

## Vascular–Interventional

# Tips and tricks: Intravenous leiomyomatosis

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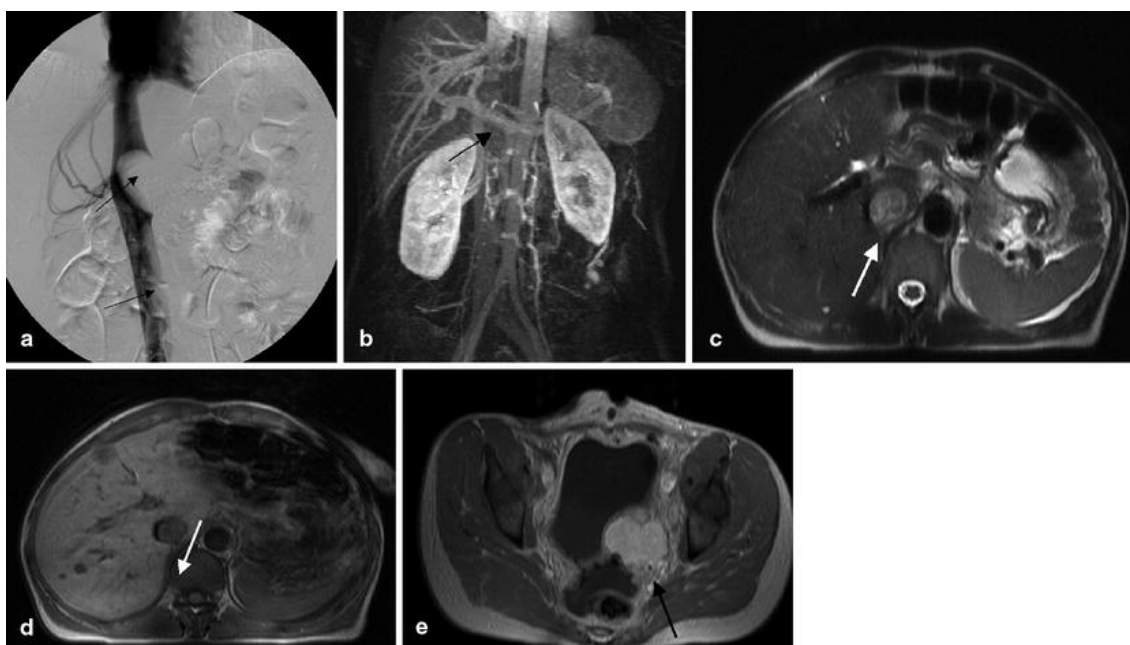
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## Case report

A 48-year-old woman had developed bulk-related symptoms of recurrent leiomyoma 2 years after hysterectomy due to uterine leiomyoma. No other symptoms were observed. After US examination and other investigations, laparotomy was planned. During surgery, severe bleeding started, and tumor extension in the inferior vena cava (IVC) was found. After right-sided oophorectomy and controlling bleeding, the surgery was stopped and the patient was referred to the radiology department for further investigations in order to rule out intravenous leiomyomatosis (IVL).

Phlebography of abdominal and pelvic veins showed a partially obstructing tumor thrombus adherent to the left lateral wall of the vena cava inferior, extending up to the level of T12/L1 (Fig. 1a). Contrast-enhanced MR angiography revealed infiltration of the venous channel (left vena ovarica, left renal vein, and IVC) by tumor and multiple homogenous masses in the small pelvis. At the level of T12/L1 the tumor thrombus was 40×30 mm in size and almost totally occluding IVC (Fig. 1b). The results of contrast-enhanced MRA were comparable to the results of the conventional phlebography. The tumor was isointense to muscles on T1-weighted images

and hyperintense on T2-weighted images (Fig. 1c,d). The tumor showed high vascularization with increased signal intensity after intravenous administration of contrast material (Fig. 1e). On the basis of characteristic MR features, the diagnosis of intravenous leiomyomatosis was made. A single-stage surgery with extirpation of small pelvic masses, left-sided oophorectomy, resection of the left vena ovarica, partial resection of the left renal vein, and thrombectomy of the tumor thrombus (as the thrombus was found to be freely moveable in the inferior vena cava), followed by reconstruction between vena cava inferior and left renal vein with prosthesis, was safely performed. Histology of the resected specimen showed a leiomyoma without malignancy.



**Fig. 1** **a** Phlebography demonstrates extension of partially obstructing tumor (*arrow*) in inferior vena cava (IVC). **b** Magnetic resonance angiography (venous phase) demonstrates tumor thrombus (*arrow*) of the IVC. **c** T2-weighted MRI shows tumor thrombus (*arrow*) in IVC which is hyperintense. **d** T1-weighted MRI shows tumor thrombus (*arrow*) in IVC isointense to muscles. **e** Contrast-enhanced MRI shows hyperintense tumor in the small pelvis compressing the urinary bladder(*arrow*)

## Discussion

Intravenous leiomyomatosis is a rare, benign, smooth muscle tumor, seen exclusively in white women with previous history of hysterectomy due to uterine leiomyoma. The IVL originates either from contiguous proliferation of benign leiomyoma into the veins or intravascular intimal smooth muscle proliferation. Cases of IVL originating from skin vessels and IVC have been reported. Involvement of the pelvic veins, inferior vena cava, adrenal and renal veins, pulmonary artery,

and cases of pulmonary and other distant (brain, bone, cardiac and lymph nodes) metastasis have been reported.

Patients with IVL may present with the symptoms of uterine leiomyoma, i.e. pelvic pain and vaginal bleeding, respiratory symptoms (dyspnea, cough) in case of pulmonary metastasis, swelling of legs in case of inferior vena cava occlusion by the tumor, and symptoms of cardiac failure due to the tumor extension in the right cardiac cavities. Surgical resection of affected veins, hysterectomy, and bilateral oophorectomy due to estrogen dependence of the tumor is the treatment of choice for IVL, and the possibility of recurrence is 30%. In order to be sure that the whole tumor is resected, Evans has recommended a 3- to 6-month follow-up. A diagnosis of IVL is easy, when the connection between intravenous mass and uterus is visualized; however, in patients after hysterectomy, IVL can only be correctly diagnosed if the pathological diagnosis of IVL is known. The signal intensity of the IVL is isointense to the muscles in T1-weighted images and hyperintense in T2-weighted images. After intravenous administration of contrast material, the signal intensity of the IVL increases. This enhancement may be due to the different amount of smooth muscle cells and fibrous-tissue-containing hyalinized vessels. The differential diagnosis of IVL include tumor thrombosis of Wilms tumor, adrenal tumor, leiomyosarcoma, primary leiomyoma of IVC, lymphoma and soft tissue sarcoma, retroperitoneal fibrosis, and metastasis. The MRT is a useful tool to assure the diagnosis of intravenous leiomyomatosis.

In conclusion, the present case demonstrates the high potential of MRT of the abdomen in the evaluation of patients for pelvic symptoms.

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