Surgical management of intravenous leiomyomatosis with intracardiac extension

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ABSTRACT

Intravenous leiomyomatosis is a rare smooth muscle tumor that is associated with uterine leiomyomas. Intravenous leiomyomatosis often presents with nonspecific abdominal and cardiac symptoms, making the diagnosis difficult. We present a comprehensive review of a case of a 52-year-old woman with intravenous leiomyomatosis with intracardiac extension, who was successfully treated with complete surgical resection. (J Vasc Surg Cases Innov Tech 2023;9:101302.)

Keywords: Intracardiac extension; Intravenous leiomyomatosis; Uterine leiomyomas

Intravenous leiomyomatosis (IVL) is a rare smooth muscle tumor that is associated with uterine leiomyomas.[1-3] According to the literature, IVL occurs in 0.1% of cases of uterine leiomyomas, and IVL with intracardiac extension (ICE) occurs in 10% to 40% of IVL cases.[4-8] Although the disease is histologically benign, its aggressive nature with rapid growth and the ability to invade tissue can reflect malignancy clinically.[1,3] IVL has four clinical stages of progression: stage I, localization in the uterus; stage II, involvement of the pelvic cavity and iliac veins; stage III, involvement of the inferior vena cava (IVC) or renal or liver veins; stage IV, involvement of the right heart system.[3,9] The presenting symptoms of IVL are often nonspecific and often mimic other diseases, making the diagnosis difficult.1,2,3 The manifestations are related to the localization and degree of tumor involvement in the venous system. Tumors confined to the pelvis will present with pelvic and abdominal symptoms, such as pain, nausea, and vomiting, and tumors with ICE will present with cardiac symptoms, such as dyspnea, syncope, and edema.[1,7,10] Early detection and surgical resection are crucial to effective management because the late findings of IVL with ICE include saddle embolus, heart failure, and death.[10,11] Given its rarity, we present the case of a 52-year-old woman with IVL and ICE who was successfully treated with surgical resection.

CASE REPORT

A 52-year-old woman with a history of uterine leiomyomas presented to the emergency department in May 2022 with a 2-day history of severe shortness of breath and associated chest tightness and a 3-week history of fatigue. Computed tomography (CT) of the chest revealed extensive saddle pulmonary embolism (PE) with clot extension into the right atrium and IVC (Fig 1). A study for deep vein thrombosis (DVT) was performed via ultrasound; however, no evidence of distal DVT was found, and further thrombophilia workup was negative. At the time of presentation, despite the concerning imaging findings, she remained hemodynamically stable and was, therefore, initially treated for an unprovoked PE with anticoagulation and admitted to the intensive care unit (ICU) for close monitoring. While in the ICU, further echocardiography and abdominal pelvic imaging studies showed a long segment of thrombus extending continuously from the level of the right external iliac vein to the right ventricular outflow tract with evidence of enhancement of some of the clot present within the intrahepatic and suprahepatic IVC. These findings raised suspicion for a neoplastic process. Follow-up magnetic resonance imaging of the pelvis demonstrated a multifibroid uterus with no radiologic evidence of leiomyosarcoma.

During treatment of her PE, her respiratory status became compromised shortly after admission. Given the significant burden of thrombus and developing vital changes, she underwent urgent intracardiac tumor resection and pulmonary embolectomy. The right atrium was opened to extract mass from the IVC; however, complete resection of the mass was not possible. Pathologic examination of the mass reported IVL with no atypia.

Postoperative CT of the body showed residual tumor in the distal IVC and right common iliac vein. At this point, vascular surgery and gynecologic oncology were consulted. A joint decision was made to delay future operations until the patient had recovered from cardiac surgery with a tentative timeframe of 6 weeks.

