CASE REPORT

Intravenous leiomyomatosis with intracardiac extension: an unusual presentation of uterine leiomyoma and evaluation with 256-slice dual-source multidetector CT and cardiac MRI

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SUMMARY

We report a case of a 40-year-old woman who presented with breathlessness, palpitations and vague pelvic pain for 6 months. The dyspnoea was non-exertional and episodic. She was a non-diabetic, non-hypertensive and had no history of prior surgeries. ECG and chest radiograph were normal. Cardiac MRI and 256-slice dual-source multidetector CT evaluation revealed the presence of a uterine and left adnexal mass with intravenous extension into the inferior vena cava via the gonadal veins, and further cephalad extension into the right atrium and ventricle. The patient was referred to the cardiothoracic surgery department and underwent two-stage surgical procedures. The first stage consisted of thoracotomy and tumour removal from the right atrium and inferior vena cava. In the second stage, hysterectomy and bilateral salpingo-oophorectomy was performed. The histological analysis of the excised specimen was consistent with an intravascular leiomyoma.

these patients. Different imaging findings and differential diagnosis are discussed in this report.

CASE PRESENTATION

We report a case of a 40-year-old woman who presented with symptoms of breathlessness, palpitations and vague pelvic pain for 6 months. The dyspnoea was non-exertional and episodic. She was a non-diabetic, non-hypertensive and had no history of prior surgeries.

INVESTIGATIONS

Ultrasonography of the abdomen revealed a bulky uterus. Colour Doppler evaluation showed increased low-resistance uterine vascularity. A well-circumscribed, lobulated, hypoechoic lesion was seen in the uterus and left adnexal region ([figure 1](#)). This lesion showed intravascular extension into the left gonadal vein and the inferior vena cava (IVC). Further imaging studies were performed to characterise and gauge the extent of the pelvic soft tissue lesion.

On two-dimensional echocardiography, tubular serpiginous hypoechoic lesions were noted in the right atrium and right ventricle, with intralesional vascularity. CT study of the abdomen and chest was performed using 256-slice dual-source CT for further evaluation. The study demonstrated a lobulated, strongly enhancing soft tissue lesion (approximately 5.5×4.5×5.0 cm in size) in the left adnexal region, contiguous with the adjacent uterine wall ([figure 2](#)). Multiple intratumoural tortuous vessels were noted. The mass lesion along with the dilated intratumoural vessels extended

BACKGROUND

Uterine leiomyoma is the most common uterine tumour. Rarely, unusual growth patterns such as intravenous leiomyomatosis may be seen. Intracardiac extension of these tumours is even rarer. The diagnosis of such cases is challenging due to the non-specific clinical presentation. This case highlights the importance of imaging studies—such as (multidetector CT) and cardiac MRI—in the diagnosis of such an unusual growth pattern; the studies aid in the appropriate management of

Figure 1 Ultrasound images in transverse (A) and oblique (B) planes showing increased uterine vascularity and a hypoechoic left adnexal mass extending into the left ovarian vein.

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Figure 2 Contrast-enhanced CT images of the pelvis showing a well-circumscribed lobulated enhancing, exophytic mass arising from uterus and extending into the left adnexal region.

into the IVC via the left gonadal vein and left renal vein, and continued cephalad to extend into right atrium and right ventricle (figures 3–5).

Cardiac MRI was performed, which revealed a lobulated mass lesion of intermediate signal intensity on T1 and T2-weighted images, in the right atrium. Multiple flow voids

were noted within the lesion on T2-weighted images, representing intralesional tortuous vessels. The lesion was seen to prolapse (through the tricuspid valve) from the right atrium into the right ventricle during diastole (figure 6).

A diagnosis of intravenous leiomyomatosis with intracardiac extension was made and the patient was posted for surgical excision of the lesion.

DIFFERENTIAL DIAGNOSIS

The diagnosis includes intraluminal IVC lesions:

- ▶ Primary leiomyosarcoma of the IVC: cannot be differentiated from leiomyoma on the basis of imaging findings alone, unless it has progressed to an advanced stage with visible infiltration and invasion of the abdominal viscera.
- ▶ Tumour thrombus in the IVC (secondary IVC tumour): contiguity with an adjacent primary tumour is seen, for example, renal cell carcinoma and hepatocellular carcinoma.
- ▶ Bland thrombus in the IVC: the thrombus displays no enhancement on the postcontrast study.

