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Femoral Vein Intravascular Synovial Sarcoma Mimicking Primary Deep Vein Thrombosis—A Rare Cause of Deep Vein Thrombosis

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Abstract

Synovial sarcomas are rare malignant mesenchymal soft tissue tumors. We presented the case of a 53-year-old woman patient presenting with acute deep vein thrombosis, later diagnosed as a deep synovial sarcoma of the femoral vein wall. The tumor was identified through cross-sectional magnetic resonance angiography and computed tomography, followed by ultrasound-guided core biopsy. The case report emphasized the importance of considering the possibility of an intravascular neoplasm mimicking thrombus, particularly if calcifications, vein expansion with intravascular cystic spaces, fluid–fluid levels, and septations within a thrombosed vein are seen in imaging.

Keywords

- deep vein thrombosis
- femoral vein malignancy
- intravascular synovial sarcoma
- intravascular tumors
- synovial sarcoma

Introduction

Sarcomas are the most frequent type of primary vascular tumors. Synovial sarcoma is a distinct type of sarcoma with specific clinical and morphological characteristics that typically affects teenagers and young adults, often develops from soft tissues near joints, especially in the knee area. Here, we present a case of an uncommon location of synovial sarcoma that originated from the femoral vein wall presenting as deep vein thrombosis (DVT).

Case

A 53-year-old woman presented with right lower limb swelling and dilated veins over medial and anterior aspect of the thigh for 3 months. She was diagnosed to have DVT of the right femoral and external iliac veins on ultrasonography. In addition, a hypoechoic lesion was noted in the right inguinal region abutting the wall of the common femoral vein. The provisional diagnosis was lymphoproliferative disease with compression of the inguinal lymph node over

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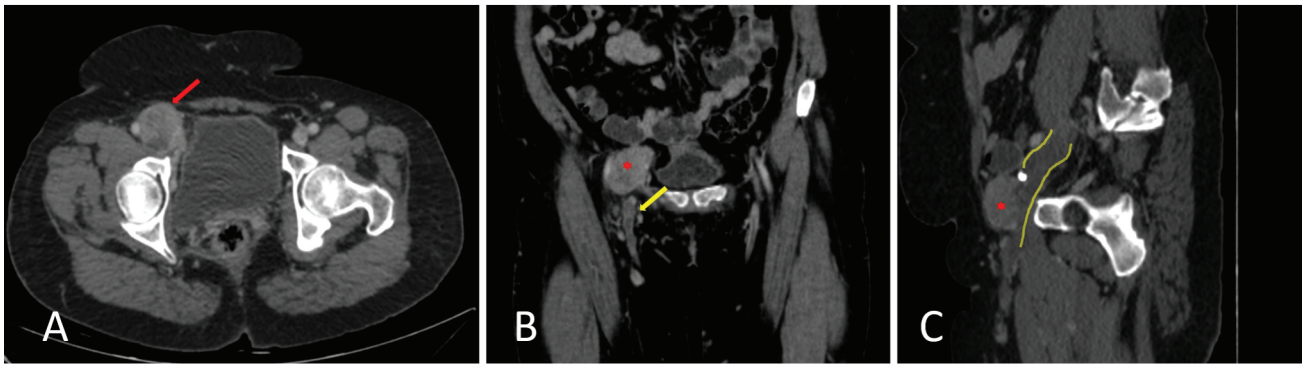

Fig. 1 (**A**) Axial contrast-enhanced computed tomography (CT) image showing enhancing mass (green arrow) likely arising from the anterior wall of the common femoral vein which is also showing a nonenhancing thrombus within the lumen. (**B**) Coronal image showing the mass (red asterisk) and a nodular enhancing filling defect in the great saphenous vein (yellow arrow). (**C**) Sagittal image showing the mass (red asterisk) with a focal calcification with intravenous extension to common femoral vein. The femoral and external iliac vein (yellow outline) filled with nonenhancing bland thrombus.

the femoral vein causing DVT, hence referred to us for inguinal (node) biopsy. Ultrasound showed the inguinal lesion extending to the femoral vein for which cross-sectional magnetic resonance angiography and computed tomography (CT) was done. Magnetic resonance imaging and CT showed enhancing ovoid mass arising from the wall of the common femoral vein with small frond-like intraluminal projections suggestive of a primary lesion in the vessel wall (►**Fig. 1**). Nonenhancing thrombus was noted in the distal femoral and external iliac veins.

