ECRONICON OPEN ACCESS

EC CARDIOLOGY
Case Report

Leiomyosarcoma of the Inferior Vena Cava Diagnosis and Treatment: Case Report

O Belmaachi*, K Farahi, S Lyazidi, A Adaoui, O El Allam and Y Ettaoumi

Cardiovascular Surgery Department, University Hospital Ibn Rochd, Casablanca, Morocco

***Corresponding Author:** O Belmaachi, Cardiovascular Surgery Department, University Hospital Ibn Rochd, Casablanca, Morocco.

Received: August 08, 2022; **Published:** July 27, 2022

Abstract

Malignant tumors of the inferior vena cava are very rare, leiomyosarcoma accounts for 95%. Leiomyosarcoma of the inferior vena cava is a rare malignant tumor developed at the expense of smooth muscle cells of the vascular wall. Preoperative diagnosis is only made in 10% of cases. CT and MRI data allow an accurate assessment of tumor extension. The only curative treatment for leiomyosarcoma of the inferior vena cava (IVC) is complete surgical excision associated with radio-chemotherapy. Recurrences are frequent. We report a surgical approach to a grade II located in the suprarenal and retrohepatic segment, between the renal veins and the suprahepatic veins of the IVC. The evolution was favorable. After 2 years of follow-up, the patient was free of any tumor recurrence.

Keywords: *Leiomyosarcoma; Inferior Vena Cava; Surgery*

Introduction

Leiomyosarcoma of vascular origin is the most common vascular malignant tumor, mainly located at the level of the inferior vena cava (VCI), it is a malignant tumor developed arising from the smooth muscle cells of the vascular wall. Leiomyosarcomas are rare, representing 0.5% of all soft tissue tumors [1,2]. It is a long asymptomatic pathology, which explains the significant diagnostic delays at the extravascular tumor stage [1]. Diagnosis is based on morphological examinations and anatomopathological evidence. Cross-sectional imaging is an important element in the diagnosis and assessment of tumor extension. Since chemotherapy and radiotherapy have not proven their effectiveness, surgical excision with healthy margins represents the only curative treatment [1,2]. IVC leiomyosarcomas are classified according to their anatomical location into three areas [1,2]:

- Zone I: Infrarenal segment of the IVC.
- Zone II: Suprarenal and retrohepatic segment of the IVC, between the renal veins and the suprahepatic veins.
- Zone III: Suprahepatic segment, between the suprahepatic veins and the right atrium.

Observation

This is a 47-year-old patient, with a history of cholecystectomy, who presented asthenia and isolated paroxysmal epigastric pain not improved by symptomatic treatment for 3 years. The clinical examination found a hard mass of the epigastrium without beating or

tenderness, mobile in relation to the superficial tissue without hepatomegaly, splenomegaly or associated lymphadenopathy. Abdominal ultrasound revealed a heterogeneous, polycyclic hypoechoic process of epigastric seat. For better characterization of the mass, a thoraco-abdomino-pelvic CT scan, without and with injection of contrast product, was performed. He specified the retroperitoneal seat lateralized to the right of the mass, at the expense of the retrohepatic inferior vena cava (VCI), which is hypodense lobulated tissue in nature, heterogeneously enhanced, measuring 60 x 50 x 100 mm. It encompasses the right adrenal gland and comes into contact with the head of the pancreas, without signs of obvious invasion or associated secondary localization (Figure 1). The patient underwent excision of the mass. The anatomopathological study was in favor of a leiomyosarcoma of the IVC and the follow-up was favorable after 2 years of monitoring.

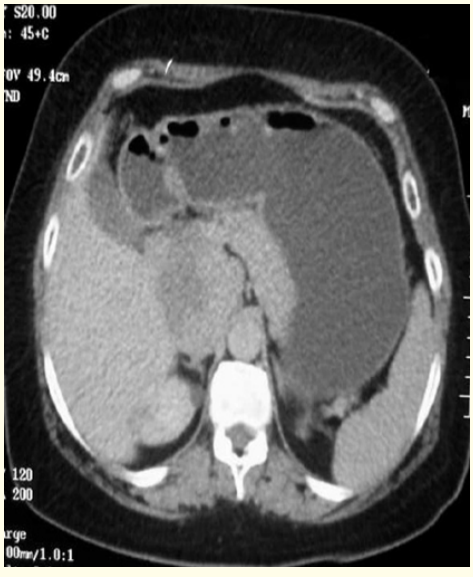

Figure 1: *Abdominopelvic CT in axial sections before injection of contrast product showing a right lateralized, lobulated, hypodense retroperitoneal tissue mass measuring 60 x 50 x 100 mm.*

