

Post hysterectomy intravenous leiomyomatosis: multimodality imaging appearances

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INTRODUCTION

Intravenous leiomyomatosis (IVL) is extremely rare with just around 300 cases reported in literature.¹ With its unusual growth pattern and clinically aggressive behavior, this benign tumor can masquerade as a malignancy and present a diagnostic challenge. Concurrent uterine fibroids or recent hysterectomy for the same often provides a hint towards the diagnosis.² The median interval between hysterectomy and diagnosis of IVL is approximately 4 years.³ We present a case of IVL detected 10-years post total hysterectomy, bilateral salpingo-oophorectomy (THBSO) and parametrectomy with emphasis on multimodality imaging with multiphase CT, MRI and digital subtraction angiography (DSA).

CASE REPORT

Our patient is a 54 year old woman with THBSO 10 years ago for uterine and broad ligament leiomyoma, and ovarian endometriosis. She presented in January 2014 for outpatient CT urography to investigate her complaint of an episode of gross hematuria. CT incidentally detected a pelvic mass with right iliac vein and IVC thrombus (Fig. 1), for which she was transferred to our institution for further management. Serum disseminated intravascular coagulation (DIC) screening (coagulation profile, D-dimer and fibrinogen levels), thrombophilia screening and tumor markers were all unremarkable. CT thorax and MRI of the pelvis (Fig. 2) were also done as part of the diagnostic work-up. A diagnosis of recurrent pelvic fibroids and IVL was made and confirmed by trans-gluteal core biopsy of the pelvic mass. Unfortunately, the biopsy was complicated by iatrogenic pelvic hematoma. This was diagnosed with CT angiography and confirmed at digital subtraction angiography (Fig. 2). The bleeding branch from the anterior division of the right internal iliac artery was embolized with metallic coils.

As the patient was clinically stable, therapeutic dose of low-molecular-weight heparin was started, and elective surgery performed. The tumor was free of the vessel wall along its entire course from the iliac vein to the right atrium. Surgical pathology concurred with the pre-operative radiological diagnosis.

DISCUSSION

IVL is characterized by a proliferation of benign smooth muscle within veins outside the confines of a leiomyoma, or even when no leiomyoma is present, with a potential to contiguously propagate through the venous system into the right atrium, ventricle and further.⁴ Just like the other protean manifestations of uterine leiomyomatosis such as disseminated peritoneal leiomyomatosis, benign metastasizing leiomyoma, parasitic leiomyoma, and retroperitoneal leiomyomatosis, IVL is extremely rare with most of the literature based on isolated case reports or short case series. The origin of IVL is debatable. According to Knauer's theory, the tumor arises from smooth muscle cells in the vessel wall, while Sitzenfray's theory states that the uterine leiomyoma itself proliferates to subsequently invade the venous channels.¹

IVL remains asymptomatic until late and often diagnosed incidentally on imaging.¹ At the time of diagnosis, extra-uterine involvement is seen in 30-80 % of cases, of which cardiac involvement is present in 10-30%.⁵ Even advanced IVL can remain quiescent, until venous obstruction or tricuspid valve impairment occurs, resulting in exertional dyspnea, arrhythmias, pedal edema, syncopal episodes, deep venous thrombosis and pulmonary embolism. Rarely, IVL presents with Budd-Chiari Syndrome or sudden death.^{3,5} Due to hormone dependence, it generally occurs in premenopausal middle-aged women, with a median age of 45 years.³ For the same reason, one of the strategies of preventing IVL is to perform THBSO besides parametrial excision with clear margins. Our patient had such a radical primary surgery, yet developed pelvic recurrence of leiomyoma and IVL. This prompted us to do a biopsy to exclude malignancy. Moreover, the condition was presenting after an unusually long interval from the previous surgery.

We got a serendipitous opportunity to image the tumor with multiphase CT, MRI and DSA. Such descriptions of IVL with multimodality correlation are scarce in literature. Our search of medical literature in English language did not yield any previous report on the CT appearance of IVL in four different phases or on its angiographic appearance. Besides the tissue characterization, the major value of CT and MRI is in depicting the extent of the lesion, relationship with the vessel wall and neighboring viscera.

