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SHORT REPORT

Primary Intravascular Synovial Sarcoma of the Femoral Vein in a Male Patient, Case Report

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Abstract Synovial Sarcoma (SS) is an aggressive neoplasm commonly affecting deep soft tissues of the extremities. In rare instances SS can arise in large veins of the lower extremities or trunk. We report the first case of intravascular synovial sarcoma (IVSS) occurring in a male patient. A biphasic tumor was diagnosed by histology and immunohistochemistry. Molecular analysis at RNA level confirmed the diagnosis demonstrating the chromosomal translocation t(X;18)(p11.2;q11.2) in the tumor. Although extremely rare, IVSS should be considered in the differential diagnosis of primary intravascular neoplasms and as a potential cause of deep vein thrombosis and thromboembolism.

Introduction

Synovial sarcoma represents the 5–10% of all soft tissue sarcomas. Synovial sarcoma arises in deep soft tissues of the lower extremities and uncommonly in large veins of the lower extremities and trunk. Five cases of intravenous synovial sarcoma have been reported, all in women.[1]

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Report

A 41-year-old man was admitted to the Emergency Department with sudden dyspnoea and right thigh swelling. Physical examination revealed a mass in the lower third of the right thigh. Duplex ultrasonography showed a highly vascularized lesion adherent to the femoral vascular bundle. Computed tomography angiography demonstrated bilateral embolic-filling defects within the main branches of the pulmonary arteries. Anticoagulation was initiated (enoxaparin 8000 change IU twice daily). Computed tomography (CT) scan of the right lower limb identified a $12 \times 9 \times 4$ cm mass with inhomogeneous contrast enhancement (Fig. 1)

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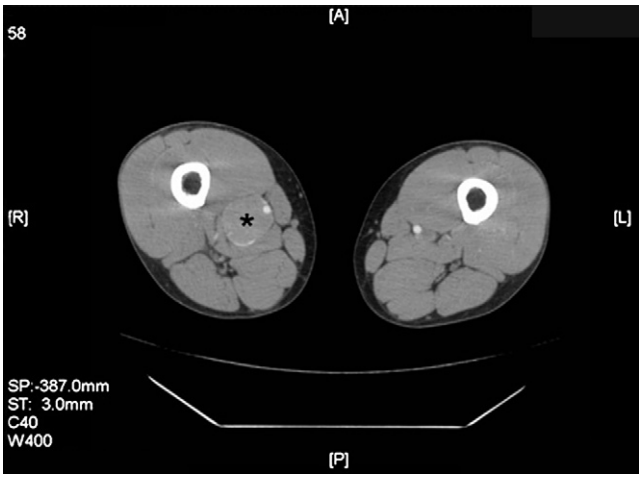

Figure 1 Contrasted-enhanced CT scan shows a 12 × 9 × 4 cm solid mass (asterisk) at the level of the right lower thigh, contiguous with the superficial femoral artery.

and popliteal vein thrombosis, with features suggesting a soft-tissue tumour involving the right femoral vein.

At surgical excision, the mass originated within the femoral vein at the adductor hiatus and involved a lateral duplication of the vein (Fig. 2a). Femoral vessels were isolated and femoral nerves spared. Affected segments of the vein were excised and ligated above the proximal and distal end of the mass. No vascular reconstruction was required. After discharge, the patient received six cycles of adjuvant chemotherapy with epirubicin and ifosfamide.

At 5 months there was no sign of either deep venous thrombosis (DVT) or local recurrence on either CT angiography or duplex scan. Therefore anticoagulation was discontinued. After 1 year the patient remained well with no sign of recurrence or metastasis on CT scan.

Histology showed a high-grade biphasic synovial sarcoma, with epithelioid and small-spindled cells (Fig. 2b). Surgical margins were negative. Immunohistochemistry was performed on a BenchMark XT platform (Ventana Medical System, Tucson, Arizona). Neoplastic cells were negative for CD31, alpha-smooth muscle actin and desmin. Epithelioid cells were cytokeratin (Fig. 2c) and epithelial membrane antigen positive and weakly S100 positive.

RNA analysis from formalin-fixed-paraffin-embedded tissue (Pinpoint Slide RNA Isolation System II, Zymo Research Corp, Orange, CA) showed the presence of the SYT-SSX1 fusion product, resulting from the chromosomal translocation t(X;18)(p11.2;q11.2).

Discussion

Synovial sarcoma is a rare tumor affecting deep soft tissues of the lower extremities near large joints. Uncommon locations include intra-articular, parapharyngeal, laryngeal, pleural, retroperitoneal, cardiac, genitourinary and intravascular sites, particularly large veins of the lower extremities and trunk.[2]

This is the first report of an intravenous synovial sarcoma in a male. Histology and molecular studies indicated that the tumor was biphasic and there was no evidence of tumor embolisation from another site. Since our patient had developed a collateral venous pathway supporting an adequate flow, we undertook en-bloc excision of the involved veins without reconstruction.

