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CLINICAL INFORMATION

# Anesthetic approach for a clinical case of intravenous leiomyomatosis: case report

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**Abstract**
*Background and objectives:* The aim of this study was to describe the anesthetic approach in a case of intravenous leiomyomatosis with invasion of the inferior vena cava and extension to the right atrium, successfully treated with surgical approach.
*Case report:* Female patient, 45 years old, apparently stable until two weeks before the admission to the emergency department with complaints of fatigue and dyspnea. Echocardiogram was performed, which detected an intracardiac mass. Therefore, elective tumorectomy was performed in the vena cava-right atrium and right ventricle transition. Histological examination of the specimen suggested uterine leiomyoma. Subsequently, to assess the extent, computed tomography was done and showed extension to the inferior vena cava and left ovary. Hysterectomy, left adnexectomy and right salpingectomy, removal of intravenous leiomyoma, and permanent filter placement in the inferior vena cava were proposed. In this article, we describe the anesthetic approach with particular emphasis on the correction of fluid requirements, as well as postoperative evolution, and we highlight possible contributions to future approach of similar cases.
*Conclusions:* Preoperative diagnosis of intravenous leiomyomatosis is extremely difficult. Treatment consists of surgical removal. This procedure involves major blood loss and, therefore, preoperative preparation was conditioned: intraoperative fluid therapy was central and invasive monitoring considered predominant to assist in fluid and electrolyte balance of the patient; the existence of a clinical laboratory accessible and with rapid response and blood gas assessment was crucial; the intensive care unit equipped with ventilator for postoperative care proved to be another requirement resulting from this case.


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## Introduction

Intravenous leiomyomatosis (IL) is a smooth muscle benign and rare neoplasm, which is characterized by growing and invading the venous system. The smooth muscle proliferation occurs most often in the venous system; although, to a lesser extent, it also occurs in uterine fibroids.[1,2] IL has a slow growth pattern; first, it is localized and in the long run it creates metastasis by venous invasion. The latter begins in the uterine or ovarian veins and later moves to the iliac and inferior vena cava (IVC). The intracardiac extension occurs in about 10% of cases described and is often clinically undetectable. Cardiac presentation is largely confused with a primary cardiac neoplasia or migrant venous thrombus.[3]

The first description of an IL case with cardiac extension in medical literature was made by Birch-Hirschfeld in 1897.[4] According to Liu et al., by 2009, there were about 110 cases of IL with intracardiac extension described.[5]

IL generally occurs in women aged between 28 and 80 years, with an average 44 years.[4] Most patients have a history of hysterectomy or symptoms due to uterine fibroids. Although many patients remain asymptomatic for years, cardiac involvement may be manifested by symptoms of right heart failure or, less often, syncope due to tricuspid valve obstruction.[4,6] Other less frequent manifestations include Budd-Chiari syndrome with secondary thrombosis, ascites, sudden death, and systemic embolism.

Next, we will present the anesthetic approach of a patient with IL, IVC invasion and extension to the right atrium (RA), treated successfully through surgical approach.

We obtained permission from the patient to publish this case report.

## Case report

The patient, Caucasian and 45 years old, was apparently stable until two weeks before going to our hospital. At that time, she presented with generalized fatigue with progressively worsening and dyspnea upon minimal exertion, which led her to the emergency department. In this sequence, the patient underwent echocardiography, in which an intracardiac mass of unknown etiology was identified. For this reason, she was assessed by the cardiothoracic surgery team who proposed elective vena cava-right atrium and right ventricle transition.

The specimen resulting from surgery revealed, histological and immunophenotypic features compatible with fragment of uterine leiomyoma and, within the appropriate clinical context, consistent with intravenous leiomyomatosis. Therefore, clotting time and activated partial thromboplastin time (CT-aPTT) tests were made, which showed that the inferior vena cava beginning at the confluence of renal veins was distended and its lumen filled with a vascularized solid mass extending to the primitive iliac veins up to its bifurcation. It was also identified the increased dimensions of the uterus and a number of solid nodules with sizes ranging from millimeter to 9 cm. This larger lesion had a center with low uptake of contrast medium, while the remaining nodes had heterogeneous uptake, but predominantly hypervascular. A cyst with 4.4 cm was also observed in the left ovary.

After discussion between the general surgery and vascular surgery teams, it was propose total hysterectomy, left adnexectomy and right salpingectomy, removal of intravenous leiomyoma (vena cava, with extension to iliac veins) and permanent filter placement in the inferior vena cava in juxtarenal position.

It was in that sequence that the patient presented herself for pre-anesthetic evaluation. She weighed 75 kg, had a history of uterine leiomyomas, open angle glaucoma, multiple sclerosis with primary demyelination of the optic nerve, and ankylosing spondylitis. Her medication on an outpatient basis consisted of timolol, dorzolamide (5 $mg \bullet mL^{-1}$ + 20 $mg \bullet mL^{-1}$), varfine (5 mg), esomeprazole (20 mg/1 tablet every 8 h), magnesium metamizol (575 mg/1 tablet every 8 h), paracetamol (1,000 mg/1 tablet SOS), estazolam (2 mg/1 tablet at night). She also reported having undergone tonsillectomy 40 years ago, under balanced general anesthesia without perioperative complications, and was allergic to norflex.