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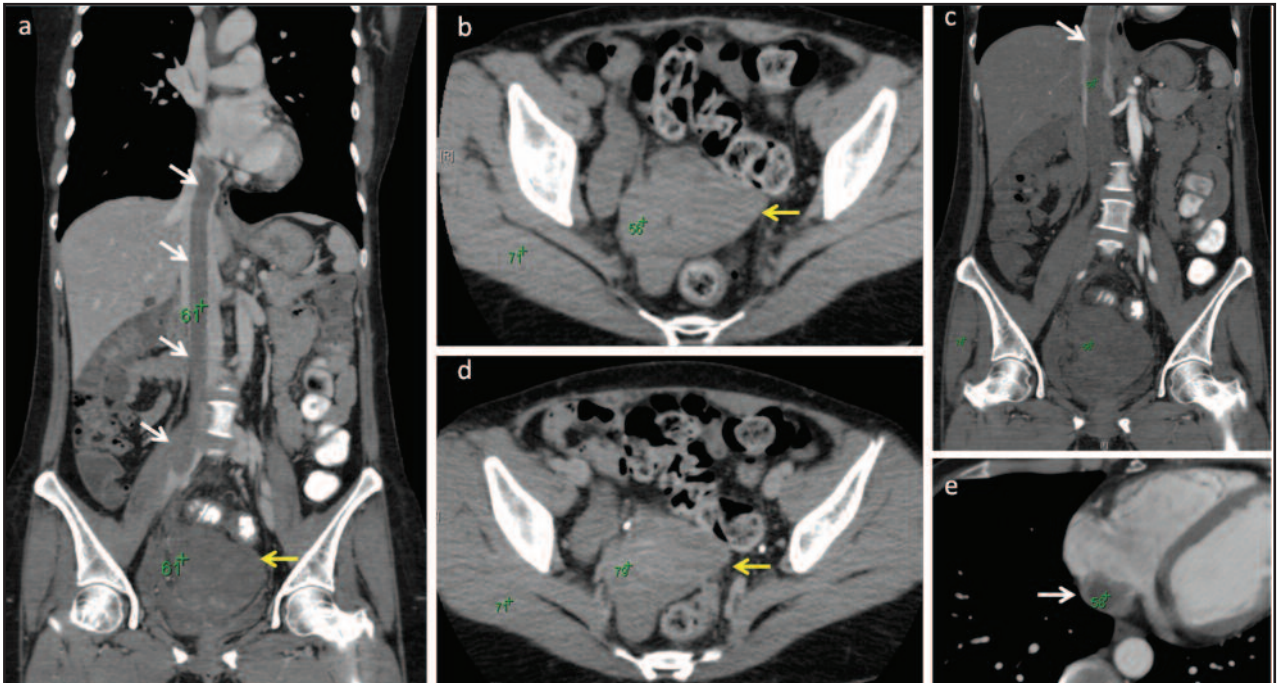


Fig. 1: The coronal venous phase CT scan (a) shows almost entire extent of the disease including the pelvic mass and intravenous leiomyomatosis growing along the right iliac vein and IVC into the right atrium. The column of contrast between the intravenous mass and the vessel wall suggests that it is free of the vascular endothelial surface. The pelvic and intravascular masses are slightly hypodense to the skeletal muscle. A similar attenuation pattern is seen in the non contrast axial image (b) and coronal arterial phase image (c). There is mild degree of delayed enhancement seen in the delayed post contrast CT scan (d). The arterial phase axial CT scan of the lower thorax shows intra-atrial component of the tumor (yellow arrow- recurrent pelvic fibroids; white arrow- intravenous leiomyomatosis; tissue attenuation in Hounsfield units written in green colored numerals).

