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# Multidisciplinary Management of Intravenous Leiomyomatosis: A Case Report

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**Running Title** Intravenous Leiomyomatosis Multidisciplinary Management

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## ABSTRACT

Intravenous leiomyomatosis (IVL) is an unusual smooth muscle tumor characterized by benign smooth muscle cells growing within blood vessels, often reaching the inferior vena cava, right atrium, and pulmonary vessels. The best treatment is complete surgical removal, and the outlook is typically positive, with recurrence occurring rarely. This report discusses a 38-year-old woman who was unexpectedly found to have intravenous leiomyomatosis extending from her uterus to the inferior vena cava and right atrium. She underwent one-stage surgery, which included total hysterectomy, bilateral salpingo-oophorectomy, and removal of the lesions from the inferior vena cava and right atrium, all without the need for thoracotomy.

## Introduction

Leiomyomas are benign tumors that show various forms of smooth muscle differentiation [1]. There is a wide range of leiomyoma types. Usually, these tumors are benign and confined to the uterus, but sometimes they can cause complications. Some types have histological traits linked to leiomyosarcoma, such as a high mitotic index and significant cellular abnormalities, while other variants can spread to the peritoneum or solid organs but remain histologically benign. Intravenous leiomyomatosis (IVL) is marked by the growth of benign smooth muscle that can extend into the uterine and pelvic veins, the vena cava, and even the heart [2].

The presentation can vary greatly, requiring multiple imaging modalities for a comprehensive evaluation of the retroperitoneal space and the heart.

IVL mostly occurs in women of reproductive age, although a few cases have been reported in postmenopausal women [3]. Symptoms can include pelvic, abdominal, or cardiac issues, depending on the extent of tumor spread. Pelvic or abdominal symptoms may consist of pain, abnormal bleeding,

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and abdominal distension [4]. Some patients may experience severe shortness of breath, an intracardiac mass, or signs of right heart failure (congestive heart failure, dyspnea, and syncope) [5].

Diagnosis of IVL is made through imaging such as computed tomography (CT) or magnetic resonance imaging (MRI), which reveals a deformed or leiomyomatous uterus with projections into blood vessels. Cardiac evaluation is conducted using echocardiography [6]. Most cases require surgical removal of the mass [7].

A multidisciplinary approach is usually necessary to perform the surgery in one stage using cardiopulmonary bypass.

## Case Presentation

A 38-year-old woman, gravida 1 para 1, was found to have a heterogeneous mass in her right adnexa during an ultrasound at a prenatal visit. Transvaginal ultrasound and Doppler imaging revealed a solid-cystic mass measuring 68 × 77 × 81 mm with internal blood flow in the right adnexa.

Abdominopelvic multidetector computed tomography (MDCT) identified a cystic-solid mass measuring 78 × 50 × 50 mm in the right adnexa, along with thrombosis in the right iliac vein and gonadal vessels extending into the inferior vena cava (IVC) and the inferior part of the right atrium, accompanied by mild free fluid.

Echocardiography was performed, which revealed a large multilobulated homogeneous mass (3.5 × 3.2 cm) in the right atrium that intermittently pushed into the right ventricle through the tricuspid valve. Given that the mass was located in the right atrium, a cardiovascular surgical consultation was requested (Figure 1).

A surgical team consisting of a vascular surgeon, cardiothoracic surgeon, gynecologic oncologist, and anesthesiologist coordinated the surgery. Before the operation, a consultation was held with the surgical team, the patient, and her husband to discuss the risks and benefits, the potential need for thoracotomy, open-heart surgery, and the necessity of hysterectomy and bilateral salpingo-oophorectomy. Both the patient and her husband agreed to proceed with the surgery.

The procedure was scheduled in an operating room equipped for open-heart surgery. A midline laparotomy was performed by the gynecologic oncology team. There was no ascites, and the peritoneal surface appeared normal. The uterus measured approximately 8 × 12 cm and had an indistinct mass on the sides and back. The ovaries appeared normal.

During the hysterectomy, a worm-like soft mass was found inside the uterus, extending into the uterine, ovarian, and infundibulopelvic vessels and the pelvic floor. The tumor had also invaded the internal iliac vein and the IVC. The patient underwent total hysterectomy and bilateral salpingo-oophorectomy.

