

Magnetic resonance venogram of intravenous leiomyomatosis

Case summary

A 47-year-old woman presented with menorrhagia and an enlarged uterus. Uterine fibroids were diagnosed on ultrasound. A few months later, the patient complained of left lower limb swelling. Deep venous thrombosis was diagnosed, anticoagulation therapy was prescribed, and total hysterectomy and bilateral salpingo-oophorectomy were subsequently performed. Pathological examination of the surgical specimen demonstrated intravenous leiomyomatosis. Postoperative computed tomography (CT) scans of the thorax, abdomen, and pelvis were performed to look for evidence of lung secondaries and intravenous extensions of the lesion. Delayed CT after contrast injection showed suspicious heterogeneous enhancement of the infrarenal inferior vena cava (IVC) and left common iliac vein. A magnetic resonance (MR) imaging scan and an MR venogram were then performed to assess the suspected intravenous lesion. On a two-dimensional time-of-flight (TOF) sequence, an abnormal filling defect was revealed in the infrarenal IVC (Fig 1), left common and internal iliac veins (Fig 2). A direct contrast-enhanced MR venogram, using contrast injection via the vascular access at the left foot, revealed the intravenous filling defect and the obstructive effect of the lesion's collateral formation (Fig 3). The enhancement of the lesion helped to make the diagnosis of an intravenous tumour lesion rather than venous thrombosis. A second operation was performed by a vascular surgeon to excise the tumour from the venous system (Fig 4).

Discussion

Intravenous leiomyomatosis is a rare disease; about 200 cases have been reported in literature,¹ all in females of reproductive age. It is an intravenous proliferation of a histologically benign smooth muscle tumour. However, it is not a benign disease: it can progress and extend along the venous system which has led to intracardiac involvement in a few cases.^{1,2} Moreover, lung metastasis has been reported as a complication of intravenous leiomyomatosis.³ While pathogenesis of the tumour remains controversial, there are two main theories regarding its origin: intravenous extension of the uterine leiomyoma, or direct tumourigenesis from the wall

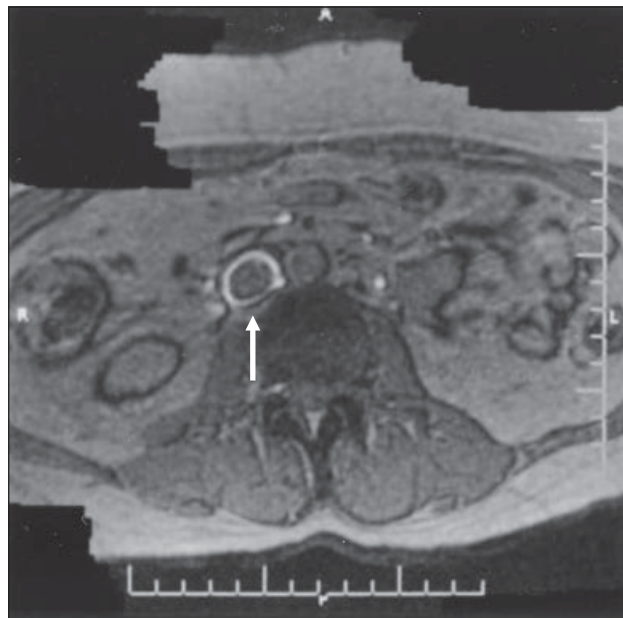


Fig 1. Axial section of time-of-flight (TOF) image at the abdomen demonstrated the low signal filling defect inside the inferior vena cava (arrow). On TOF sequence, flowing blood is of high signal

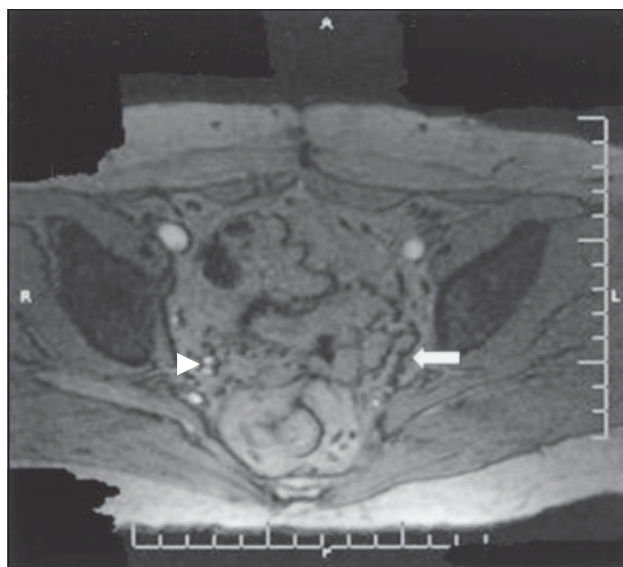


Fig 2. Axial section of time-of-flight image at the pelvis did not demonstrate high signal at the distended left internal iliac vein and its branches (arrow). On comparison, the right internal iliac veins are well demonstrated as high-signal areas (arrowhead) on right side of pelvis

of the venous system. Most cases have been associated with uterine leiomyoma, and the most common