Post-operative chemotherapy may offer a survival benefit in high risk patients.

Tumor size $>$ 5 cm in diameter, age $>$ 40 years, areas of poor differentiation and lack of initial gross total resection are adverse prognostic factors, associated with local recurrence and metastasis.[2,3] There is no evidence that adjuvant local radiotherapy is of benefit.[3–5]

Although extremely rare, intravenous synovial sarcoma should be considered in the differential diagnosis of primary vascular neoplasms, leiomyosarcomas, intimal sarcomas and malignant peripheral nerve sheath tumors.

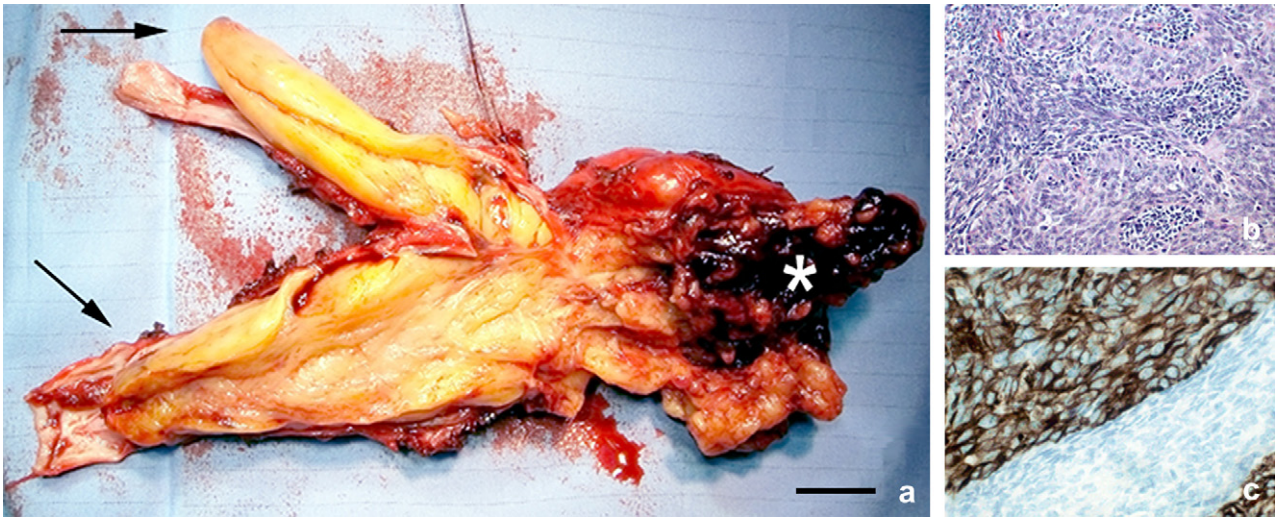

Figure 2 a) Large soft-tissue tumor extended into the right superficial femoral vein (lower arrow) and involved a short lateral duplication of the femoral vein (upper arrow). Popliteal vein thrombosis was also evident (asterisk) (scale bar = 10 mm). b) IVSS displayed a typical biphasic pattern composed of epithelioid and spindled cells (stain, hematoxylin-eosin; 200×). c) Epithelioid cells were cytokeratine positive, whereas spindled cells were negative (stain, pancytokeratin; 400×).

Moreover it should be included in the differential diagnosis of venous thrombosis and thromboembolism.

References

1 Tong GX, Goldblum JR, Qiu W, Skacel M, Downs-Kelly E, Contractor S, et al. Primary intravascular synovial sarcoma: a disease of young adult women? Report of a case diagnosed by aspiration biopsy and review of the literature. *Diagn Cytopathol* 2006;**34**:834—8.

2 White JS, Medlicott SA, Brown H, Moore R, Temple W. Intravascular synovial sarcoma of the external iliac vein and reconstruction with the superficial femoral vein. *J Vasc Surg* 2005;**42**:365—7.

3 Ferrari A, Gronchi A, Casanova M, Meazza C, Gandola L, Collini P, et al. Synovial sarcoma: a retrospective analysis of 271 patients of all ages treated at a single institution. *Cancer* 2004;**101**:627—34.

4 Robertson NJ, Halawa MH, Smith ME. Intravascular synovial sarcoma. *J Clin Pathol* 1998;**51**:172—3.

5 Deshmukh R, Mankin HJ, Singer S. Synovial sarcoma: the importance of size and location for survival. *Clin Orthop Relat Res* 2004;**419**:155—61.