After completing the hysterectomy, the vascular and cardiothoracic surgery teams began their procedures simultaneously. The incision was extended up to the xiphoid process, and upon entering the abdominal cavity, a standard exploration was performed. We opted to proceed without using cardiopulmonary bypass. Throughout the operation, the cardiac surgery team remained on standby for any urgent interventions if necessary. Following the dissection of the linea alba, the right colon was mobilized and shifted to the left side, allowing full exposure of the accessible inferior vena cava, from the lower edge of the liver down to the iliac veins.

The renal and gonadal veins were controlled with tapes, along with the infrahepatic segment of the IVC. A venotomy was performed on the IVC with an incision length of 8 to 10 cm. The proximal portion of the mass was gently pulled downward through the vessel lumen, allowing easy extraction of the mass from the right atrium with minimal resistance. Subsequently, the proximal IVC lumen was closed using a circumferential tape, which rapidly controlled the bleeding. Intraoperative transesophageal echocardiography confirmed complete removal of the mass from the right atrium.

In the second stage of the procedure, the mass was removed from the distal IVC by applying continuous gentle traction, extracting it from the iliac and gonadal veins. The shape of the mass indicated that the tumor was entirely removed (Figure 2). The venotomy site was then repaired, and the abdominal wall was closed. The estimated blood loss during the surgery was approximately 700 to 1000 cc. The patient was transferred to the intensive care unit and recovered quickly, without complications related to intraoperative bleeding. She was discharged four days later with a prescription for new oral anticoagulant (NOAC) therapy.

The pathologist's report of the inferior vena cava tumor specimen confirmed an intravenous leiomyoma with hyaline degeneration (Figure 3).

