

Intravenous leiomyomatosis disguised as a large deep vein thrombosis

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ABSTRACT

Intravenous leiomyomatosis is a benign smooth muscle tumor which despite its histology can have devastating consequences. Furthermore, the clinical manifestations are variable and nonspecific, typically leading to delayed or missed diagnosis. Thus, it is critical for clinicians to be aware of this condition and have a high index of suspicion in a middle-aged woman with a history of uterine leiomyoma presenting with an inferior vena cava mass to enable early diagnosis and treatment. We report a case of a large intravenous leiomyoma which was initially considered to be a very large deep venous thrombosis; with thorough preoperative planning, it was successfully removed intact and in entirety with a single-stage operation.

CASE REPORT

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Clinical & Imaging findings

A 53 year old Caucasian woman presented to her primary care physician reporting dyspnea on exertion, 10 pound weight loss, and myalgias. She had hypertension but no other prior medical or surgical history, and she took no regular medications. A mass was palpable posterior to the uterus, but her physical exam was otherwise unremarkable. Laboratory abnormalities included AST 181 (normal, 8-41 units/liter) and ALT 365 (normal, 12-48 units/liter). This prompted an abdominal ultrasound (Figs. 1a and 1b), which showed a clot extending from right the gonadal vein into the inferior vena cava. To further characterize this mass, CT of the abdomen and pelvis was obtained, which showed marked uterine enlargement with several masses, the largest of which measured approximately 6cm in maximal diameter. Additionally, dilation of the right gonadal vein was noted, with what was initially presumed to be thrombus extending the full length of the right gonadal vein into the inferior vena cava.

(Figs. 2a-e). Upon further review of the CT scan, the differential diagnosis was expanded to include intravascular tumor rather than thrombus given the enhancement pattern. Transthoracic echocardiogram was performed to further characterize motility and composition of the structure, which showed a large amount of echogenic material in the inferior vena cava extending to approximately 2cm from the caval-atrial junction. This structure was very mobile and appeared more compatible with intravascular tumor than thrombus (Figs. 3a and 3b). Furthermore, Doppler studies of the bilateral lower extremities were obtained which showed no evidence of deep venous thrombosis (Figs. 4a-d). An MRI was not obtained as part of the pre-operative evaluation.

Management

She was started on therapeutic anticoagulation and was taken to the operating room and underwent laparotomy with total abdominal hysterectomy and bilateral salpingo-oophorectomy. Given the considerable embolic risk by either tumor or clot, she also underwent a planned sternotomy,

cannulation and cardiopulmonary bypass, circulatory arrest, and removal of a large, mucoid inferior vena cava mass intact and in entirety from its origin at the right gonadal vein (Figs. 5a and 5b). Final pathology was consistent with intravenous leiomyomatosis with no evidence of thrombus. She also had small implants of leiomyoma within the uterine veins.

Follow-up

She had no further indication for anticoagulation and had an uneventful postoperative recovery.

DISCUSSION

Introduction:

Uterine leiomyomas are extremely common benign tumors found in approximately 30% of reproductive age women. Rarely, these neoplasms exhibit unusual growth patterns that are beyond the domain of a gynecologist [1, 2]. One such manifestation is intravenous leiomyomatosis (IVL), an uncommon condition wherein the smooth muscle tumor grows into venous channels without invading the tissues [3, 4]. This tumor is typically confined to the pelvis but can sometimes extend into the thoracic cavity or heart via the inferior vena cava [5]. IVL was first described in 1896 by Birch-Hirschfeld, and since then, less than 300 cases have been reported in the English literature [6, 7]. Herein we present a case of intravenous leiomyomatosis grossly extending from the origin of the right gonadal vein and extending to the proximal inferior vena cava, which was initially thought to represent a very large intracaval thrombus.

Etiology & Demographics:

The pathophysiology of IVL is not clearly established, but two theories have been proposed [8, 9]. One theory suggests that the neoplasm arises from a vessel wall. The other theory implies that the leiomyoma directly extends into the myometrial veins and can further spread to other veins by contiguous growth within the lumen. The latter explanation seems more applicable to the index case. The tumor most commonly enters the lumen of the iliac vein and extends from there to the inferior vena cava; in some cases, it may reach the right atrium, ventricle, or pulmonary artery [8, 9].