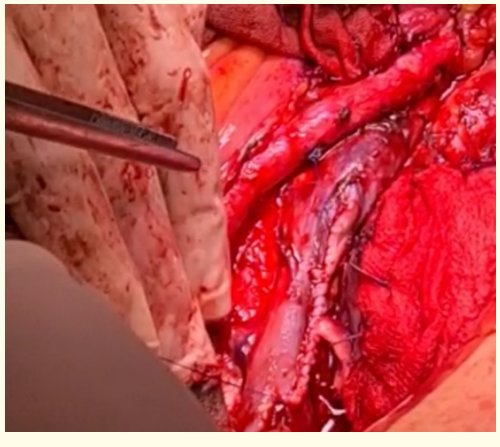

Figure 2: *Image representing repair of the IVC after resection of a leiomyosarcoma.*

Discussion

IVC tumors are rare. They would represent 0.07‰ cases of an autopsy series [1]. Of these tumors, 95% are leiomyosarcomas [2,11]. The remaining 5% are represented by: fibrosarcoma, endothelioma, cystic (lymphangioma) or solid (embryonic and lymph node remnants) formations [7]. Vascular localizations of leiomyosarcoma are also rare. Involvement of the IVC is seen in 50% of cases [7]. Leiomyosarcoma of the IVC is defined as a malignant mesenchymal tumor developed at the expense of the smooth muscle fibers of the venous wall. Most of the cases reported in the literature (80%) are women [7,11]. The average age at diagnosis is 54.3 years [10]. IVC leiomyosarcoma is diagnosed intraoperatively in 63% of cases and post-mortem in 27% of cases [11]. The clinical diagnosis is only mentioned in 10% of cases [11]. This is due to the fact that the symptomatology is often not very suggestive, preceding the discovery of the tumor by several years. In this situation, the different clinical manifestations depend on the characteristics of the tumor. The tumor can develop extra-vascular, intra-vascular. The clinical symptomatology of the tumor depends on the site of its development on the IVC. Atrio-hepatic segment involvement may manifest as cardiac disorders, pulmonary embolism [2,11] or acute or chronic Budd-Chiari syndrome [12]. When the tumor originates from the supra-renal (58% of cases [2]) or sub-renal segment, the symptomatology is dominated by pain that may evoke a hepato-biliary, renal or colic origin [2,3,11]. Edema of the lower limbs is rare due to the slow progression of the tumor which allows time for collateral venous circulation to develop [2,3,7,12]. The tumor is palpable in less than 50% of cases [3]. Certain general signs, such as a deterioration in general condition or a fever, may be present and prompt the search for neoplasia by carrying out additional explorations [2,7]. Ultrasound is an important step in the preoperative diagnosis. It shows a mass of heterogeneous hypoechoic retroperitoneum closely related to the IVC. Visualization of dense echoes inside it should raise the suspicion of thrombosis [7]. CT and MRI [10] make it possible to highlight the tumoral process, to study its extension in relation to the IVC and in relation to neighboring organs, to assess the impact of the tumor on blood flow and to look for secondary locations [12]. Confirmation of the diagnosis is provided by histological or cytological analysis of the biopsy fragments. They can be obtained either by ultrasound-guided or CT-guided biopsy [12], or sometimes by transjugular biopsy when the tumor protrudes into the lumen of the vein [7,12]. The interest of this examination lies in the possibility of performing chemotherapy associated or not with preoperative radiotherapy [6,8,12]. The evolution of IVC leiomyosarcoma is slow and remains local. A cleavage plane is usually present with the neighboring organs. The latter are very rarely invaded or only at very advanced stages of tumor progression [3]. Metastases, when present (30 to 50% of cases), involve the liver, lung, lymph nodes and bone [2,6,7,10]. The prognosis remains severe due to a late and unforeseen diagnosis (63% of these tumors are discovered intraoperatively) making the surgical procedure difficult [2]. Surgery is the radical treatment for leiomyosarcoma of the IVC [12]. The goal of treatment is complete excision of the tumor, preservation of venous return and prevention of recurrence [10]. The need for a technical platform for vascular surgery must be foreseen in the face of any retroperitoneal tumor [11]. The tumor must be excised completely without worrying about the IVC, which will be resected in most cases, respecting a safety margin [5, 10] for some it would be 1 cm [9]. The macroscopic appearance of the specimen is that of a greyish-red polylobed tumour. On section, the tumor closely resembles uterine fibroids, forming fasciculate vortices [10]. If the venous resection is partial, direct suture or angioplasty will be performed [11]. The use of a vascular prosthesis to ensure the continuity of venous flow does not seem to be unanimous and remains subject to many controversies [3,11,12]. Due to Infectious and thromboembolic risks. The placement of a prosthesis will be discussed on a case-by-case basis depending on whether or not there is collateral circulation [2,12]. Vascular replacement would be recommended in the event of extensive resection of the IVC with a single right kidney (absence of collateral circulation) or during excision of the retrohepatic segment above the suprahepatic veins [7]. The use of polytetrafluoroethylene (PTFE) prostheses is the material of choice which exposes to little morbidity [3,4,10]. The existence of an invasion of neighboring organs should not stop the excision. Extensive resections of the aorta and liver [4,11] are possible. Similarly, the presence of metastases should not contraindicate tumor excision. Complete resection of the latter, even as a palliative measure, makes metastases susceptible to adjuvant therapy [7,9,10]. No study has proven the effectiveness of either chemotherapy or radiotherapy [3,7,10,11]. Some sporadic cases of their effectiveness are reported [10]. The rarity of the disease and the small number of patients who can form homoge-