She subsequently underwent ultrasound-guided biopsy of the lesion. Microscopic examination showed a neoplasm composed of sheets and fascicles of round to oval cells with dilated vascular channels and mitotic activity (►**Fig. 2**). The cells were diffusely positive for vimentin and CD99 and focally for epithelial membrane antigen and PanCK. They were negative for S100, smooth muscle antigen, desmin, myogenin, and STAT6 with intact nuclear expression for SMARCB1 (INI1) suggestive of synovial sarcoma (►**Fig. 3**).

The patient subsequently underwent wide local excision of the tumor along with segmental venous resection and reconstruction of the femoral vein segment. Patient was advised routine postoperative follow-up.

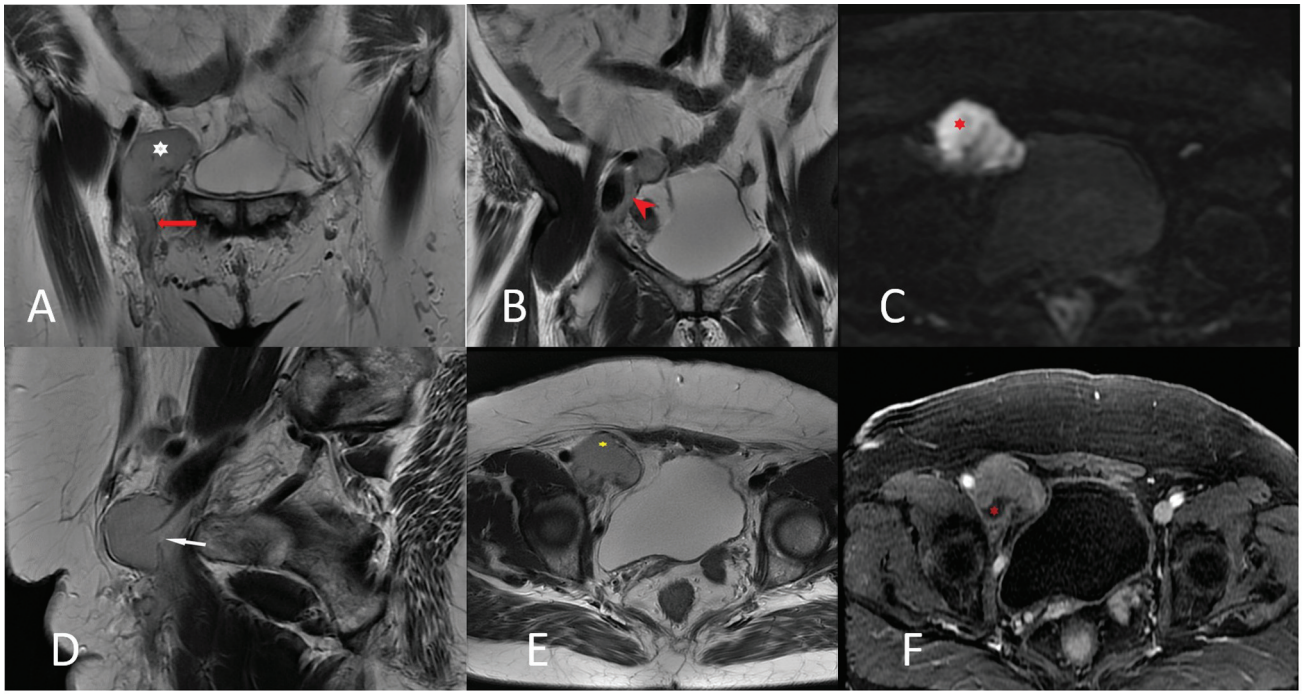

Fig. 2 (**A**) Coronal T2-weighted image showing the hyperintense mass (white asterisk) which is seen extending to the great saphenous vein (red arrow). (**B**) The mass (red arrow) is extending intraluminally into the common femoral vein with thrombosis of the vein below the level of the lesion. (**C**) The lesion (red asterisk) is showing restricted diffusion. (**D**) T2-weighted sagittal image and axial image (**E**) showing the lesion breaching the anterior wall of the vein (white arrow) and extending intraluminally. (**F**) Contrast-enhanced T1 axial section showing the enhancing lesion with intraluminal extension and the presence of nonenhancing thrombus within the vein (red asterisk).