IVL occurs exclusively in females. A review of the literature demonstrates that the majority of women with IVL have previously undergone hysterectomy [10]. Our patient had not undergone hysterectomy, but not surprisingly, she had leiomyoma within the uterus. The median age at presentation is 47 years old, but cases have been reported in women ranging 26 to 76 years of age. Because this is a hormone-dependent tumor, most (but not all) patients are premenopausal and multiparous [2, 11].

Clinical & Imaging Findings:

Clinical manifestations are various and nonspecific [12]. Patients may be asymptomatic with IVL discovered only incidentally, or IVL may present with symptoms that mirror the distribution of IVL extension. For example, patients with inferior vena cava occlusion may have lower extremity edema

and abdominal swelling, whereas those with intracardiac obstruction may have dyspnea, chest pain, congestive heart failure, or even sudden death [6, 13].

Echocardiography is helpful in defining the distal extent of the lesion. Computed tomography and magnetic resonance imaging offer cross-sectional views with multiplanar capability and thus are the most useful imaging modalities for IVL. Magnetic resonance imaging is particularly helpful in distinguishing IVL from bland thrombus by visualization of heterogeneous enhancement after administration of intravenous contrast, unlike a bland thrombus which would not enhance [2, 4]. Despite imaging findings, a definitive diagnosis relies on histopathology [6, 11].

Treatment & Prognosis:

The treatment of choice is radical surgical resection of the entire lesion with a multidisciplinary team to remove the pelvic organs (uterus, fallopian tubes, ovaries, and any parametrial leiomyoma), intravascular tumor, and any intracardiac components. Prior authors have compared one-stage versus two-stage operations to separate chest surgery from abdominal-pelvic surgery with varying results [14-16]. Wang et al. reported on 30 patients treated for IVL at a single center, the largest known series to date, and found that patients with a good performance status and minor or no intracardiac extension can safely undergo a single-stage operation [12]. At this time, it seems uncertain and variant upon individual characteristics as to whether a single-stage or two-stage operation is superior to remove IVL.

In the study by Wang et al, recurrence rate was 30% and notably was significantly higher in patients who underwent incomplete rather than complete resection (50.0% versus 21.4%, $p=0.016$) [12]. Similar rates to that reported by Wang et al. have been previously reported in the literature [17]. Complete resection and adjuvant anti-estrogen therapy appear to be the cornerstone of prevention or minimization of extent of recurrence [12, 18].

Differential Diagnosis:

Upon noting an intravascular mass within the inferior vena cava, differential diagnoses include intravenous thrombosis, right atrial myxoma, leiomyosarcoma, malignant tumor embolus, and intravenous leiomyoma [6, 12]. Detailed preoperative imaging may help to reveal the underlying lesion, except in the case of leiomyosarcoma, where histopathology is required for definitive diagnosis as these tumors appear radiographically indistinct. Intravenous thrombi ("bland thrombi") appear similar to IVL on CT scan or pelvic ultrasound but differ in that they do not enhance after administration of gadolinium-based contrast with MR [4]. Right atrial myxomas are usually confined to the cardiac chambers and do not involve the IVC; they have a characteristic appearance of a narrow-based mass attached to the inter-atrial septum. Additionally, they have no association with uterine fibroids [6]. Malignant tumor emboli most commonly arise from renal cell carcinoma; CT or MR of the abdomen enables visualization of the primary tumor, which typically grows in an antecedent fashion and involves the renal veins [4,6].

TEACHING POINT

Although intravenous leiomyomatosis is rather uncommon, it typically occurs in a specific clinical setting, and thus physicians and other providers should include this condition in their differential diagnosis of an intravascular mass in a reproductive age woman regardless of symptoms.

