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CASE REPORT

Rare Case Presentation of Leiomyosarcoma as IVC Thrombus

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Background

Leiomyosarcoma (LMS) of vascular origin is uncommon. This leiomyosarcoma is seen occurring from the tunica media of major blood vessels. Leiomyosarcoma of venous origin is five times more common those of arterial origin. Most cases are presented in woman with a median age of 50 years [1]. We are presenting a case where histopathology was instrumental in making the diagnosis of LMS in the thrombus and confirmed by IHC.

A 46-year old female presented with bilateral swelling of lower limbs and distention of abdomen. Her CT, MRI, and color Doppler were suggestive of IVC, hepatic, and renal vein thrombosis. Histopathology of thrombus revealed leiomyosarcoma and was confirmed by IHC. Conservative treatment was given and asked for a regular follow-up. This case report gives us an opportunity to think of a differential diagnosis in a patient presenting with IVC thrombus

Introduction

LMS accounts for 5–10% of soft tissue sarcomas. Leiomyosarcoma occurring within major blood vessel is very rare comprising only a few hundred cases in the literature [2]. More than half of all vascular LMS occurring from the inferior venecava accounting or only 0.5% of all soft tissue sarcomas in adults [3].

Case Report

A 46-year-old female presented to hospital with 2-week history of swelling of lower limbs and distention of abdomen. CT abdomen revealed IVC, hepatic veins, renal veins, and left ovarian vein thrombosis. MRI, color Doppler, and CT venogram were also suggestive of IVC right atrium junction. There was also extension into right atrium (confirmed on ECHO). There was mild dilatation of IVC and some contrast uptake in the thrombus. PET CT did not reveal any uptake in the thrombus or rest of the body. Her procoagulant and PNH work up was negative. Angiogram-guided thrombolysis was done. Thrombus was sent for histopathological analysis.

Pathological Findings

Gross examination: Received multiple brownish tissue bits measuring 2 × 2 × 2 cm. Microscopy revealed blood clot, thrombus, and fragments of either cellular or edematous tumor tissue. The tumor cells are spindle-shaped with elongated fusiform nuclei arranged as fascicule (Images 1, 2, and 3). The cytoplasm is scanty, eosinophilic, and fibrillar. Moderate nuclear pleomorphism and occasional mitosis are seen. Few binucleate and tumor giant cells are seen (Image 2).

A diagnosis of leiomyoma with atypia was given and IHC was advised. IHC revealed spindle cells staining positive for smooth muscle actin and desmin and negative for CK. CD 34 is seen positive in the lining of the tumor vasculature. Ki67 is 10–20%. A final diagnosis of low-grade leiomyosarcoma was made based on all above findings.

Multidisciplinary meeting was done. Option of IVC excision with construction of new IVC was considered. Patient was unwilling to proceed for surgery. Conservative treatment was started. Patient is on follow-up and doing well.

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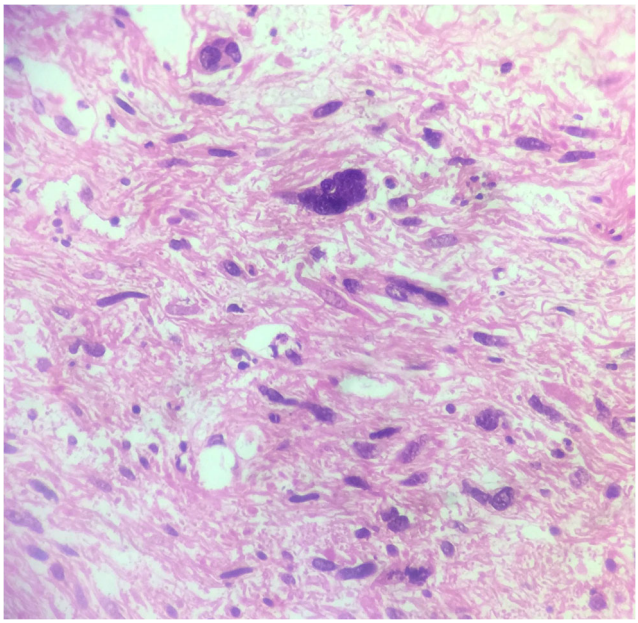