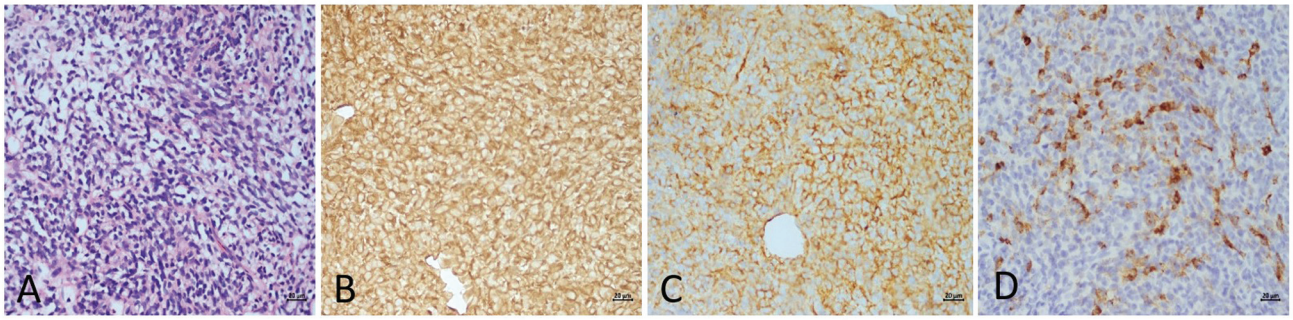

Fig. 3 Monophasic synovial sarcoma (**A**) with cells positive for vimentin (**B**) and CD99 (**C**). Epithelial membrane antigen (EMA) immunoreactivity is present focally (**D**). **A**, hematoxylin and eosin; **B–D**, immunoperoxidase. Magnification = scale bar (**A–D**: 20 µm).

Discussion

Synovial sarcomas are malignant mesenchymal soft tissue tumors comprising approximately 6 to 9% of all adult soft tissue sarcomas. They usually occur around 15 and 35 years and almost usually manifest before the sixth decade of life.[1] The term "synovial" is derived from its morphologic origin and its histological resemblance, and is easily misinterpreted to mean that the tumor comes from the synovium. They occur due to reciprocal translocation between chromosomes X and 18, leading to gene SS18 and SYT-SSX fusion protein.[2,3] Two major histologic subtypes exist: monophasic and biphasic synovial sarcomas. The classical synovial sarcoma has biphasic appearance, consisting of a combination of epithelial and spindle cells in varying proportions. Monophasic synovial sarcomas, on the other hand, are made up entirely of sarcomatous components and are often difficult to diagnose.[4]

Synovial sarcomas have been found in uncommon locations such as the head and neck, mediastinum, lungs, abdominal wall, intra-abdominal region, kidney, and retroperitoneum. There have also been isolated reports of synovial sarcomas occurring in rare locations such as the vulva, skin, blood vessels, and nerves.[5,6] About 60% of cases occur in the lower limb, 20% in the arm, and the remainder in the trunk, head, and neck. Only nine cases of intravascular synovial sarcomas have been reported previously, of which six cases have been reported in the femoral vein. The incidence of primary intravascular synovial sarcoma is extremely rare compared to angiosarcomas and leiomyosarcomas.[7–15]

The differentiation of intravascular tumor and bland thrombus within a vein is not often possible with grayscale ultrasound. The presence of calcifications, vein expansion with intravascular cystic spaces, fluid–fluid levels, and septations within a thrombosed vein should raise the suspicion of an intravascular neoplasm mimicking thrombus. Abundant blood flow signals in color Doppler flow imaging and spectral Doppler imaging and heterogeneous enhancement in contrast-enhanced ultrasound examination also points out to a neoplasm. Cystic changes, calcifications, and fluid–fluid levels are described in intravascular synovial sarcomas. Enhancement of the intravascular mass in the arterial phase in cross-sectional imaging is an accurate indicator of tumor with 87.5% sensitivity and 100% specificity. Bland thrombi are isointense to muscle on T2-weighted images whereas homogeneous T2 hyperintensity will be seen in leiomyosarcomas and some synovial sarcomas. A triple signal pattern (hyperintense, hypointense, and isointense signal areas) on T2-weighted imaging specific to synovial sarcomas and helps in distinction from the more common leiomyosarcomas, however, is seen in larger lesions only.[16]

The primary treatment for synovial sarcoma is surgery. Chemotherapy and radiation therapy can be used as supplementary treatment options or in cases of recurrence. The prognosis for patients is not favorable, with almost 50% of patients experiencing local recurrence or metastases to lungs.[2] New treatments targeting the SYT-SSX junction peptide and retinoic acid derivatives have shown promise; however, the efficacy is still being evaluated.[17,18]

In conclusion, this case highlighted the importance of identification of the various imaging features of intravascular tumors which can masquerade DVT. Awareness of the characteristic imaging findings is important for accurate diagnosis and appropriate management.

Conflict of Interest

None declared.

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