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Case Report

Chronic graft thrombosis after en bloc resection of inferior vena cava leiomyosarcoma. A case report[☆]

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ABSTRACT

Leiomyosarcoma (LMS) is a rare malignant tumor originating from smooth muscle cells. Primary leiomyosarcomas arising from vessels' walls are extremely rare (2%), with LMS of inferior vena cava being the most frequent subtype. We present the case of a 45-year-old man with a past medical history of resected leiomyosarcoma of the right calf, presenting with a follow-up CT showing a retroperitoneal mass arising from the inferior vena cava, which proved to be IVC leiomyosarcoma at histopathology. The patient underwent surgical resection of the mass with prosthetic reconstruction of the IVC. Three days after surgery he developed complete thrombosis of IVC graft which persisted at 3-months follow-up imaging and was treated with pharmacological therapy. Although there are many references reporting the association between LMS of the inferior vena cava and postoperative deep vein thrombosis, to our knowledge there are no reports currently available regarding complete thrombosis of IVC vascular graft after surgical resection of IVC LMS.

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Introduction

Leiomyosarcoma (LMS) is a rare malignant tumor originating from smooth muscle cells. Primary leiomyosarcomas originat-

ing from the smooth muscle of vessels' walls are extremely rare (2%) [1,2]. The most frequent location of vascular LMS is inferior vena cava (IVC); this location has a poorer prognosis when compared to leiomyosarcomas of other anatomic sites with the same histopathological features [1].

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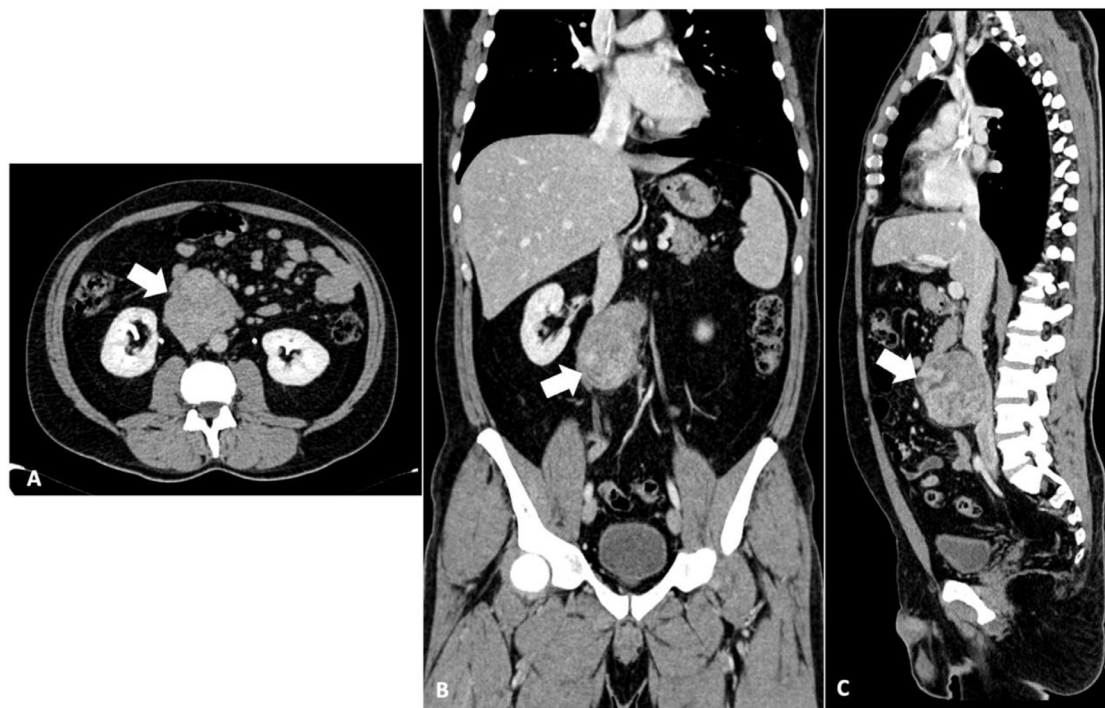


Fig. 1 – Axial (A), coronal (B), and sagittal (C) preoperative contrast-enhanced CT demonstrating a partially necrotic soft-tissue retroperitoneal mass (white arrows) compressing the infrarenal inferior vena cava.

We present the case of a 45-year-old man with IVC leiomyosarcoma who developed a persistent postoperative thrombosis of the prosthetic vascular graft after surgery.