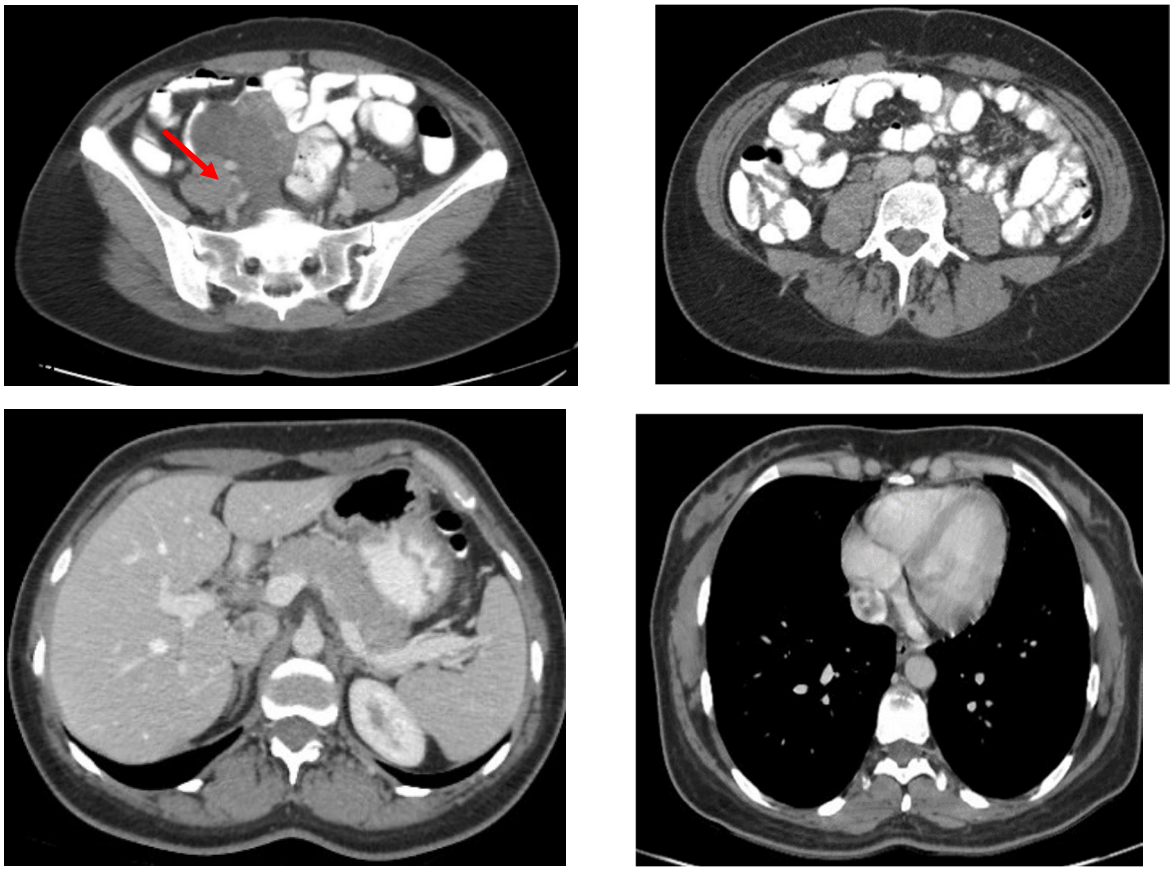

**Fig. 1.** Abdominopelcic CT scan with IV and PO contrast axial images: a) at pelvic level, there is right adnexal cystic lesion.filling defect in right external illiac vein is present (red arrow). b) at distal IVC level, filling defect in IVC is visualized.c) IVC at the level of hepatic vein confluence reveals luminal thrombosis.d)suprahepatic IVC shows luminal filling defect

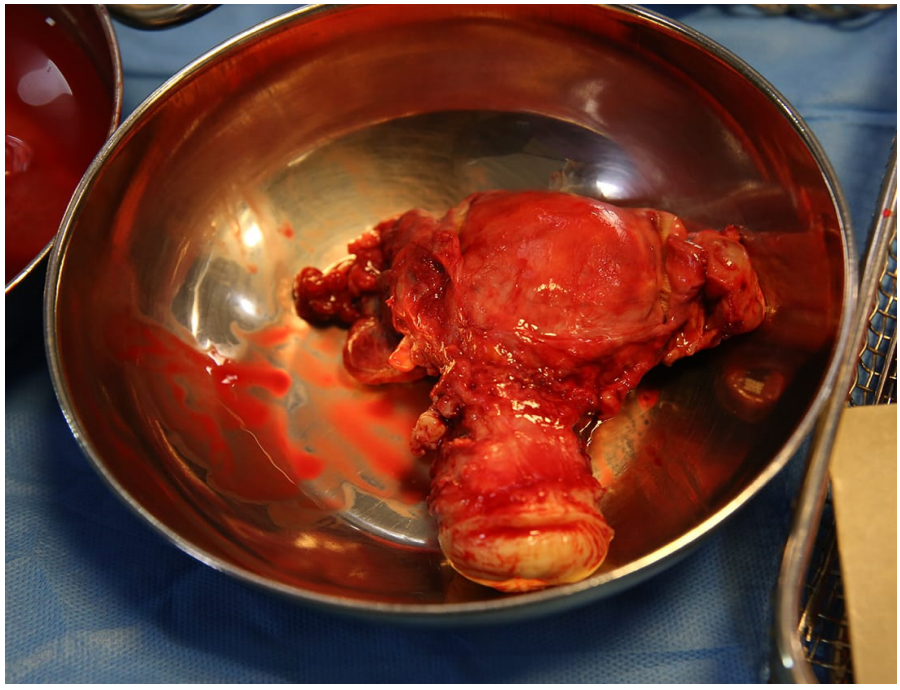

**Fig. 2.** Total abdominal hysterectomy and bilateral salpingo-oophorectomy

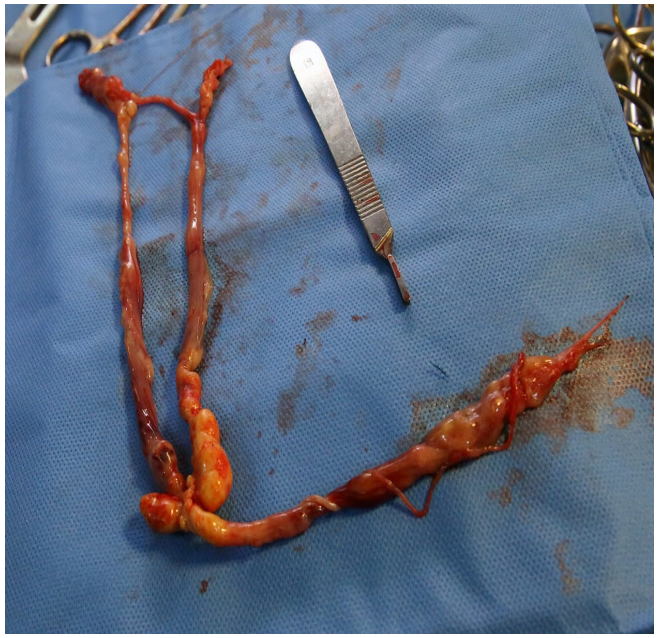

**Fig. 3.** Gross specimens of intra-cardiac and vena cava lesions after extraction

### Pathological Report

#### Macroscopic Examination of the Sample

The sample labeled "IVC mass" includes several pieces of stringy tissue measuring 25 cm in total length. In the center of this stringy tissue, there is a cream-white to cream-yellow mass with a solid cross-section and a relatively firm consistency, measuring 4.5 × 3.5 × 2 cm. The boundaries of the mass are well defined.

