Intravascular synovial sarcoma of the external iliac vein and reconstruction with the superficial femoral vein

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Intravascular synovial sarcoma is a rare neoplasm that arises in large veins of the junctional zone between the proximal leg and lower trunk in adult women. Herein we report the first case of an intravascular synovial sarcoma of the external iliac vein. It was successfully treated with neoadjuvant radiotherapy, radical excisional surgery, and combined arterial and venous reconstruction. Intravascular synovial sarcoma should be considered in the differential diagnosis of any adult woman with deep venous thrombosis and a mass of the femoral or iliac venous system. (J Vasc Surg 2005;42:365-7.)

Synovial sarcoma (SS) is a rare tumor that comprises 5% to 10% of all soft tissue malignancies.1 Its approximate incidence in the United Kingdom and United States is 2 or 3 per million population per year.2 Classic SS involves large joint tendons, bursas, or capsules in the extremity.3 This is the first report of an intravascular SS of the external iliac vein.

CASE REPORT

A 56-year-old woman presented with swelling and pain in her right leg. Doppler ultrasound scanning confirmed thrombosis of the right common and external iliac veins. A computed tomographic scan showed a soft tissue mass deforming the external iliac vein just proximal to the inguinal ligament (Fig 1). The vein was completely obstructed, and numerous venous collaterals existed. Fine-needle aspiration cytology of the mass was consistent with a biphasic SS. A ventilation/perfusion scan revealed asymptomatic bilateral pulmonary emboli.

Anticoagulant therapy was initiated, and a spacer was placed in the right pelvis to protect the small bowel from radiotherapy injury before definitive surgery. A dose of 5000 cGy was administered in 25 fractions and was well tolerated. Surgery followed 6 weeks later.

At operation, a mass arising from the right external iliac vessels extended posteriorly and inferiorly to the obturator fossa. Excision was en bloc and included a 1-cm cuff of normal external iliac artery and vein about the mass, peritoneum, pelvic fascia, and a 1-cm cuff of psoas muscle posteriorly. The right ureter, obturator, and femoral nerves were preserved. A segment of the left superficial femoral vein was harvested to maintain the profunda femoris nerve. A segment of the left superficial femoral nerve was preserved. A segment of the left superficial femoral vein was harvested to maintain the profunda femoris nerve and vein about the mass, peritoneum, pelvic fascia, and a 1-cm cuff of psoas muscle posteriorly. The right ureter, obturator, and femoral nerves were preserved. A segment of the left superficial femoral vein was harvested to maintain the profunda femoris nerve and vein about the mass, peritoneum, pelvic fascia, and a 1-cm cuff of psoas muscle posteriorly. The right ureter, obturator, and femoral nerves were preserved. A segment of the left superficial femoral nerve was preserved. A segment of the left superficial femoral vein was harvested to maintain the profunda femoris nerve and vein about the mass, peritoneum, pelvic fascia, and a 1-cm cuff of psoas muscle posteriorly. The right ureter, obturator, and femoral nerves were preserved. A segment of the left superficial femoral nerve was preserved.

Histology confirmed a biphasic epithelioid and spindle cell neoplasm focally extending into the vein wall (Fig 3). The neoplasm was confluent with organizing thrombi at its longitudinal apices. Epithelioid areas were acinar with pseudopapillae, luminal mucin, and rare lamellar psammoma bodies. Spindle cells, juxtaposing epithelial regions, were arranged in loose fascicles with focal hyalinization and scant ropy collagen deposition. No osteoid formation and no poorly differentiated areas were seen.

Immunohistochemistry. Antibodies studied were carcinoembryonic antigen (1:20,000; Dako Diagnostics, Mississauga, ONT, Canada), epithelial membrane antigen (EMA, 1:200; Dako Diagnostics), cytokeratin AE1/3 (prediluted; Ventana Medical Systems, Tucson, Ariz), S-100 (Ventana; prediluted), and vimentin (Ventana; prediluted). The Envision anti-mouse and antirabbit antibody (Dako, Copenhagen, Denmark) and the DAB-Plus kit (Dako Diagnostics) were used. Epithelioid cells were carcinoembryonic antigen, EMA, pancytokeratin AE1/3, and vimentin positive. Spindle cells were vimentin positive and weakly EMA and AE1/3 positive. S-100 focally stained both cell types.
Reverse transcriptase-polymerase chain reaction. The gene primers SSA and SSX were used; these express an 87-base pair product of an SYT-SSX1 or SYT-SSX2 gene fusion. Paraffin-embedded tumor was the substrate. Polymerase chain reaction products were Southern-blotted onto nylon membranes and probed with an end-labeled internal oligonucleotide for the SYT-SSX fusion gene. A chromosomal t(X;18) translocation was identified in the tumor.