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The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

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https://doi.org/10.1016/j.jvscit.2023.101302

Journal of Vascular Surgery Cases, Innovations and Techniques
December 2023

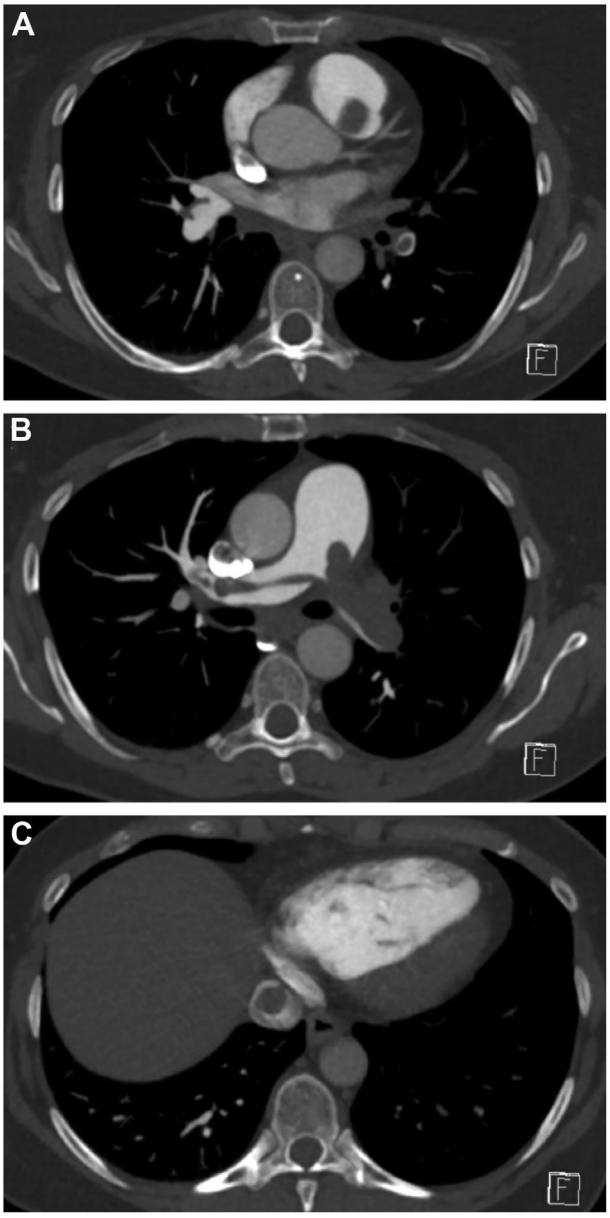

Fig 1. Axial computed tomography (CT) of the chest showing extensive saddle pulmonary embolism (PE) with clot extension from the right atrium to inferior vena cava (IVC).

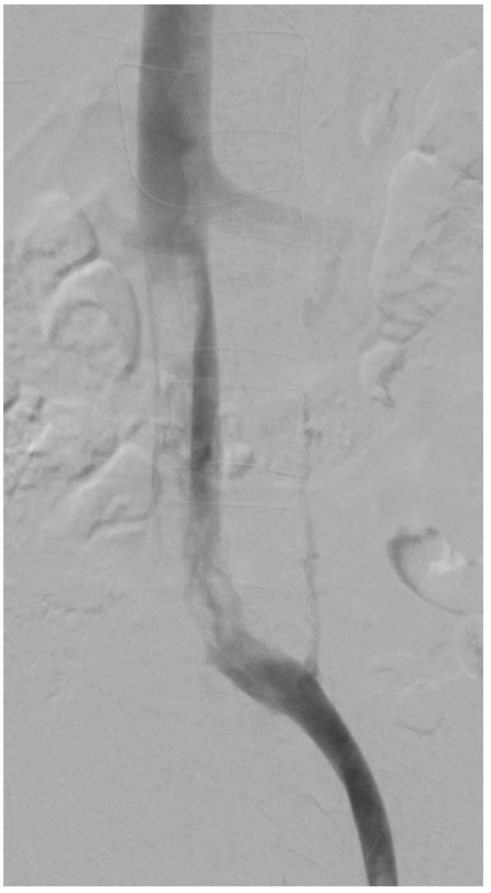

Fig 2. Intraoperative venography displaying a large filling defect of the inferior vena cava (IVC) and no flow through the right iliac vein system.

