

## Leiomyosarcoma of the inferior vena cava

**Ben Abid Sadri,**  
**Attaoui Mohamed Amine,**  
**Mzoughi Zeineb, Miloudi Nizar,**  
**Gharbi Lassad,**  
**Mohamed Tahar Khalfallah**  
**General Surgery Department, Mongi Slim hospital, La Marsa, Tunisia**

### Abstract

Vascular leiomyosarcoma (LMS) are unique. The inferior vena cava (IVC) is the most affected organ (about 38% cases). We report the observation of a 50-year old woman who consulted for right upper quadrant pain. Imaging studies revealed a retroperitoneal mass that mimic a LMS of the IVC. The patient was operated. A resection of the IVC along with the tumor was performed without reconstruction. The management of LMS is surgical and depends upon the location and tumor characteristics.

### Introduction

Leiomyosarcoma (LMS) of the inferior vena cava (IVC) is a slowly developing tumor which arise from smooth muscle cells of the wall of IVC in 38% cases.<sup>1</sup> This tumor reside at the level of segment I represented by sub-renal IVC in 38% cases.<sup>2</sup>

### Case Report

A 50-year old patient, with no previous medical history, consulted for upper quadrant pain. Physical examination revealed a lower extremities edema. Abdominal palpation showed a non-pulsating mass on the right flank region. There was also hepatosplenomegaly. Laboratory workup was normal. Ultrasound studies and computed tomography (CT) scan (Figures 1, 2) revealed a multilobulated right mass, centered by the IVC invading along 7 cm. The right renal hilum was not invaded. There was no hepatic metastasis. Magnetic resonance imaging (MRI) showed an isointense T1 mass and hyperintense T2 emerging from the IVC adjacent to the right kidney (Figure 3). CT scan guided biopsy found a fusiform cells tumor. The patient was operated. During surgery, there was a sub-renal IVC tumor, totally obstructive, located 1 cm from the right renal

vein ostium and 5 cm from iliac vein (Figure 4). A resection of the IVC along with the tumor (Figure 5) was performed after clamping the IVC above the right renal vein. An IVC reconstruction wasn't done (Figure 6). Pathology report of the surgical specimen confirmed the diagnosis of IVC LMS. The post-operative period was complicated by the appearance of lower extremities deep vein thromboses at Day 20 which well evolved with anticoagulant therapy. Currently, the patient has well recovered. There is no relapse or secondary localization with a 1-year follow up.

### Discussion and Conclusions

LMS of the IVC is characterized by its great clinical latency. The diagnosis is established pre-operatively in only 10% cases.<sup>3</sup> In tumors affecting segment I and II, there is usually a colicky pain which can be either hepatic or renal. Lower extremities edema is a rare manifestation which is due to the slow tumor growth that gives time for venous collateral circulation to develop. In our observation, the patient presented with a painful mass on the right flank with slight edema. Ultrasound examination is an important step in diagnosing LMS of the IVC. It allows the detection of retroperitoneal hypoechoic heterogeneous tumor. CT-scan and abdominal MRI allowed to precisely locating the tumor relatively to the IVC, its vasculature and within neighboring organs. The cavography have low interest as additional diagnostic tool for caval tumors. It doesn't provide the ability to differentiate between a caval tumor and thrombosis. A biopsy, either CT guided or transvenous is to be discussed. It can be indicated when there is doubt in establishing the diagnosis or neoadjuvant chemotherapy is planned. In our case, preoperative biopsy allowed establishing the diagnosis of sarcoma.

A complete R0 surgical resection is the unique cure of IVC LMS. Segment I is the localization best suited for resection. If the tumor is not obstructing and limited to one venous wall, a lateral resection with suture can be done. If the tumor is large, totally obstructive, as in our case, wide excision with interruption of the IVC is performed. Reconstruction is rarely performed as it is not needed in most cases; because a well developed alternative circulation usually exist in the totally obstructive tumors.<sup>4</sup> In suprarenal IVC, a prosthetic reconstruction might be necessary. The indications for prosthetics are incomplete obstruction of the vena cava or total resection along with many collaterals. However, prosthetic replacement remain a solution but controversial because of the risk of thrombotic events and infections. Invasion of the renal veins espe-

Correspondence: Attaoui Mohamed Amine, General surgery department CHU Mongi Slim, La Marsa, Tunisia.

E-mail: attaouiamine@gmail.com

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**Figure 1.** Abdominal computed tomography scan showing a retroperitoneal tumor that emerges from subrenal inferior vena cava.