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FIGURES

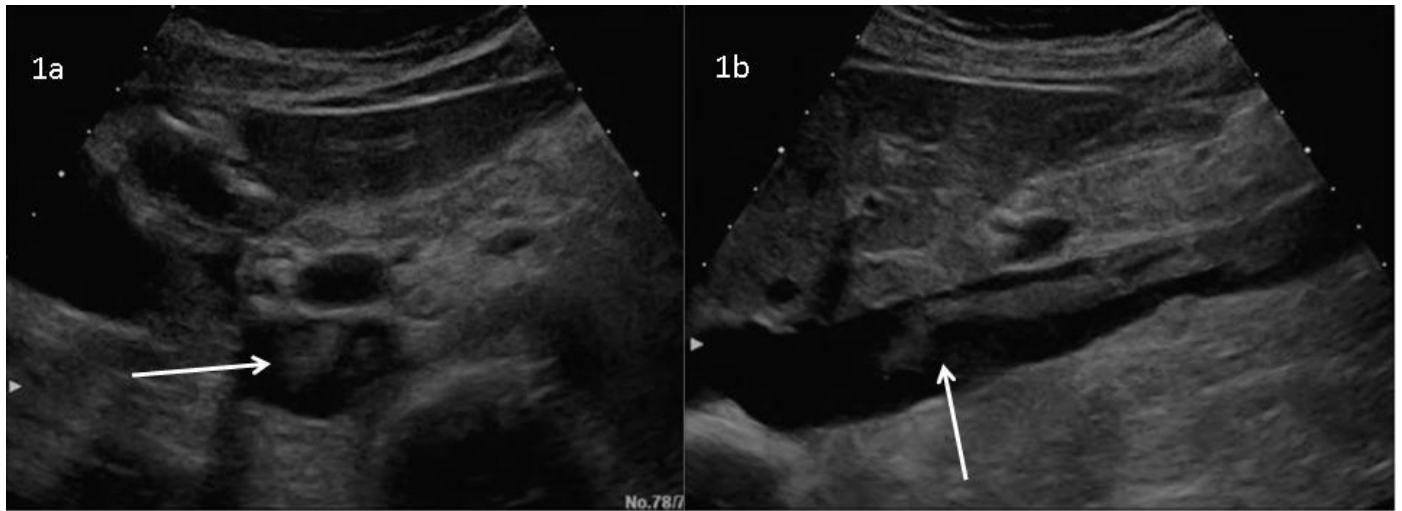


Figure 1: 53-year-old female with a large leiomyoma with extension into the right gonadal vein and inferior vena cava. Findings: Real time longitudinal and transverse images focused on the right upper quadrant of the abdomen. 1a) Transverse image demonstrating an echogenic filling defect within the inferior vena cava (white arrow). 1b) Longitudinal image demonstrating an echogenic filling defect within the inferior vena cava (white arrow). TECHNIQUE: Longitudinal and transverse sonographic images, focused on the right upper quadrant of the abdomen using a convex 5-1 MHz transducer.