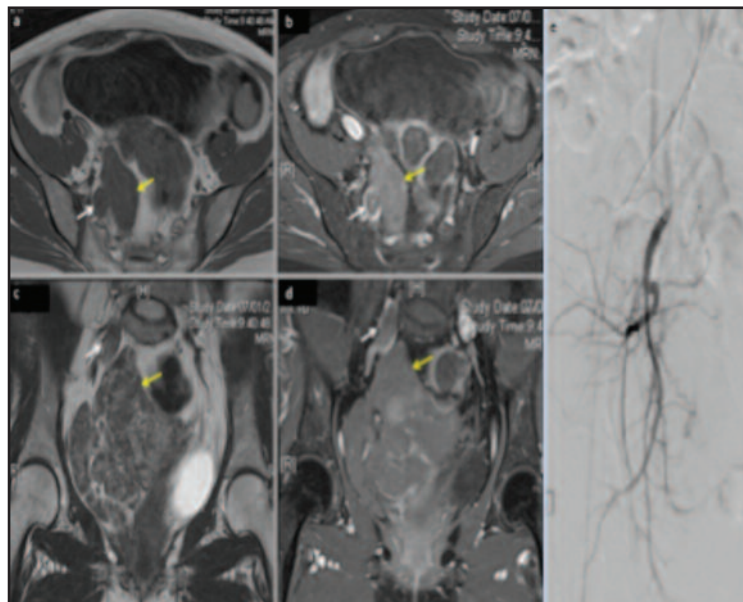


Fig. 2: The axial pre contrast T1 weighted (a) and post contrast fat saturated T1 weighted (b) images at the same level shows the lobulated pelvic mass isointense to the skeletal muscles squeezing into a branch of internal iliac vein. The intravascular component is better appreciated on the Fig 2b where almost 2/3 rd of the tumor thrombus is surrounded by contrast and a relatively narrow stalk connects it to the rest of the pelvic tumor. The coronal T2 weighted (c) image shows the marked internal heterogeneity and the cranio-caudal extent of the pelvic tumor as well as the intravenous tumor extending longitudinally. In the post contrast T1 weighted (d) image along a similar plane as Fig 2c the intravascular component is better demonstrated. The pelvic and intravascular tumors have similar signal intensity on all the sequences with mild post contrast enhancement. (yellow arrow- recurrent pelvic fibroids; white arrow- intravenous leiomyomatosis. The digital subtraction angiogram (e) of the right internal iliac artery shows mild capillary-phase blush of the pelvic tumor. No hypertrophic/ tortuous feeding artery, obvious hypervascularity, intra-tumoral shunts or prominent draining vein is detected.

The differential diagnoses for IVL include primary leiomyosarcoma, malignant thrombus (eg. from metastatic renal cell carcinoma), endometrial stromal sarcoma and bland thrombus. The intra-atrial component often tends to be misdiagnosed as a right atrial myxoma; however they are unlikely to extend down into the pelvic veins and generally show marked hyper-intensity on T2WI. On imaging, primary leiomyosarcoma cannot be differentiated from IVL unless the disease has progressed to an advanced stage with visible tumor invasion of the abdominal viscera. Sometimes, in early disease, the mural origin of a primary caval leiomyosarcoma may be demonstrable, unlike IVL which is separate from the vessel wall. A biopsy is often required to exclude vascular invasion by pelvic sarcoma. A malignant thrombus is unlikely in the absence of a primary abdominal malignancy. A bland tumor is often easily excluded by its lack of enhancement.

Surgery is the treatment of choice for IVL, with successful clinical management based on THBSO, and removal of the intra-venous tumour.² There may be a potential role for adjuvant anti-estrogen therapy with tamoxifen, though it's clinical efficacy is yet to be established.¹ The prognosis is excellent if complete surgical resection is achieved. There is no consensus on the optimal timing and technique for follow-up surveillance imaging. A baseline CT may be done at 6 months post surgery, and subsequently every 2 to 5 years depending on surgical excision and extent of disease.¹

CONCLUSION

In conclusion, diagnosis of IVL requires the radiologist to be aware of this entity and pro-actively seek for evidence, both radiological and clinical. Our report also provides insight into certain multimodality imaging features of IVL that have seldom been described in literature.

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