#### Microscopic Examination of the Sample

Tumoral cells were seen in a diffuse and whorled pattern, with uniform elongated nuclei and wide areas of hyaline degeneration. Mitotic activity is negligible. In IHC staining number 416, the following results were obtained:

- **SMA:** Positive
- **Desmin:** Positive
- **S100:** Negative
- **CD34:** Negative
- **STAT6:** Negative
- **Ki67:** Positive about 1% of nuclei

### Diagnosis

**DX: Inferior vena cava tumor**

- Intravenous Leiomyoma with hyaline degeneration

## Discussion

IVL is an uncommon condition marked by the growth of benign smooth muscle cells within blood vessels. It most often originates from the uterus and can extend through the pelvic veins and inferior vena cava, occasionally reaching the right atrium, ventricle, and main pulmonary artery. To date, around 300 cases have been documented, with fewer than 100 involving cardiac extension. No prospective studies have been conducted in this area so far [8,9].

IVL most commonly affects women in their fifth decade of life, and its symptoms vary depending on the extent and location of tumor spread. This condition should be considered as a possible diagnosis before gynecological surgery, especially when a patient presents with a large pelvic mass or a broad ligament fibroid [10].

Signs and symptoms of IVL include pelvic pain and abnormal uterine bleeding caused by a myomatous uterus; swelling of the lower limbs due to obstruction of the IVC; chest pain and shortness of breath resulting from pulmonary embolism; fainting episodes caused by tumor obstruction at the tricuspid valve opening; electrocardiogram abnormalities linked to enlargement of heart chambers, impaired systolic function, and involvement of cardiac valves; and sudden death caused by blockage of the tricuspid

valve or the right ventricular outflow tract [11].

Since intracardiac extension is uncommon and patients with IVL often remain asymptomatic until the tumor reaches the heart, early detection of the condition is challenging [12].

Typical imaging techniques for assessing uterine intravenous leiomyomatosis include ultrasonography (US), magnetic resonance imaging (MRI), and computed tomography angiography (CTA). Preoperative imaging evaluations often involve echocardiography, thoracic CT, and abdominopelvic CT, as well as magnetic resonance venography or CT angiography to determine the extent of the disease and plan surgical intervention [13].

The most effective treatment for the tumor is surgical excision. Depending on various factor such as the patient's ability to tolerate surgery, the tumor's anatomical characteristics, and the surgeon's expertise a single-stage or two-stage surgical approach may be selected [14,15]. Sternotomy combined with cardiopulmonary bypass, along with laparotomy performed either in a single or two-stage procedure, may be utilized to manage tumors extending into the cardiovascular system [8]. Many studies consider hysterectomy with bilateral salpingo-oophorectomy to be the standard treatment approach [16].

In this case, the patient was incidentally found to have a mass in the right adnexa during pre-pregnancy evaluation. Further comprehensive diagnostic workup revealed that the lesion had extended into the inferior vena cava and the right atrium. Consequently, a multidisciplinary surgical plan was developed. The patient underwent a single-stage surgery, beginning with hysterectomy and bilateral salpingo-oophorectomy. Without performing thoracotomy or using cardiopulmonary bypass, the laparotomy incision was extended, and the mass was removed via venotomy of the inferior vena cava. The incision was then extended upward to the sternum to open the vena cava, allowing extraction of the intravenous tumor from both the vena cava and right heart through this approach, thereby avoiding thoracotomy. Eighteen months postoperatively, the patient remains in good general condition with no signs of disease recurrence on clinical examination or imaging studies.

The timing of postoperative recurrence differs among patients. As a result, long-term monitoring is strongly advised, with recurrence surveillance conducted through ultrasound, transthoracic echocardiography, and CT scans [17].

## Conclusion

Intravenous leiomyomatosis should be suspected in patients with uterine leiomyomatosis, particularly when the tumor involves the broad ligament. Timely and appropriate imaging, such as pelvic Doppler ultrasound, is essential for accurate diagnosis and assessment of tumor extent. Moreover, in cases where the tumor extends into the heart, thoracotomy may be avoidable under certain conditions, allowing for less invasive surgical management.

## Ethical Considerations

### Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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### Conflict of Interests

The authors have no conflict of interest to declare.

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