Case report

A 45-year-old man with a previous leiomyosarcoma of the right calf (resected in 2020) presented to our oncological department in December 2021 with a follow-up contrast-enhanced CT showing a partially necrotic soft-tissue retroperitoneal mass apparently arising from the inferior vena cava with maximum diameters of $7.3 \times 5.6 \times 7.3$ cm (Fig. 1).

The neoplasm had a mass effect on the IVC, the duodenum, the head, and the uncinate process of the pancreas, the abdominal aorta, and the right iliopsoas muscle.

Multiple paracaval and right retrocrural adenopathies (maximum short axis of 8 mm) were also detected. The patient underwent a biopsy of the paracaval mass; histopathology showed a G2 leiomyosarcoma arising from the IVC (Fig. 2).

Contrast-enhanced CT performed 2 months after neoadjuvant chemotherapy with Doxorubicin-Dacarbazine didn't show any significant change in the lesion's size and contrast enhancement. No distant metastases were detected.

The case was reviewed by our multidisciplinary sarcoma board, which opted for surgical treatment.

Surgical resection of the IVC mass, prosthetic reconstruction of IVC, and abdominal nodulectomy were performed.

Three days after surgery, the patient developed dyspnea and lower limb swelling and pain. A chest-abdomen and lower

limbs contrast-enhanced CT was performed to rule out pulmonary embolism and deep venous thrombosis. CT showed complete graft thrombosis and bilateral thrombosis of iliac and common femoral veins (Fig. 3).

No pulmonary embolism was detected.

The patient underwent pharmacological therapy with low-molecular-weight heparin at a therapeutic dosage (100 IU/kg every 12 hours).

A 3-day and 3-month follow-up venous CT-angiography showed persistence of thrombosis of the IVC graft and iliac-femoral venous axis (Fig. 4).

The patient continued anticoagulation therapy with novel oral anticoagulants (NOAC) and was in good clinical conditions at follow-up physical examination, with improvement of lower limb swelling.

Discussion

Inferior vena cava leiomyosarcoma (IVCLMS) has an insidious onset and most patients may present with nonspecific symptoms such as poorly localized pain, nausea, and vomiting. In some cases, metastatic disease may be the first manifestation of the tumor [1].

Computed tomography is the most useful imaging modality in the management of IVCLMS, providing excellent anatomic details and being a useful guide for biopsy and treatment choice [4].

Complete resection of the tumor, when feasible, is the first-choice treatment [1-3]. Surgery might be combined with

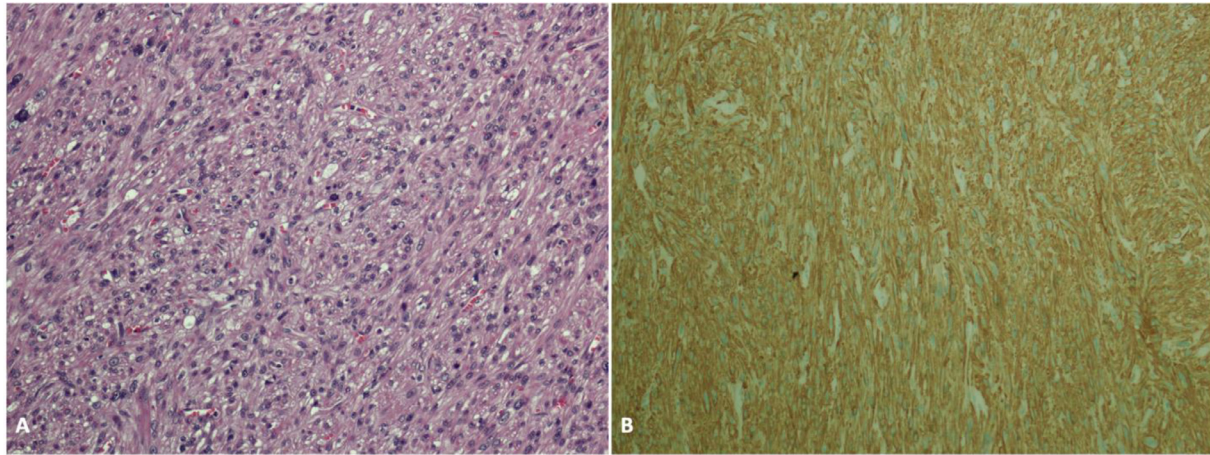


Fig. 2 – (A) Histologically, the tumor consists of spindle cells with eosinophilic cytoplasm arranged in sheets and whorled, intersecting fascicles, with vascular necrosis (20% of the neoplasia area); there was pronounced nuclear pleomorphism and a mitotic rate of 9 mitoses on 1734 mmq, also with atypical mitosis (4x magnification); **(B)** Tumor cells with positive reaction to H-Caldesmon (10x magnification).