The preoperative laboratory tests showed no changes worth recording. Enoxaparin (40 mg) was administered for prophylaxis of deep venous thrombosis. Airway was considered easy to approach. The anesthetic plan was discussed with the patient. Because she was evidently anxious, premedication with midazolam (3 mg) was administered. Balance general anesthesia (BGA) was proposed. In the operating room, peripheral venous access was obtained with a catheter (16 G), induction of anesthesia with fentanyl (0.5 mg) and propofol (85 mg). Ventilation with mask showed no difficulties. Tracheal intubation was facilitated by atracurium (30 mg) and performed by direct laryngoscopy, introducing an endotracheal tube nº 7.5, cuffed. BGA was maintained with a mixture of oxygen/air/desflurane and successive re-administrations of opioid. Ventilation was monitored by volume. Other peripheral access n° 16 G was obtained, and arterial catheterization in the left radial artery and central venous catheter in the right jugular vein were performed, all uneventful. Monitoring consisted of pulse oximetry, electrocardiogram, of carbon dioxide concentration in expiratory tidal volume, concentration of volatile anesthetic, fraction of inspired and expired oxygen, invasive blood pressure, central venous pressure, and urine output monitoring.

Total intraoperative analgesia was paracetamol (1 g) and fentanyl (3.25 mg). Prophylaxis of nausea and vomiting was made with droperidol (1.25 mg). During surgery, the Trendelenburg position was adopted.

During surgery, fluid requirements resulting from fasting, insensible losses, and gastric and urinary drainage were corrected. Blood loss from surgical field was corrected and crystalloids, colloids, and 10 units of packed erythrocytes, eight units of fresh frozen plasma, 2 g of fibrinogen, 1 pool of platelet concentrate were administered uneventfully, which helped maintain hemodynamic stability, with a mean arterial pressure of 79 mmHg, correct level of hemoglobin,

maintain diuresis and acid base balance within normal values.

There were no complications in the surgical procedure or anesthesia complications.

The approximate duration of anesthesia was 270 minutes and 200 minutes of surgery. The approximate blood loss was 3,400 mL.

According to the evolution in the postoperative period, the patient was kept under sedation and analgesia and volume-controlled ventilated and transferred to postoperative intensive care unit. She was extubated after 5 hours of post-anesthesia care and remained clinically stable and controlled from the standpoint of analgesia. She was discharged after 10 days without complications.

## Discussion

Preoperative diagnosis of IL is extremely difficult. The differential diagnosis is made with endometrial sarcoma, leiomyosarcoma, thrombus recanalization, atrial myxoma, renal cell carcinoma with intravascular invasion, adrenal carcinoma, and hepatocellular carcinoma.[1,2,4]

Proper treatment of this neoplasm is complete surgical removal. In cases of inadequate surgical margins, which may occur after surgical intervention, treatments such as Gn-RH analogues, oophorectomy or the ablation of ovarian function by irradiation have been advocated, as the growth on the natural history of the disease is slow.

Several surgical techniques have been described, such as extraction at one or two surgical procedures, with the use of cardiopulmonary bypass and circulatory arrest with deep hypothermia.[3,5-8]

In our clinical case, the surgical approach of IL was done in two surgical procedures with a month interval. The perspective of difficulty in diagnosing this pathology at an outpatient clinic is reinforced. The scarce frequency in the medical literature does not favor the standardization of surgical technique.

In the last decade, more cases of IL have been observed. In cases early diagnosed, with the evolution of imaging techniques and cardiopulmonary bypass, the surgical removal of the tumor is made in a single surgical procedure and with lower percentage of complications. Here, a transesophageal ultrasound seems to have a role.[4,6-9] Kullo et al. have emphasized the morphological characteristics of intracardiac IL and reported that the presence or absence of adhesions at the right atrium determines the intervention. In the absence of adhesions, laparotomy with venotomy may be sufficient to remove the tumor, in contrast to the sternotomy and cardiopulmonary bypass, in the presence of adhesions latter.[4,10]

The first intervention by the cardiac surgery team was directed to the uterus, the primary origin of the tumor. In the second intervention, to which we restrict ourselves, important aspects should be noted. This is a surgery that involves major blood loss that affects the proper preoperative preparation, in which we stress the discontinuation of oral anticoagulation, with subcutaneous replacement. The anticipation of problems of anemia and haemostasis put us in touch with the Immune-Hematology Service. Intraoperative fluid therapy is central to the outcome. Invasive monitoring, particularly the arterial line to maintain the tension profile; analytical control and central venous catheter control; central venous pressure monitoring; administration of blood products; and fluid therapy administration are crucial for fluid and electrolyte balance.

This type of surgery requires a good communication between the surgical and anesthetic teams. The sequential blood gas assessment requires functional equipment nearby, with adequate response time. Accessible to a clinical laboratory with quick response is important. A multidisciplinary and well trained team is imperative, with intensive care unit equipped with ventilator.

Fortunately, preoperative hemodynamic stabilization difficulties were not observed in our patient. Capnography showed no anomalous change of the curve or reduced values of $etCO_2$. Pulmonary embolism is described in IL cases. Kim D et al. reported a case of a patient with IL who underwent surgery and detected a non-characteristic capnography curve, which was assigned to the mobile and slack nature of tumor tissue, as well as its location. The authors recommend transesophageal echocardiography to exclude pulmonary embolism.[11]

The follow-up of these patients is essential for the reduction of morbidity and mortality in IL, because they are associated with recurrence of 30%.[4,11]

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