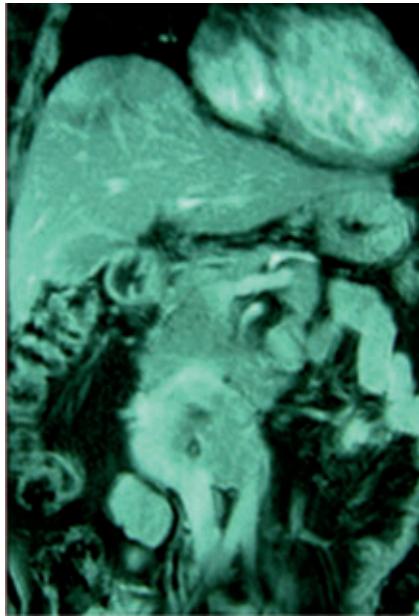
**Figure 2.** Abdominal computed tomography scan showing non-invaded renal veins.

cially right renal vein is a problem that can be encountered during surgery in tumors of segment I and II. Left renal vein can be ligated. Left renal vein drainage is done through lumbar gonadal veins or suprarenal veins. If the right renal vein is invaded, a reimplantation of the renal vein over the prosthetics, on the por-

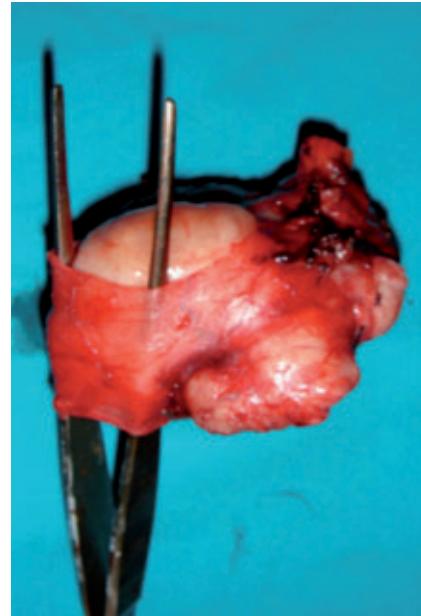
tal vein or renal autotransplantation in the iliac fossa is necessary to avoid right nephrectomy.<sup>5</sup> Adjuvant treatment of sarcoma is controversial. LMS are more sensitive than other retroperitoneal in the association of gemcitabine-docetaxel. Our patient didn't receive any adjuvant therapy.

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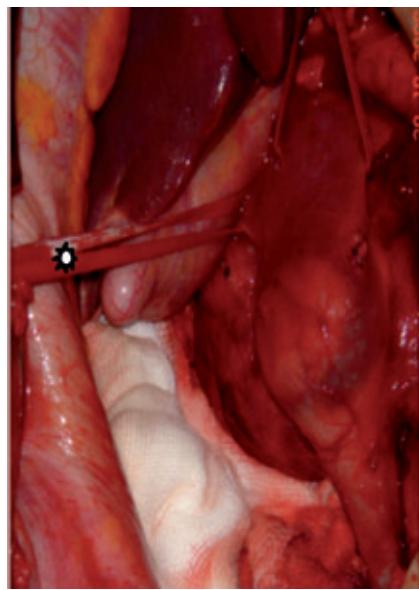
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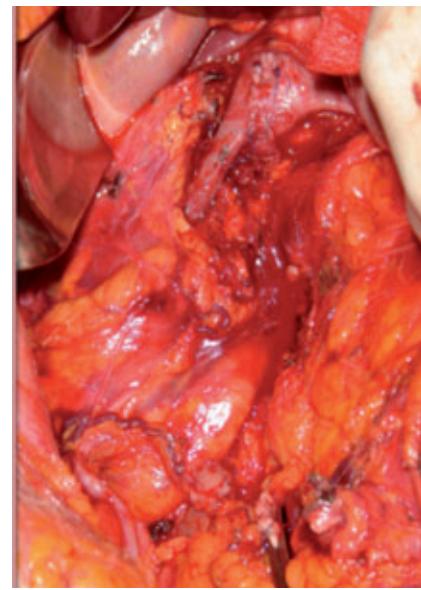
**Figure 3.** Abdominal magnetic resonance imaging reveals an isointense T1 tumor that emerges from inferior vena cava adjacent to right kidney.



**Figure 5.** Gross-view of the tumor emerging from the venous wall.



**Figure 4.** Gross-view of the totally obstructive tumor located 5 cm from right renal artery ostium.



**Figure 6.** Gross-view of the inferior vena cava after tumor resection.