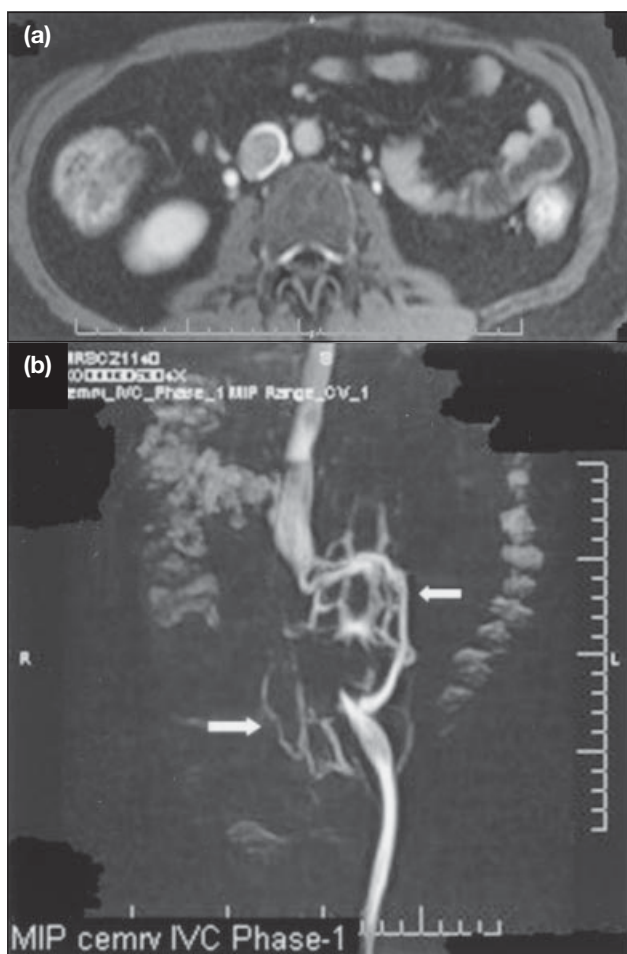


Fig 3. (a) Axial section of post-contrast T1 image of the abdomen showed the enhancing tumour lesion within the inferior vena cava (IVC) surrounded by enhanced blood; (b) Maximum intensity projection of contrast-enhanced magnetic resonance venogram illustrated the blockage of left iliac veins and part of IVC with collateral vessel formation (arrows)

presentation was uterine enlargement with leiomyoma. The diagnosis had not been suspected in most of the patients before hysterectomy; some patients presented few years later and the diagnosis was probably missed at the time of operation.

Various imaging examinations are available for the detection and diagnosis of intravenous leiomyomatosis. Conventional venogram is an invasive procedure involving catheter cannulation and irradiation. Ultrasound is operator-dependent, bowel gas obscuration is always a problem, and assessment of deep structures in abdomen and pelvis is not always optimal. On the other hand, echocardiogram is helpful in cases with intracardiac extension. Good images can be obtained with direct contrast-enhanced CT venograms, but this involves radiation and there is always the potential problem of contrast reaction and complications.



Fig 4. The surgical specimen of the excised intravenous leiomyoma, which is tubular in shape conforming to the lumen of the vessel

Magnetic resonance imaging is particularly helpful in assessing patients with suspected intravascular lesions due to its multiplanar capabilities, better soft tissue-contrast resolution, and development of much faster imaging sequences, as well as its unique ability to assess blood flow without contrast injection.⁴ Magnetic resonance imaging can provide important information unavailable with other imaging examinations. It is also a safer examination, as the gadolinium chelate is a much safer contrast agent compared with the water-soluble contrast used in CT scanning, and there is no irradiation to the patient. The TOF technique, based on flow-related enhancement phenomena, assesses the extent of the intraluminal lesion without an injection of contrast. Internal iliac veins usually do not show up on direct contrast-enhanced venograms, but they can be readily assessed with TOF techniques. While TOF techniques provide good evaluations of intraluminal lesions in the venous system, contrast injection is required to make the important differentiation of tumour from thrombus. The imaging appearance of intravenous leiomyomatosis is non-specific, but the major role of an imaging examination is to define the extent of the lesion. The treatment of intravenous leiomyomatosis should be complete removal by surgery.⁵ Therefore, accurate preoperative assessment of the extent of the lesion is critical. Anti-oestrogens like tamoxifen have been used to treat the disease, but their efficacy remains uncertain.

Conclusion

Intravenous leiomyomatosis is a rare disease. Although it is caused by pathologically benign lesion, it is not a benign disease. Complete surgical resection of the

tumour is the treatment of choice. Magnetic resonance imaging and MR venogram are well suited to assess the extent of the lesion, which is critical to surgical planning.

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