Image 1 Pathological findings:Spindle cells with marked pleomorphism, tumor giant cells, increased N:C ratio, vescicular chromatin, prominant nucleoli

Discussion

Vascular LMS is a rare tumor occurring within the media of smooth muscle. Perl L [4] reported first case in German literature in 1871. It is seen in the 6th decade with female preponderance. The clinical manifestation of quickly growing tumor may cause paresthesia, edema depending on compression, and occlusion of vital structures. Metastatic disease is reported in fewer than 505 of cases. Dzsinich et al. [5] reported 13 cases of LMS (A retrospective analysis was done in 13 patients who had undergone surgical resection of primary tumor occurring at these sites ie., 8 cases in iliac, 2 in IVC, 2 in saphanous vein).

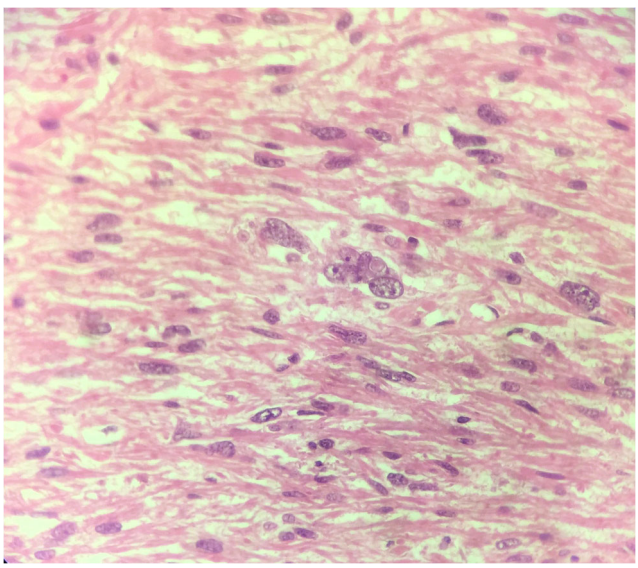

Image 2 Pathological findings: moderate nuclear pleomorphism and occasional mitosis

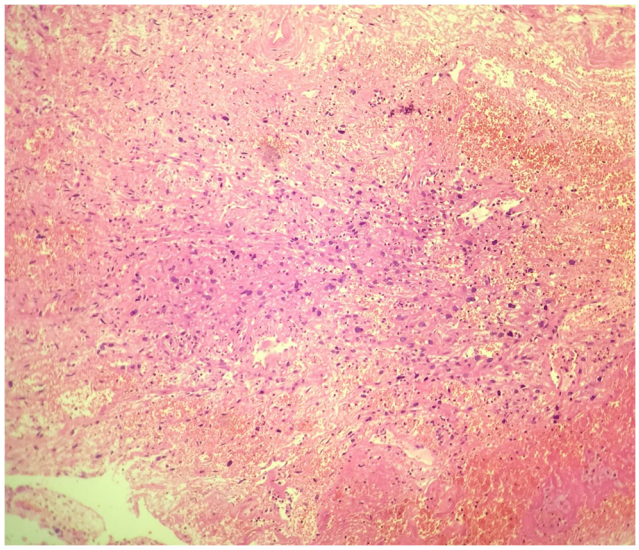

Image 3 Pathological findings: spindle-shaped cells with elongated fusiform nuclei arranged as fascicule

Conclusion

Leimoyosarcoma forms a very rare diagnosis in a thrombus. It forms a differential diagnosis in the absence of adequate response to anticoagulant therapy. biopsy through fluoroscopfy guided is suitable in such instances for confirmation, followed by chemotherapy, radiation therapy and close follow up for the betterment of the patient.

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