Case Study of the Month

Pregnant Woman Presenting with a Gross Retroperitoneal Mass: Surgical Treatment with Caval Replacement

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A 40-year-old woman in the twenty-fifth week of pregnancy presented with a gross retroperitoneal mass. At the end of the pregnancy, the patient was submitted to surgery, and the gross infiltration of the inferior vena cava wall required the resection of the vena cava with its prosthetic substitution. The histopathological examination demonstrated the presence of a leiomyosarcoma of the inferior vena cava. An electronic video supplement showing the most important intraoperative passages is available online at doi:10.1016/j.eururo.2008.06.074.

1. Case report

We report the case of a 40-year-old woman in the twenty-fifth week of pregnancy, who presented sudden right flank and right lumbar pain. The woman underwent abdominal ultrasound examination and magnetic-resonance imaging, which evidenced the presence of a gross mass of apparent adrenal origin. The decision to go through with her pregnancy was made.

At the end of the pregnancy, the woman recovered in our urological department and was subjected to an angio-computer tomography (CT) scan, which confirmed the presence of a 12.5-cm mass. This large mass seemed to anteriorly dislocate the right kidney and to laterally dislocate the pancreas and the duodenum (Figs. 1–3). Moreover, the mass seemed to anteriorly dislocate the inferior vena cava (IVC), with such a compression that the flow was reduced to about 30%. The patient then underwent surgery.

Through a xifo-subumbilical incision, and after complete derotation of the intestinal mass, a gross mass was evident that had clearly infiltrated the IVC (Fig. 4). Initially, an attempt to excise only the mass was made, but due to the macroscopic infiltration of the IVC wall, the decision to resect the vessel became unavoidable. Therefore, the mass was...
entirely removed along with the right kidney, the right adrenal gland, and part of the IVC wall. After removal of the mass, the wall of the IVC and of the left renal vein were compromised. After having discussed the case with the vascular surgeon, the decision to resect a segment of the infrarenal IVC with a prosthetic substitution and to place a patch on the left renal vein was made.

Therefore, a widening polytetrafluoroethylene (PTFE) patch was placed on the left renal vein wall.

The resected segment of the IVC was replaced with a PTFE Gore-Tex 20 mm (W.L. Gore & Assoc, Flagstaff, Arizona) prosthetic substitution through a Prolene 4-0 suture (Fig. 5). (The video can be watched online at doi:10.1016/j.eururo.2008.06.074.) Surgical time was 340 min, with an IVC-clamping time of 65 min. Estimated blood loss was 2500 ml. No perioperative complications occurred, and the woman was discharged from the hospital 12 days after the procedure. Ten days after surgery, an angio-CT scan was repeated and showed the clarity of the prosthesis, with neither periprosthetic clots nor infection (Fig. 6).

The histopathological report showed a leiomyosarcoma of the IVC, G3, pN0, infiltrating the right perirenal and periadrenal tissues, with negative...
surgical margins. At 8 months after surgery, the patient is alive with no evidence of disease.

2. Discussion

Leiomyosarcoma of the IVC is an extremely rare disease, with about 300 cases reported in the literature, and it is more common in females (female-to-male ratio of 4.5 to 1) [1]. The presentation of this tumor is usually late, due to delayed appearance of the symptoms, which are usually not specific, such as abdominal pain, weight loss, mass, fever, weakness, and Budd-Chiari syndrome. Primary tumors of the IVC can occur in three different segments of the IVC: segment I (below the inflow of the renal veins), segment II (from the inflow of the renal veins to the inflow of the hepatic veins, excluded), and segment III (from the inflow of the hepatic veins up to the right atrium) [2].

In most of the reports, cases are referred by general and vascular surgery authors [2–4]. However, it has been reported that IVC leiomyosarcoma can mimic urological tumors [5]. Indeed, the preoperative imaging studies of this patient initially interpreted this tumor as a mass of adrenal origin.

Therapy of leiomyosarcoma of the vena cava consists of wide excision of the tumor [3]. Standard surgical resection of leiomyosarcoma en bloc with the affected portion of the IVC should attain the goals of complete excision of the tumor and preservation of venous return [3]. Technical difficulties are linked to tumor localization. There are controversies regarding the use of extracorporeal circulation and the prosthetic replacement of the IVC. We performed a wide resection of the interrenal and infrarenal vena cava with an associated right radical nephrectomy, without extracorporeal circulation, even if the cardiosurgical team was alerted for this possible strategy. The decision to replace the vena cava after the resection was made due to the absence of venous collateral circulation. The prosthetic substitution was performed using a PTFE prosthesis, which is the material of choice of many authors [6,7]. As an alternative, banked venous homograft replacement of the IVC, has been recently reported [8]. Right nephrectomy is frequently required for IVC leiomyosarcoma involving IVC segment II, even if the kidney is not directly involved [4].

The prognosis of patients affected by leiomyosarcoma of the IVC is variable, with up to 50% showing recurrence, and the majority of these recurrences occurring within 30 months of resection [9]. Three-year and 5-yr disease-specific survival rates are on the order of 76% and 33%, respectively [9]. There are controversies concerning the postoperative adjuvant therapy. However, to date, neither radiotherapy nor chemotherapy has demonstrated an improvement in either overall survival or local recurrence rates. Therefore, IVC leiomyosarcoma should be managed with wide surgical resection in order to achieve negative margins, when possible [10].

In conclusion, IVC leiomyosarcoma is a rare entity, but the misleading appearance of preoperative imaging and the retroperitoneal location of the
tumor must push urologists to be familiar with this disease.

Conflicts of interest

The authors have nothing to disclose.

Appendix A. Supplementary data


EU-ACME question

Please visit www.eu-acme.org/europeanurology to answer the below EU-ACME question on-line (the EU-ACME credits will be attributed automatically).

Question:

What is the currently recommended treatment option for the leiomyosarcoma of the inferior vena cava?

A. Radiotherapy and chemotherapy  
B. Surgical resection with adjuvant chemotherapy  
C. Surgical resection alone  
D. Surgical resection and adjuvant radiotherapy

References