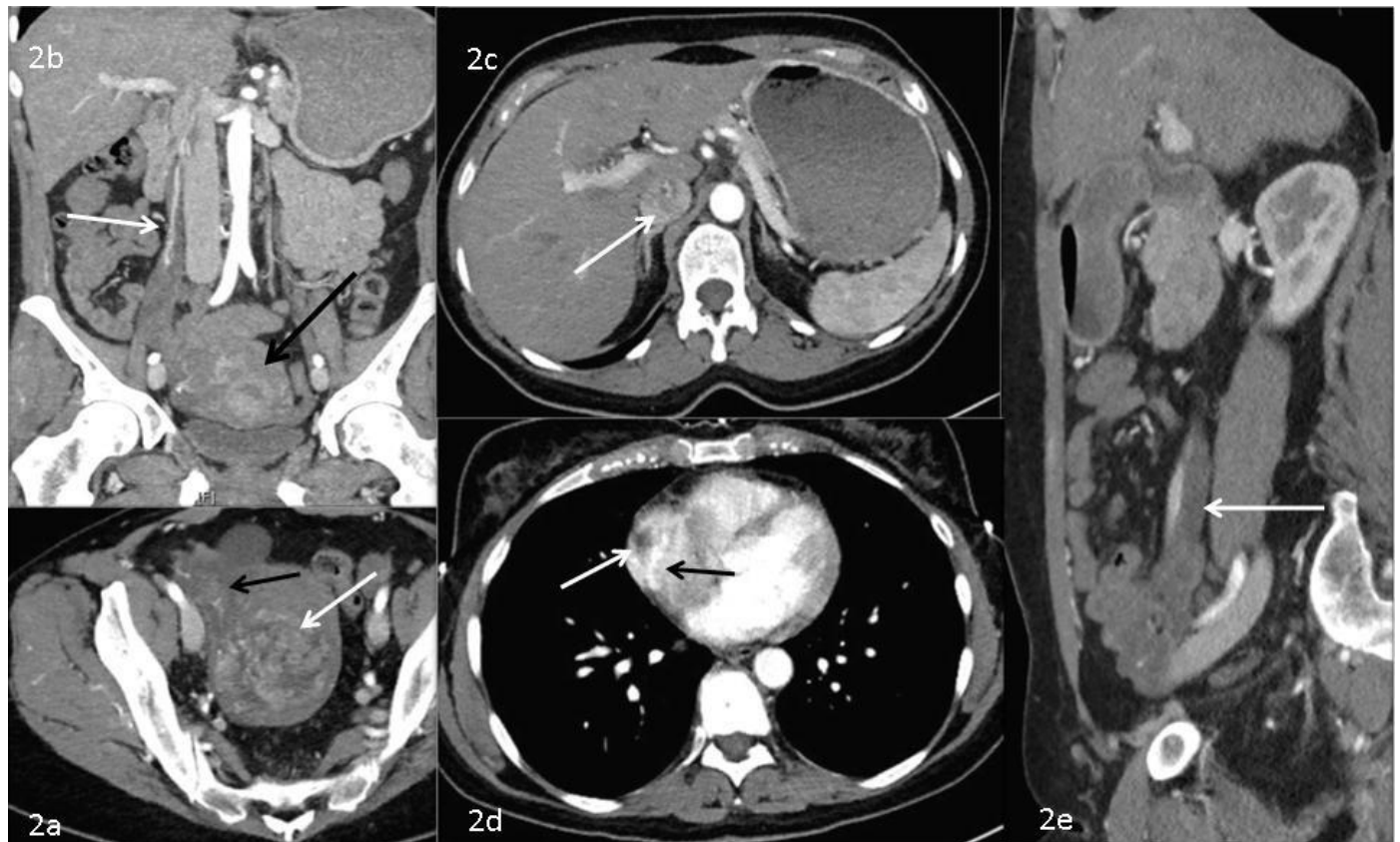


Figure 2: 53-year-old female with a large leiomyoma with extension into the right gonadal vein and inferior vena cava. Findings: Axial, coronal, and sagittal contrast enhanced images of the abdomen and pelvis. 2a) Axial image demonstrates a large uterine fibroid within the posterior fundus (white arrow) with extension into the right gonadal vein (black arrow). 2b) Coronal image demonstrating soft tissue density with linear arterial contrast enhancement within the right gonadal vein (white arrow). Large uterine fibroid (black arrow). 2c) Axial image demonstrating soft tissue density with central arterial contrast enhancement within the proximal inferior vena cava (white arrow). 2d) Axial image demonstrating hypoattenuating soft tissue (white arrow) with adjacent linear contrast enhancing tissue within the right atrium (black arrow). 2e) Sagittal image demonstrating soft tissue density with linear arterial contrast enhancement in the right gonadal vein (white arrow). TECHNIQUE: Axial, Coronal, and Sagittal contrast enhanced CT, kV 100, mAs 185, 3mm slice thickness, 85cc of Isoviev 370 intravenous contrast.

