tissue reconstruction, was performed successfully.

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