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Case Study

Leiomyosarcoma of the Superficial Femoral vein – A rare tumor at an unusual site.

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Abstract

Background: The leiomyosarcomas most commonly arise from smooth muscles of the uterus and gastrointestinal tract. The vascular origin of the leiomyosarcoma is very rare; however, veins are more frequently affected in such cases.

Case Presentation: A 23-year-old boy presents with left thigh painless swelling. Subsequently, he underwent multimodality radiological examinations, which showed left superficial femoral vein mass, which later on histopathological evaluation came out to be grade III leiomyosarcoma of the superficial femoral vein.

Management & Results: The patient was given chemotherapy, but unfortunately, after three cycles of chemotherapy patient developed pulmonary metastases. Currently, the patient is under palliative treatment.

Conclusion: Primary vascular leiomyosarcomas are very rare entities. The knowledge of these rare tumors is essential for the reporting radiologists and clinicians to properly diagnose these aggressive tumors to prevent any delays in the management.

Keywords

Leiomyosarcoma, Vascular Leiomyosarcoma, Vascular Tumor, Lung Metastases, Metastatic Vascular Leiomyosarcoma.

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Introduction

The leiomyosarcomas (LMS) are rare mesenchymal tumors of smooth muscles origin, which account for about 8% of the soft tissue tumors and are considered the malignant leiomyoma counterpart[1]. Vascular leiomyosarcoma is an extremely rare subtype of LMS which arises from the smooth muscles of the blood vessels and usually involve the major veins like the inferior vena cava[2]. Unfortunately, the prognosis of vascular leiomyosarcomas is poor and requires a multimodality radiological approach for the diagnosis and a multidisciplinary therapeutic approach, including surgery, chemotherapy, and radiotherapy.[3]

Case Presentation

A 23 years old young boy presented with swelling in the left thigh for four months. On examination, the firm to hard painless mass was noted in the femoral triangle, which was extending underneath/above the inguinal ligament. Femoral pulses were not appreciated below mass. The patient was then referred to the ultrasound department for further evaluation of the mass, which revealed a heterogeneous oblong-shaped mass that was inseparable from the femoral artery causing its lateral displacement. The femoral vein was not visualized. Subsequently, contrast-enhanced MRI was advised to further characterize the mass, which showed heterogeneously enhancing soft tissue mass circumferentially involving the superficial femoral vein with intraluminal extension (Fig 1. A-F). The case was discussed at the oncology conference. Upon the board's recommendations, an excisional biopsy of the mass was done, which revealed grade III leiomyosarcoma of the superficial femoral vein (Fig 2. A-D), followed by radiotherapy.

Management & Results

The patient was then kept on chemotherapy, but unfortunately, after three cycles of chemotherapy patient developed pulmonary metastases. Currently, the patient is under palliative treatment.

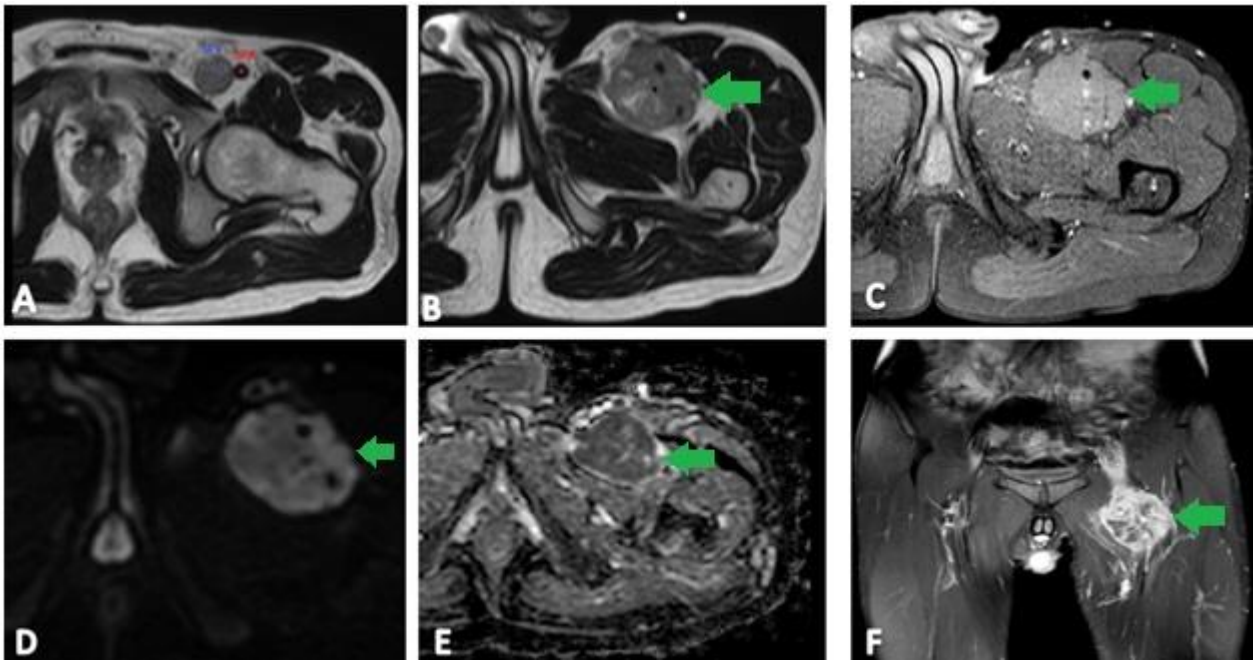

Figure 1: (A-F). Axial T1 (A), axial T2 (B), axial T1 post-contrast (C), DWI (D), ADC (E), Coronal T1 post-contrast fat-suppressed (F) showing heterogeneously enhancing soft tissue mass circumferentially involving the superficial femoral vein with intraluminal extension (green arrow).

