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SHORT REPORT

Surgical Treatment of Intravenous Leiomyomatosis: The Role of the IVC Filter

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Introduction: Intravenous leiomyomatosis is a rare, life-threatening intravenous tumor associated with uterine leiomyomata.
Report: This report describes the case of a 45-year-old woman with a history of weakness and exertional dyspnea, and an extensive intracaval mass extending to the right side of the heart. The tumor was successfully removed in a two-stage surgical procedure with an inferior vena cava (IVC) filter deployed before the second stage. An extensive DVT was observed postoperatively.
Discussion: Surgical removal is the only effective treatment for intravenous leiomyomatosis, and the rate of recurrence remains unclear. An IVC filter should be placed routinely to prevent postoperative or late (in case of recurrence) pulmonary embolism.
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INTRODUCTION

Intravenous leiomyomatosis (IVL) is a rare condition associated with uterine leiomyomata. It consists of the benign proliferation of smooth muscle cells, starting at the uterine or gonadal veins, extending to the venous drainage system of the abdomen and pelvis, and potentially reaching the right heart and the pulmonary arteries. Although the tumor is nonmalignant, the obstruction to flow can lead to severe cardiopulmonary complications and death.

CASE REPORT

A 45-year-old woman presented with a 3-week history of weakness and exertional dyspnea. Transthoracic echocardiography revealed a large, free-floating mass in the chambers of the right side of the heart.

Computed tomography (CT) scan confirmed those findings, showing a thrombus-like hypodense mass (4.5 × 3.0 cm) in the right atrium extending to the right ventricle and involving the inferior vena cava (IVC) and both iliac veins. The renal veins were free, and pulmonary embolism (PE) was excluded. Transesophageal echocardiography excluded structural or functional changes of the heart (Fig. 1A).

The patient was submitted to a two-stage surgical procedure, first through a median sternotomy, cardiopulmonary bypass and moderate hypothermia, removal of the atrial tumor, and partial resection of the IVC mass was performed. The patient's recovery was uneventful.

Histology and immunohistochemical studies were compatible with uterine leiomyoma, and the diagnosis of IVL was established.

At the second stage, 7 weeks later, a laparotomy was performed for total hysterectomy, left salpingo-oopherectomy, and right salpingectomy. Additionally, the tumor was removed from the IVC and iliac veins, and a TRAPEASE IVC filter (Cordis; Johnson & Johnson, New Brunswick, NJ, USA) was deployed (Fig. 1B).

Histology confirmed uterine fibroleiomyomas similar to those previously removed from the heart.

Postoperative recovery was complicated by iliocaval deep vein thrombosis (DVT) below the IVC filter 3 weeks later. The patient presented with lower limb edema while undergoing prophylactic anticoagulation. This was treated successfully with therapeutic oral anticoagulation. (Fig. 1C).

At the 24-month follow-up the patient remained asymptomatic with no evidence of recurrent disease and showing recanalization of the thrombus on CT scanning.

DISCUSSION

IVL was first described in 1896 by Birch-Hirschfeld and later defined as a rare uterine neoplasm characterized by nodular masses of histologically benign smooth muscle growing within veins as worm-like projections.[1–3]

The etiology remains unclear and the clinical presentation varies widely.[1,4,5] Patients can be asymptomatic or

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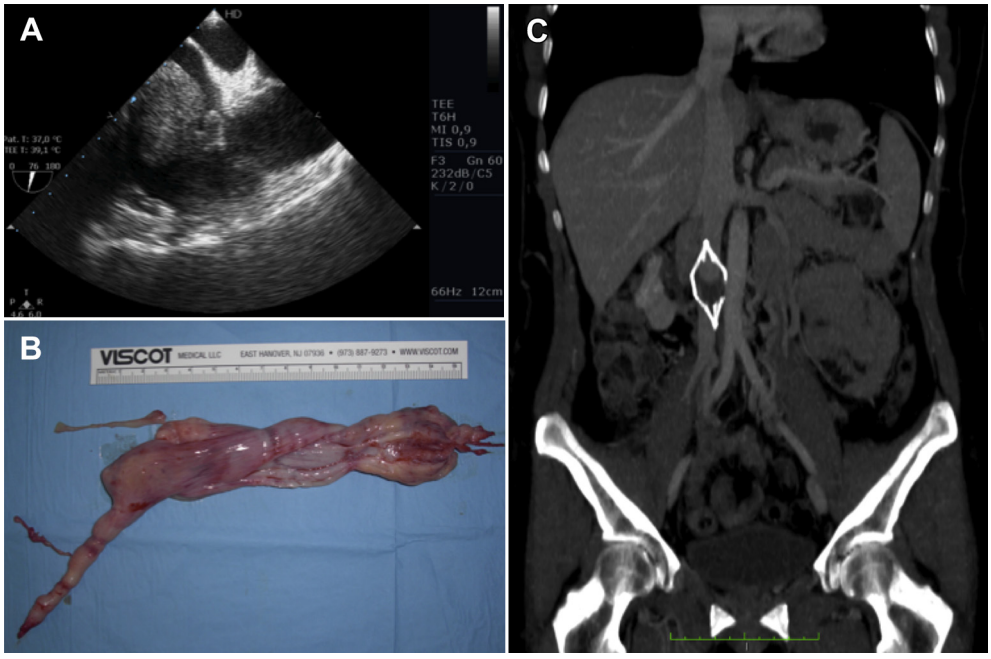

Figure 1. (A) Transesophageal echocardiography showing the intracardiac tumor in the right atrium. (B) Surgical specimen removed from the inferior vena cava (IVC) and iliac veins during the second procedure. (C) Computed tomography showing recanalization of the thrombus trapped in the IVC filter 4 months after the iliocaval deep vein thrombosis.

present with uterine leiomyoma without signs of venous invasion. Venous involvement can be a major problem, leading to complications such as Budd—Chiari syndrome, PE, outflow obstruction of the right side of the heart, heart failure, or even death.[1]

Surgical removal is the only effective treatment. Currently, there are two main surgical approaches for IVL with intracardiac extension: a one- or two-stage procedure.[2]

Given the patient's condition in this case, a two-stage approach, which is a less invasive and makes for a quicker recovery, was selected.

During the first stage, the intracardiac portion and part of the intracaval tumor are extracted through a median sternotomy. Depending on the patient's recovery, the second stage is performed via laparotomy as soon as possible. This stage includes the removal of the tumor form the IVC and the iliac veins, as well as hysterectomy and salpingo-oopherectomy.[2]

In this case, a decision to deploy an IVC filter was taken after completion of the second stage procedure in order to prevent postoperative PE—a decision considered to be appropriate owing to extensive DVT proximal to the IVC filter.

Much controversy has been raised concerning the ideal time for deployment of the filter. Some have defended retrograde placement, acting during the cardiac surgical procedure, as described by Barskdale et al.[3] Others have argued that the ideal time comes after completion of the second operation. This was what was performed in this case. However, whatever the option selected, the placement of an IVC filter seems to be required because of the high risk of DVT and PE associated with IVL resection.

Patients submitted to IVL resection extending to the IVC should have an IVC filter to prevent PE associated with venous thrombosis. IVL carries a risk of recurrence, and is unpredictable owing to it slow growth rate.[4] Patients with this condition should therefore be maintained under long-term clinical, ultrasound, and CT surveillance.

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CONFLICT OF INTEREST

None.

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