In the interim, the patient continued taking a direct oral anticoagulant after discharge. Perioperatively, an unfractionated heparin infusion was started.

A collaborative open resection was conducted by vascular and gynecologic surgery. On the operating table, venography was first completed to assess the clot burden, which showed a large filling defect of the IVC and no flow through the right iliac vein system (Fig 2). Intravascular ultrasound was also used to confirm the location of the tumor with respect to renal and hepatic veins. A midline laparotomy was performed, and the right colon and duodenum were mobilized medially to expose the infrarenal IVC. The right gonadal vein was divided at the origin of the right renal vein and resected as part of the specimen. The internal iliac veins were exposed down to the third order branches, and the right external iliac vein and the left common iliac vein were dissected for eventual control. We chose to open the IVC from the iliac vein to the level of the renal veins to completely remove the tumor under direct visualization and avoid tumor embolus. On entry into the IVC, a large amount of tumor was encountered; however, the IVC was noted to be completely normal. We were able to easily remove the tumor essentially in one long cord with continuous traction, because it was not adherent to the vessel wall (Fig 3). The cava above the level of the renal veins appeared normal with no evidence of tumor or thrombus. The dissection was then continued all the way down to the iliac bifurcation, allowing for removal of all remaining tumor in deeper branches of the internal iliac veins. We elected to repair the IVC with a bovine pericardial patch, because we did not see a benefit in removing and reconstructing an otherwise normal IVC. Once we believed the bulk of the tumor thrombus was safely removed, gynecologic surgery performed total abdominal hysterectomy and bilateral salpingo-oophorectomy. She had a history of endometriosis and evidence of stage IV endometriosis, with both ovaries densely adherent to the posterior uterus and pelvic sidewall noted intraoperatively. This operation lasted 5 hours, with an

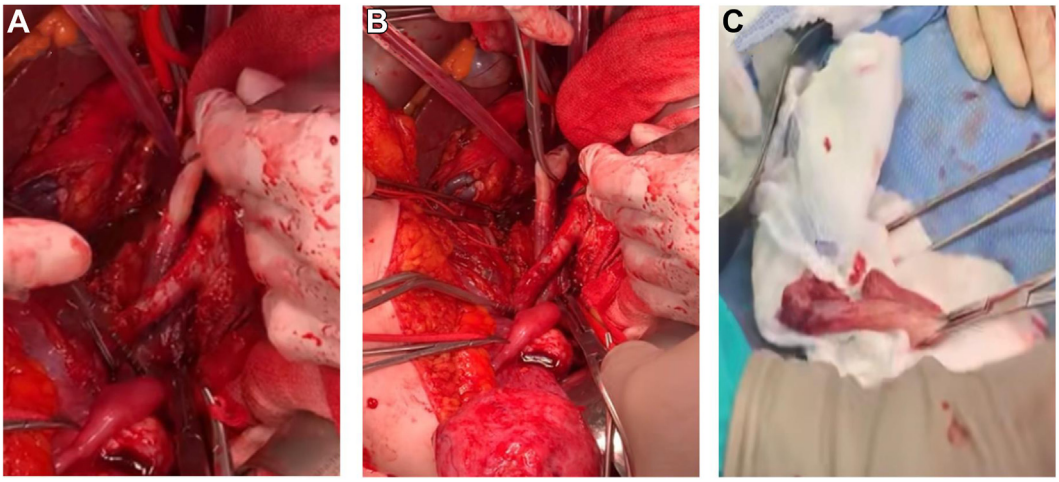

Fig 3. A, A large tumor was encountered on entry into the inferior vena cava (IVC). **B,** It was removed in one long cord with continuous traction. **C,** Photograph showing resected segment visible on a sponge.

estimated blood loss of 800 mL. The patient's hospital admission lasted 13 days, and her postoperative course was only complicated by right obturator neuropathy. This was confirmed by electromyography and thought to be self-limiting; therefore, no further management was required. The final pathology examination again reported IVL and uterine leiomyomas with no atypia. Thus, she was recommended clinical follow-up with no adjuvant therapy. Postoperatively, the patient was transitioned to low-molecular-weight heparin and ultimately discharged home with apixaban as guided by the consulting hematology service, which will follow-up the patient for complex venous disease. She was seen at 1 month after surgery by vascular surgery and was noted to have had an uncomplicated recovery with no changes to the management required. The patient provided written informed consent for the report of her case details and imaging studies.