DISCUSSION

SS is a soft tissue malignancy with a propensity for the extremities. The neoplasm has a peak incidence between 15 and 35 years, has a slight male preference, and almost always manifests before the sixth decade of life. Symptoms of a localized mass effect and tenderness develop insidiously and delay clinical presentation. More than 60% involve the lower limb, and 20% involve the arm. Most of the remaining are trunk or head and neck lesions. Unusual sites for primary SS include intra-articular, parapharyngeal, laryngeal, pleural, chest wall, retroperitoneal, cardiac, and genitourinary locations.

Only three intravascular SS have been previously reported. One SS arose in the femoral vein of a 34-year-old woman; the vein was filled with thrombus, and a 5-cm wall segment in the mid thigh was replaced by tumor. The mass was locally excised, and a patch from the long saphenous vein closed the defect. The tumor recurred 5 years later, and this necessitated a formal reconstruction. Both the femoral artery and vein were then widely resected, and the patient remained well 11 years after diagnosis. The second example was in a 31-year-old woman who presented with acute right abdominal pain. Ultrasound and computed tomographic scanning showed a mass occluding the inferior vena cava. The patient had a cardiac arrest 3 hours thereafter and required emergent embolectomy. Large quantities of clot and tumor were extracted from the cava, heart, and pulmonary arteries. The patient remained hypotensive and died 1 day after operation. An autopsy confirmed an SS arising in the wall of the inferior vena cava. The third case was a SS of the superficial femoral vein in a 34-year-old woman who presented with a painful thigh mass. Magnetic resonance imaging confirmed an adductor canal mass adjacent to the neurovascular bundle. At operation, an SS was arising in the wall of the superficial femoral and profunda femoris vein junction and was completely occluding the former. The involved segment was excised, and adjuvant chemoradiotherapy was administered.

Our case replicates the clinical scenario proposed by Robertson et al in 1997: intravascular SS originates in the large veins of the junctional zone between the proximal leg and lower trunk in adult women. At 54 years, our patient presented at an advanced age compared with cases of SS in general and intravascular SS in particular. Chan et al have suggested that older subjects are prone to SS arising in unusual sites.

Surgery and radiotherapy for truncal and extremity SS results in an overall survival of 51% at 10 years and a 3% 5-year local recurrence rate. Tumor size is a key prognostic factor: those smaller than 5 cm in diameter are associated with a 10-year survival of 88%, whereas those larger than 10 cm have an 8% survival rate. Other adverse prognostic factors include age older than 40 years and areas of poor histologic differentiation. SS is most likely to recur locally within 2 years after excision, but metastasis can occur after a prolonged interval.

The superficial femoral vein is a versatile autologous conduit ideal for arterial and venous reconstruction in limb-sparing oncologic surgery. When combined arterial and venous resection is required, reconstruction of both vessels is desirable to avoid the long-term sequelae of chronic venous hypertension. Use of an autologous substitute for venous reconstruction is advisable to enhance long-term patency. Simple ligation of the venous segment with isolated arterial reconstruction has been used in the acute trauma setting, with subsequent femoral/femoral
In summary, intravascular SS is a rare neoplasm of the lower trunk/upper limb large veins in adult women. Although SS was previously deemed a tumor of the young adult, our case confirms that it can occur in women in the sixth decade of life. This tumor should be considered in the differential diagnosis of any woman with a deep venous thrombosis and a femoral or iliac venous system mass. Local control and long-term survival can be achieved by a combination of preoperative radiotherapy, radical surgery, and appropriate vascular reconstruction with an autologous superficial femoral vein conduit.

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