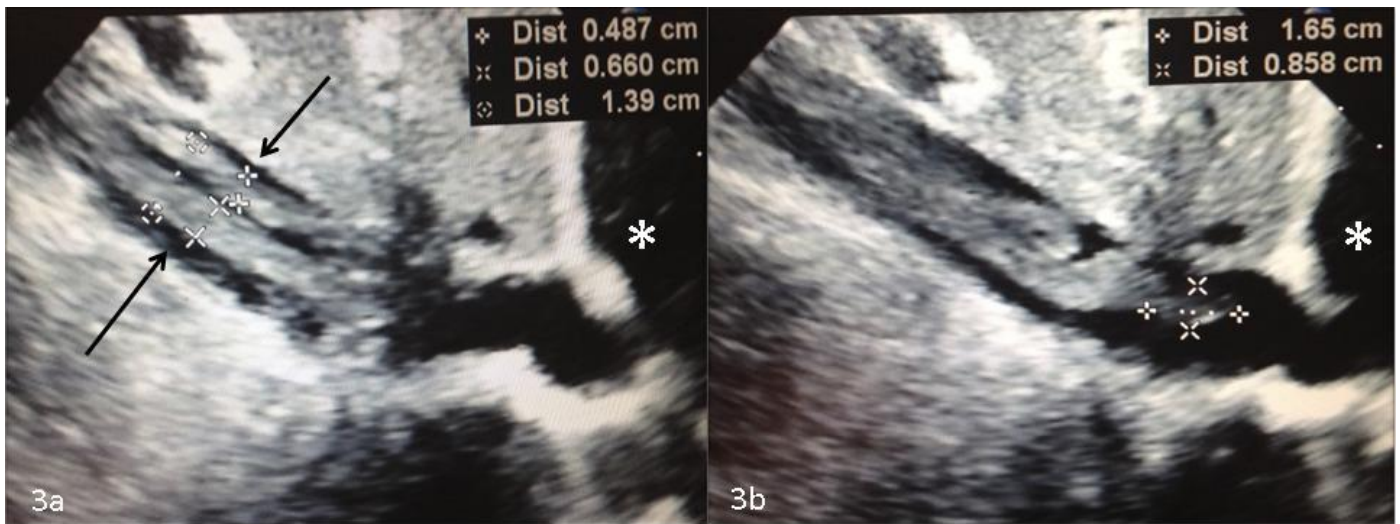


Figure 3: 53-year-old female with a large leiomyoma with extension into the right gonadal vein and inferior vena cava. Findings: Transthoracic echocardiogram images were obtained. Right atrium (*). 3a) Two linear, echogenic soft tissue densities (black arrows) demonstrated in the inferior vena cava. 3b) The distal aspect of the echogenic soft tissue density is located within the inferior vena cava approximately 2cm from the right atrium (*). **TECHNIQUE:** Echocardiography using M-mode, complete 2D, complete spectral Doppler and color Doppler with a 5-1 MHz phased array transducer.