Fig. 3 – Contrast-enhanced CT scan performed 3 days after surgery, showing complete thrombosis of the venous graft (white arrows in A and B) and bilateral complete thrombosis of the iliac veins (blue arrowheads in B).

neoadjuvant chemotherapy and radiotherapy to improve the outcome, quality of life, and overall survival of the patient.

The technical challenges presented by the anatomical characteristics of the disease may raise issues regarding multivisceral resection and vascular reconstruction techniques [5–8].

Vascular reconstruction with synthetic tube grafts is recommended whenever the patency of the IVC is confirmed at preoperative imaging. Five-year and 10-year progression-free survival rates after successful surgical intervention may reach 30% and 7%, respectively [9].

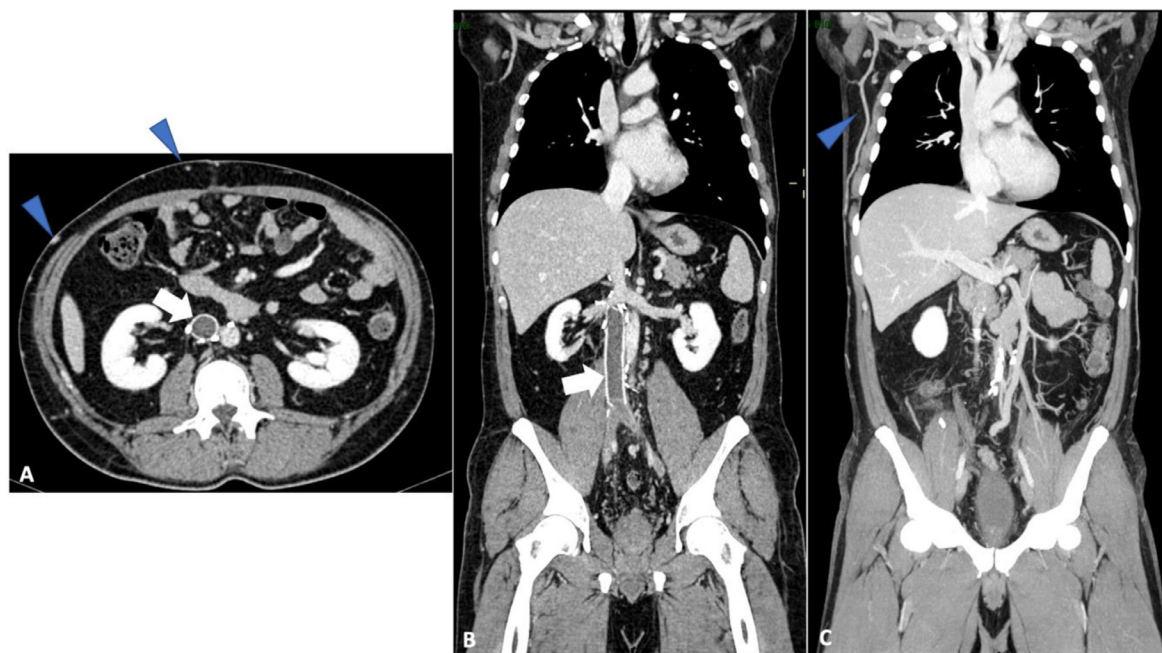


Fig. 4 – Chest-abdomen CT-venography performed 3 months after surgery, showing persistent thrombosis of the IVC venous graft (white arrows in A and B), with multiple compensatory dilated subcutaneous collateral veins (blue arrowheads in A and C).

The most frequent postsurgical complications include surgical site infections, sepsis, and venous thromboembolism [10].

Since IVC reconstruction can be performed safely with low venous thromboembolism-associated morbidity, routine anticoagulation might not be recommended in these patients, but early postoperative screening for thrombosis should be considered, especially in cases with large tumor burden or when graft reconstruction is performed [10].

IVC thrombosis is often an underdiagnosed condition because most commonly pulmonary emboli are thought to arise from a lower extremity deep venous thrombosis.

Patients with IVC thrombosis are at particularly high risk, with reports of catastrophic pulmonary emboli occurring in up to 8% of patients [10].

Conclusion

Persistent complete graft thrombosis is a must-know condition in patients undergoing surgical resection of IVC leiomyosarcoma and prosthetic reconstruction of IVC.

Graft thrombosis can lead to serious complications; being aware of this condition is essential for an early-stage diagnosis and treatment, in order to improve patients' outcome.

Patient consent

Informed consent was obtained from all individual participants included in the study.

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