DISCUSSION

Our case demonstrates successful management of stage IV IVL, which has the highest risk of morbidity, mortality, and surgical complexity.[1,3,11] Nonspecific cardiac findings such as fatigue and dyspnea, in addition to a diagnosis of PE in a woman, especially one with a history of uterine leiomyomas, should raise suspicion for IVL warranting surgical resection for management.[1,3,6,11]

The operation can be performed as a single or multistage procedure with comparable postoperative outcomes.[1,12] For our patient, a multistage approach for resection of tumor thrombus was followed predominantly because of the clinical presentation. Given this patient's acute decline in the ICU, urgent cardiac and pulmonary thrombus removal was necessary, and additional major abdominal and pelvic operative interventions would likely not have been safe simultaneously. At the time of the abdominal and pelvic components of her treatment, the associated significant endometriosis made dissection quite challenging during both the IVC dissection and the hysterectomy. Despite the surgical complexity, we were able to achieve complete resection of the tumor with minimal postoperative complications, including prolonged fatigue and right leg adduction weakness. This was found to be consistent with an intraoperative stretch injury to the obturator nerve with an expected recovery in 12 to 16 weeks.

The diagnosis of IVL is best made using CT or transthoracic echocardiography looking for ICE. CT is crucial for preoperative planning because it provides important information about the tumor burden and range and path of the lesion and can additionally assess the gynecologic structures in great detail.[1,9] Although diagnostic imaging is necessary for both diagnosis and planning, it is important to confirm the diagnosis histologically.[6] Patients with confirmed IVL with ICE have a high risk of venous thrombosis and require both preoperative and postoperative anticoagulation therapy for ≥3 months.[3]

Although IVL is not considered a malignant pathology, it shares molecular cytogenetic characteristics with both uterine leiomyomas and leiomyosarcoma, potentially explaining its partial malignant behavior.[13] Although malignant transformation has not been described in the literature, cases of pulmonary metastases have been reported.[14] Given its invasive nature, IVL poses a risk of fatal obstruction of the cardiac and/or pulmonary vasculature. Therefore, complete resection of the tumor provides patients with the lowest risk of morbidity, mortality, and recurrence.

The risk factors for recurrence include large tumor (≥7 cm), involvement of the broad ligament, and incomplete surgical resection.[3,15] In cases in which complete resection cannot be achieved, the risk of recurrence is ≤30%, necessitating postoperative medical management and longitudinal follow-up.[1,3,15,16] IVL is estrogen sensitive; therefore, various hormonal treatments have been trialed as an adjunct to surgery

to reduce the risk of recurrence; however, currently, no standard therapy or guidelines have been established.[10,12] Furthermore, the evidence is insufficient to support neoadjuvant hormonal therapy for patients for whom complete surgical resection is considered to be too high risk initially.[1] Further investigation is required to determine the role of adjuvant hormonal therapy in IVL recurrence prevention vs the standard of care after hysterectomy and bilateral salpingo-oophorectomy.

CONCLUSIONS

IVL is a rare association of uterine leiomyomas, and IVL with ICE is even rarer. Therefore, IVL should be considered when women, especially those with a history of uterine leiomyomas, present with nonspecific abdominal or cardiovascular symptoms. Once identified, IVL can be successfully managed with complete surgical resection. Adjuvant hormonal treatment can be considered owing to the risk of recurrence.

DISCLOSURES

None.

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Submitted Jun 16, 2023; accepted Aug 3, 2023.