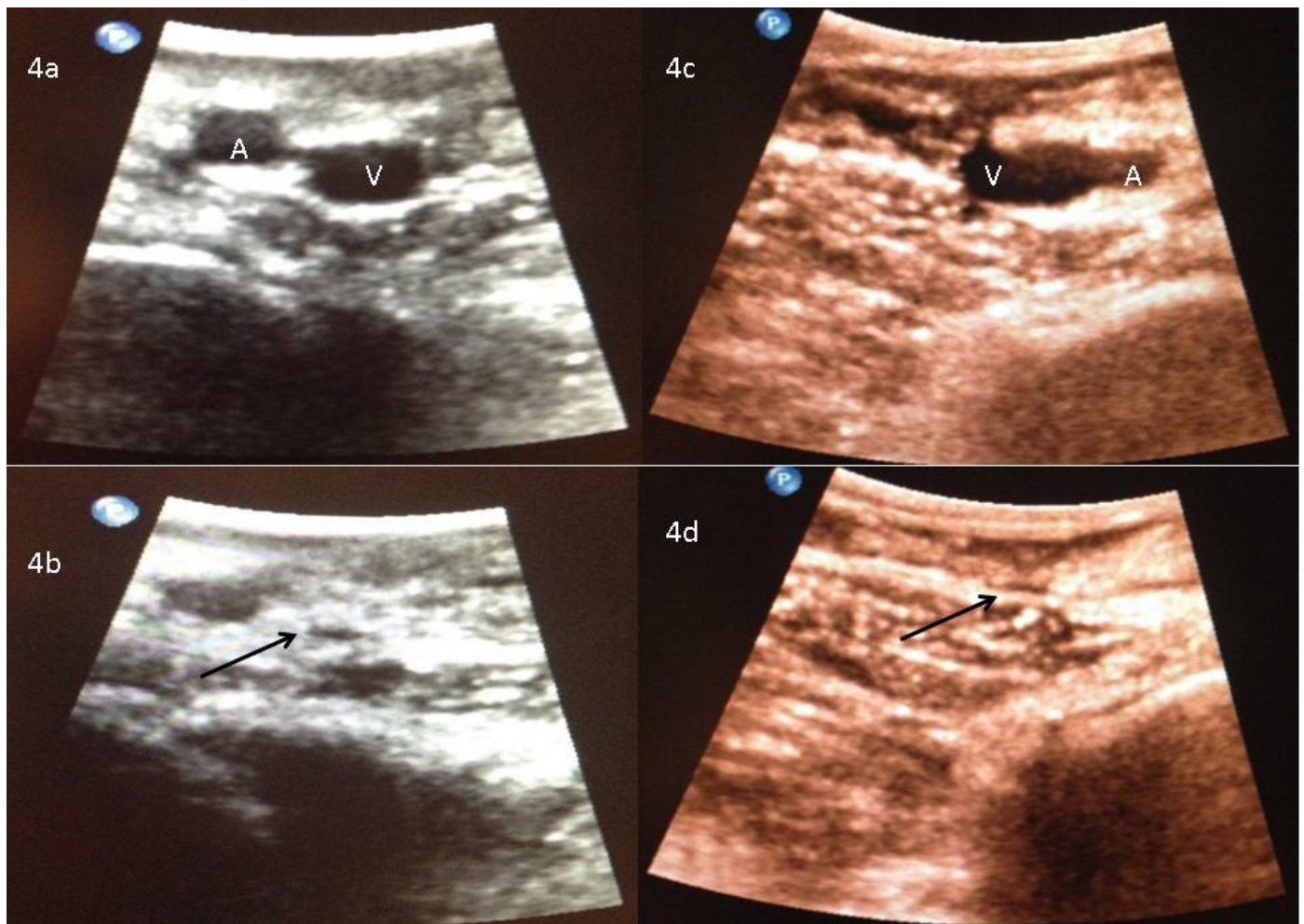


Figure 4: 53-year-old female with a large leiomyoma with extension into the right gonadal vein and inferior vena cava. Findings: Transverse and longitudinal sonographic images of the bilateral lower extremities. 4a) Right common femoral artery (a) and vein (v) which appear patent without evidence of thrombus. 4b) Compression of the right common femoral vein (black arrow). No evidence of thrombus. 4c) Left common femoral artery (a) and vein (v) which appear patent without evidence of thrombus. 4d) Compression of the left common femoral vein (black arrow). **TECHNIQUE:** Longitudinal and transverse sonographic images of the bilateral lower extremities using a 5.0-13.0 MHz linear transducer.