neous groups make it difficult to determine the therapeutic protocol [6,10]. The rate of complete resectability of the tumor is around 60% of cases (56.9 - 61.5% of cases [15,18]). After analysis of the results of complete excision of the tumour, from the international register of LMS of the VCI, operative mortality is 2.5% of cases, venous thrombosis of the lower limbs is 17.5% of cases and the risk of recurrence is 57.3% of cases after a follow-up of 32 +/- 4 months [12]. The risk of death after resection increases significantly when there is lower limb edema, Budd-Chiari syndrome, occlusion of the IVC, intravascular tumor proliferation and development of the latter from the segment suprashepatitis of the IVC [12]. Survival at 5 years and 10 years is 49.4 and 29.5% respectively in selected patients: without preoperative metastasis and without microscopic residues after resection [11]. Tumors originating from the middle segment of the IVC have a significantly better prognosis than those originating from the lower segment [10]. Invasion of the excision limits is a poor prognosis. In another work [11], 5-year survival is 0% if the resection margins are invaded, compared with 68% of cases if the margins are healthy. None of the following factors appeared as an element of prognosis: Age, sex, tumor size and lymph node status [5,6,10,12].

Conclusion

Leiomyosarcoma of the inferior vena cava is a rare tumour, often diagnosed late and therefore with a poor prognosis. The use of medical imaging, CT and especially MRI, makes it possible to have a precise lesion assessment and to predict surgical difficulties. Complete excision of the tumor passing through healthy resection margins on the IVC is the reference treatment. The recidivism rate remains high. Aggressive surgery associated with radiochemotherapy would be a better treatment for LMS of the IVC and could guarantee better survival.

Bibliography

1. Soualy K., *et al*. "Leiomyosarcoma of the inferior vena cava: a new case report". *Urology Annals* 37.4 (2003): 177-179.
2. Malajati H., *et al*. "Leiomyosarcoma of the inferior vena cava". *Radiology Patient Leaflets* 49.2 (2009): 105-109.
3. Leong Tan GW and Hoong Chia K. "An unusual case of leiomyosarcoma of the inferior vena cava in a patient with double inferior vena cava". *Annals of Vascular Surgery* 23.2 (2009): 13-19.
4. El Malki HO., *et al*. "Leiomyosarcoma of the inferior vena cava: diagnostic problem and treatment". *Progrès en Urologie* 13 (2003): 293-296.
5. Bonnet S., *et al*. "Leiomyosarcoma of the middle segment of the inferior vena cava: tactical issues raised by vena cava and renal revascularization". *Journal des Maladies Vasculaires* 31 (2006): 79-84.
6. Soury P., *et al*. "Leiomyoscarcomas of the inferior vena cava". *EMC-Cardiology Angiology* 2.1 (2005): 90-96.
7. Ben Hassouna J., *et al*. "Leiomyosarcoma of the subrenal inferior vena cava". *Journal de Chirurgie* 143.5 (2006): 325-327.
8. N Hammoune., *et al*. "Leiomyosarcoma of the inferior vena cava: a case report". *The Pan African Medical Journal* 20 (2015): 283.
9. Guarino G., *et al*. "Leiomyosarcoma of the inferior vena cava: a case report". *Annali Italiani di Chirurgia* 71 (2000): 717-720.
10. OEl Malki., *et al*. "Leiomyosarcoma of the inferior vena cava: diagnostic problem and treatment". *Progrès en Urologie* 13 (2003): 293-296.
11. Huguet C., *et al*. "Leiomyosarcoma of the inferior vena cava extending to the liver. Complete resection with venous reconstruction". *Gastroentérologie Clinique et Biologique* 16 (1992): 714-717.

12. Kabbaj N., *et al*. "Leiomyosarcoma of the inferior vena cava and nuclear magnetic resonance imaging". *Annals of Clinical Radiology* 39 (1996): 188-192.

Volume 9 Issue 6 August 2022
All rights reserved by O Belmaachi*., et al.*