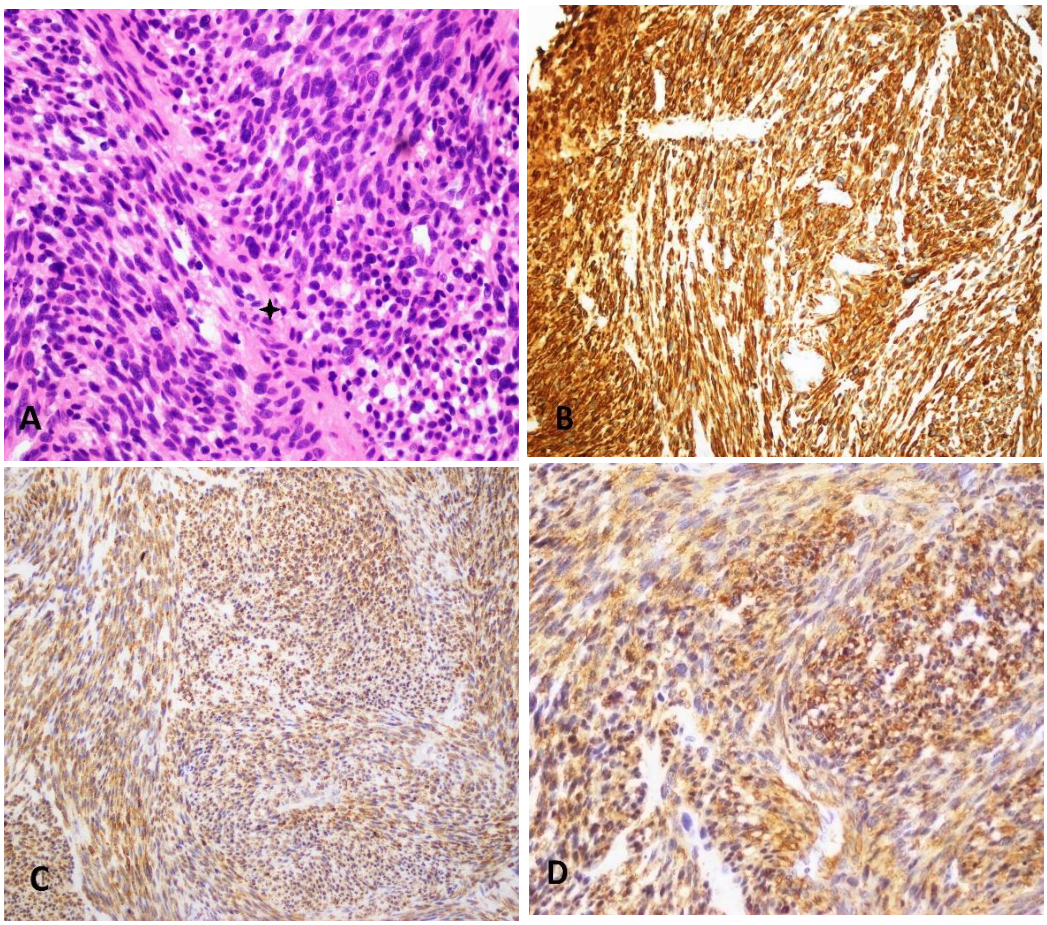

Figure 2: (A-D) High-resolution histopathological view showing spindle tumor cells surrounding vessel (asterisk) (A), tumor positive for Desmin stain (B), negative MyoD1 stain (C), and positive SMA stain (D).

Discussion

Langenbeck first reported primary vascular leiomyosarcoma (VLMS) in 1861[4]. It usually originates from the major veins like the inferior vena cava (60%) and very rarely from the lower extremity deep vessels. These tumors usually have a high-grade malignant potential[4] and exhibit 3 main patterns of growth, i.e., extraluminal, intraluminal, and combined with the extraluminal pattern being most common (62%)[3, 5]. Clinically, these tumors exhibit slow growth, which explains their late presentation and scarcity of clinical symptoms. These may remain asymptomatic or can present with palpable, painless mass, tingling, or pain due to compression of surrounding nerves. In advanced cases, distant metastasis, especially in the lungs and livers, has been reported[4].

Radiologically, the imaging features are variable, non-specific, and require a multimodality approach; however, CT scan and MRI remain the modality of choice for demarcation of the site of origin, extent, staging, and relation with surrounding structures. MRI is best to assess the relation of the tumor with the vessel wall, invasion into surrounding soft tissues, and extension into the joint cavity. Depending upon size and internal liquefactive contents, larger tumors are more heterogeneous with iso to hypointense signals to muscles on T1, hyperintense on T2 and STIR, and heterogeneous post-contrast enhancement. Surgical resection is the standard treatment, along with vascular resection and grafting followed by adjuvant radiotherapy and chemotherapy[6].

Conclusion

Primary VLMS are aggressive and rare tumors with a high potential for metastasis and recurrence. A multidisciplinary approach should be considered for management.

Conflicts of Interest

The authors have declared that no competing interests exist.

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