TREATMENT

Our patient was referred to the cardiothoracic surgery department and underwent two-stage surgical procedures. The first stage consisted of thoracotomy and tumour removal from the

Figure 3 Coronal (maximum intensity projection) images showing extension of the uterine mass along with multiple tortuous tumoural vessels into the left ovarian vein (A and B) and inferior vena cava (C).

Figure 4 Axial contrast-enhanced CT images of the cardiac chambers showing a hypodense, heterogeneously enhancing mass lesion with large intratumoural vessels in the right atrium.

Figure 5 Volume rendered technique image showing extension of the uterine mass along with multiple tortuous tumoural vessels, into the left ovarian vein and inferior vena cava.

right atrium and IVC (figure 7). In the second stage, hysterectomy and bilateral salpingo-oophorectomy were performed.

OUTCOME AND FOLLOW-UP

Postoperative histopathological evaluation revealed interlacing bundles of spindle-shaped smooth muscle having elongated nuclei. For further confirmation immunohistochemistry was performed, which was positive for smooth muscle cell markers such as smooth muscle actin, vimentin and desmin. The histopathological features aforementioned were compatible with a diagnosis of intravenous leiomyoma. The patient has clinically improved and is on regular follow-up with echocardiography and ultrasonography of the abdomen and pelvis. Follow-up imaging with CT and echocardiography did not reveal any residual lesion in the pelvis or cardiac chambers.

DISCUSSION

The spectrum of uterine tumours extends from benign leiomyoma to endometrial carcinoma. Leiomyoma is the most common benign tumour of the uterus, and is usually confined to the uterine contour. Rarely, this tumour might show atypical growth patterns with extrauterine extension, including intravenous leiomyomatosis, disseminated peritoneal leiomyomatosis and benign metastasising leiomyoma.^{1 2} These unusual patterns make the diagnosis and subsequent management difficult. When present, a synchronous uterine leiomyoma is indicative of the diagnosis. Birch³ originally described intravenous leiomyomatosis in an autopsy case. Subsequently, Durk reported the first case of intravenous leiomyomatosis with intracardiac extension.⁴

Intravenous leiomyomatosis is a rare condition that is histologically benign but clinically aggressive; it is characterised by intravenous extension of the uterine leiomyoma, commonly into

Figure 6 Axial T1-weighted (A and B) and T2-weighted (C and D) images showing a lobulated intermediate signal intensity mass lesion in the right atrium and right ventricle.

Figure 7 Intraoperative photograph showing a tubular mass lesion in the right atrium (A), specimen morphology (B).

the gonadal vein.^{5–7} These smooth muscle tumours can extend into the IVC and even, rarely, into the right-sided cardiac chambers, and may cause symptoms of obstruction.

Ultrasonography usually shows a bulky uterus with a well-circumscribed, hypervascular, hypoechoic lesion in a submucosal, intramural or subserosal location, with or without extension into the broad ligament. A vascularised thrombus might be seen in the IVC and cardiac chambers. Extension into systemic veins, the IVC and, particularly, cardiac chambers, is well demonstrated with CT. Its multiplanar reconstruction capability helps delineate the entire extension of the tumour from the pelvis to

the heart. The continuity of the uterine mass into the gonadal veins can be evaluated clearly with CT or MRI.^{8 9}

Appearance of the lesion on MRI depends on the relative amount of smooth muscle cells and fibrous tissue. The typical appearance is low to intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted images.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Leiomyoma is the most common benign tumour of the uterus; however, in some rare cases, it may show extension into the venous system, this is known as intravenous leiomyomatosis.
- ▶ This rare growth pattern of intravenous leiomyomatosis requires careful evaluation of the entire venous system for caval or intracardiac extension. Contrast-enhanced CT and MRI is invaluable in assessing the entire extent of the lesion.
- ▶ Intravenous leiomyomatosis may be considered in middle-aged women with cardiac symptoms presenting with a right-sided cardiac mass along with a pelvic mass.
- ▶ Radiologists and clinicians must be aware of this rare entity to avoid misdiagnosis.

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