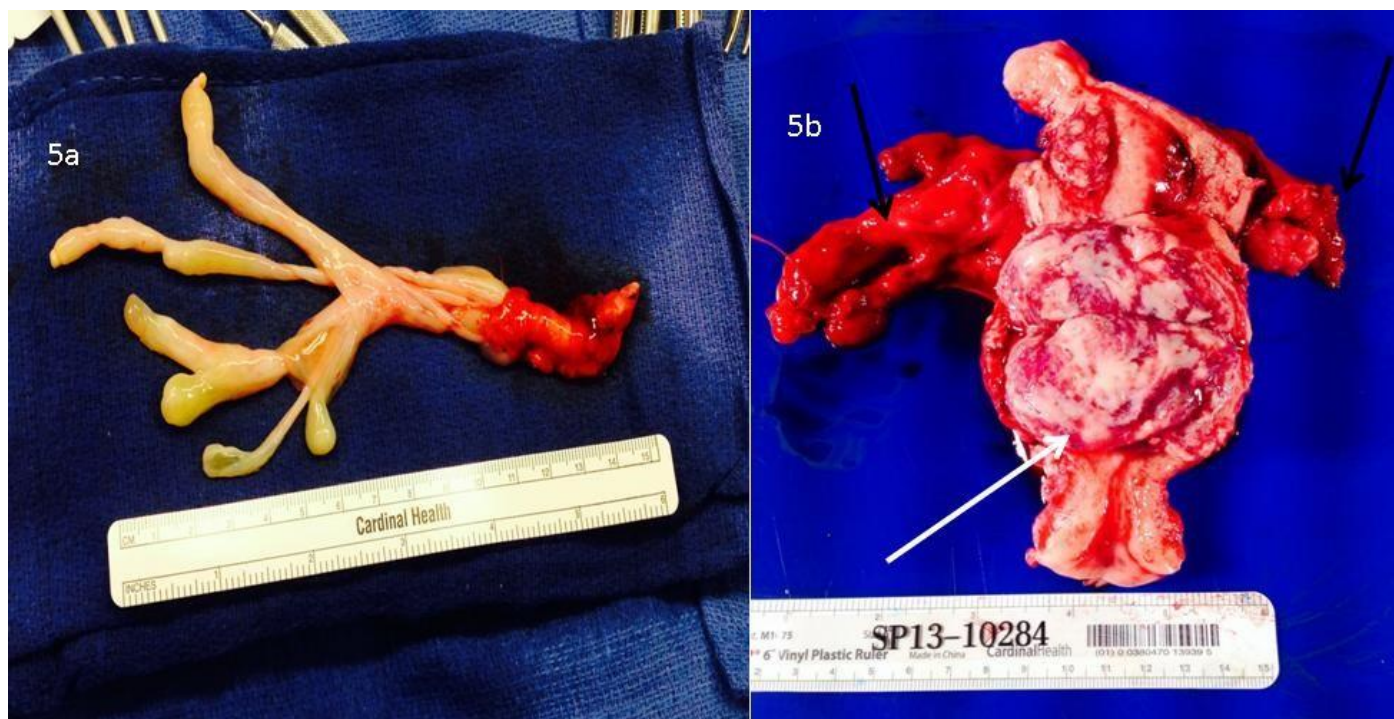


Figure 5: 53-year-old female with a large leiomyoma with extension into the right gonadal vein and inferior vena cava. a) Removed intraoperatively is a yellow-tan, smooth, glistening mass measuring 11.8 x 2.2 x 1.3cm recapitulating the vascular branches from which it was removed. 5b) The gross specimen includes the opened uterine cavity with a bulging 6cm, white, whorled mass (white arrow) displacing the endometrial cavity. Also noted are the bilateral adnexa (black arrows).

Etiology	Benign smooth muscle tumor thought to either arise from a vessel wall or directly extend into myometrial veins and then further extend into other veins via contiguous intraluminal growth
Incidence	Rare; no exact incidence defined as only 300 cases have been reported
Gender Ratio	Occurs solely in females; no analogous process has been identified in males
Age predilection	Typically occurs in reproductive aged women; included within the literature are reports of women age 26-76
Risk factors	<ul style="list-style-type: none"> • Uterine fibroids • Reproductive age • Multiparity
Treatment	Radical surgical resection with excision of the entire lesion via a multidisciplinary team (gynecology, vascular surgery, cardiothoracic surgery) with either a one-stage or two-stage operation
Prognosis	Typically excellent. Recurrence is ~30% but is more common with incomplete resection.
Findings on imaging	<p>CT:</p> <ul style="list-style-type: none"> • Associated uterine leiomyomas • Hypoattenuating intravascular filling defects <p>MRI:</p> <ul style="list-style-type: none"> • Tubular intravascular mass with “sausage-like appearance” • Intraluminal tumor growth from uterine veins • Heterogeneous enhancement after intravenous gadolinium <p>Echocardiogram:</p> <ul style="list-style-type: none"> • Elongated mobile mass extending into IVC, right atrium, or right ventricle • Vascularized thrombi within the pelvic veins and IVC

Table 1: Summary table of intravenous leiomyomatosis.

	CT	MRI	Echocardiogram
Intra-vascular leiomyomatosis	<ul style="list-style-type: none"> • Associated uterine leiomyomas • Hypoattenuating intravascular filling defects 	<ul style="list-style-type: none"> • Tubular intravascular mass with “sausage-like appearance” • Intraluminal tumor growth from uterine veins • Heterogeneous enhancement after intravenous gadolinium 	<ul style="list-style-type: none"> • Elongated mobile mass extending into IVC, right atrium, or right ventricle • Vascularized thrombi within the pelvic veins and IVC
Deep vein thrombosis	<ul style="list-style-type: none"> • Hypoattenuating thrombus with no evidence of internal enhancement on contrast CT 	<ul style="list-style-type: none"> • Lack of enhancement following administration of gadolinium based contrast 	<ul style="list-style-type: none"> • Rare involvement of thrombus into major veins such as IVC • Extremely unlikely to extend to heart

Table 2: Differential diagnosis table for intravenous leiomyomatosis.

ABBREVIATIONS

ALT: Alanine aminotransferase
 AST: Aspartate aminotransferase
 CT: Computed tomography
 IVL: Intravenous leiomyomatosis
 MR: Magnetic resonance

KEYWORDS

Intravenous leiomyomatosis; leiomyoma; intravascular; intracardiac; inferior vena cava; computed tomography; magnetic resonance

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