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CASE REPORT
Cardiac imaging

Case report: intravenous leiomyomatosis with intracaval and intracardiac growth

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Background Intravenous leiomyomatosis (IVL) is a rare, benign smooth muscle cell tumour that extends beyond the pelvis. These tumours grow within vascular channels and can progress to involve the heart and pulmonary vasculature.

Case Summary A 44-year-old female initially presented to her primary care physician for subacute bloating. In the weeks leading up to her presentation, she was in good health. On admission, computed tomography (CT) imaging of the abdomen and pelvis was notable for a mixed solid and cystic mass arising from the fundal myometrium with invasion into the inferior vena cava (IVC). Transthoracic echocardiogram (TTE) was notable for mobile mass in the right atrium originating from the IVC. The mass was further evaluated by cardiac magnetic resonance (CMR) imaging before a multidisciplinary, single-staged thoracoabdominal resection was performed. The procedure was well tolerated, and the entire mass was successfully removed without complication. Subsequently, pathological analysis of the resected tumour revealed benign smooth muscle cells, confirming the diagnosis of IVL.

Discussion Intravenous leiomyomatosis is a rare cause of right-sided cardiac tumours but should be considered in premenopausal females, even those with a prior history of hysterectomy. The clinical presentation of patients with IVL is varied and imaging including CMR, CT, and TTE to evaluate the tissue characteristics and source of the cardiac mass should be performed. Finally, while imaging revealing a freely mobile pelvic mass extending into the IVC and right heart chambers is strongly suggestive of IVL, definitive diagnosis requires pathological analysis of resected tissue.

Keywords IVL • Intravenous leiomyomatosis • Cardiac tumour • Intracaval tumour • Case report

ESC Curriculum 2.3 Cardiac magnetic resonance • 2.1 Imaging modalities • 6.8 Cardiac tumours • 7.5 Cardiac surgery

Learning points

- Intravenous leiomyomatosis (IVL) is a rare cause of right-sided cardiac tumours but should be considered in premenopausal females, even those with a prior history of hysterectomy.
- Imaging, including transthoracic echocardiogram, computed tomography, and cardiac magnetic resonance, revealing a freely mobile pelvic mass extending into the inferior vena cava is strongly suggestive of IVL, but definitive diagnosis requires histological analysis of tissue samples.
- Definitive management of IVL involves surgical resection; however, recurrence may occur in up to 30% of cases.

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Introduction

Intravenous leiomyomatosis (IVL) is a rare smooth muscle cell tumour that exclusively affects females.[1] Although histologically benign, IVL can be clinically aggressive, invading vascular channels and extending beyond the pelvis.[1] The diagnostic imaging features of these tumours remain poorly described.[2,3] Herein, we describe the features of IVL on echocardiogram, computed tomography (CT), and cardiac magnetic resonance (CMR) imaging from the case of a healthy female who initially presented for abdominal distention and was found to have a pelvic mass with intracaval and intracardiac involvement.

Timeline of clinical course

Day −7	Patient presents to her primary care physician for 4 weeks of abdominal bloating
Day −5	CT scan reveals a large pelvic mass
Day −3	TTE reveals a freely mobile mass in the right atrium with extension into the IVC concerning for rhabdomysarcoma vs. IVL
Day −1	Patient admitted to an outside hospital
Day 0	Patient transferred to Emory University Hospital for further evaluation and management
Day 1	CT scan with contrast of the chest, abdomen, and pelvis (*Figure 1*) reveals a large pelvic mass extending into the abdomen and IVC
Day 3	Cardiac MRI with contrast and velocity flow mapping (*Figure 2* and *Table 2*) is notable for a mass with tissue characteristics concerning for rhabdomysarcoma
Day 12	Patient undergoes a single-staged thoracoabdominal resection involving gynaecological oncology, urological oncology, surgical oncology, and cardiothoracic surgery (*Figure 3A* and *B*)
Day 19	Patient discharged home
Day 20	Pathology (*Figure 3C* and *D*) confirms the diagnosis of IVL
Day 30	Institutional tumour board recommends initiating an aromatase inhibitor to reduce the risk of recurrence
6 months	Surveillance CT scan of the chest, abdomen, and pelvis shows no evidence of recurrence

CT, computed tomography; IVC, inferior vena cava; IVL, intravenous leiomyomatosis; MRI, magnetic resonance imaging; TTE, transthoracic echocardiogram.

Case presentation

A 44-year-old woman, with no past medical history and no family history of cancer, presented to her primary care physician for several weeks of abdominal distension. She was evaluated by a CT scan of the abdomen, which was notable for a pelvic mass. Transthoracic echocardiogram (TTE) revealed a cardiac mass involving the right atrium (RA). She was subsequently transferred to Emory University Hospital for further management.

In the weeks leading up to her presentation, she was in good health and denied constitutional symptoms. She reported occasional palpitations but had no symptoms indicating hemodynamic disturbance, including pre-syncope, syncope, or dyspnoea. On admission, vitals were unremarkable and physical exam was only revealing for non-tender abdominal distension. No additional heart sounds or evidence of volume overload was appreciated. Initial laboratory studies for tumour markers (*Table 1*) were only notable for elevated levels of CA 125.

Given this patient's pelvic mass and concomitant mass in the RA, leiomyosarcoma with thrombus vs. direct tumour metastasis and IVL were considered most likely. Additional imaging studies were performed to investigate these differentials. Computed tomography imaging of the chest, abdomen, and pelvis with intravenous contrast (*Figure 1*) revealed a heterogeneous mixed solid and cystic mass arising from the fundal endometrium with a large cystic component extending into the epigastric region. Computed tomography imaging also revealed a filling defect extending from the right ovarian vein into the inferior vena cava (IVC), RA, and right ventricle (RV) (*Figure 1B*). The filling defect in the RA and RV observed on CT scan was further evaluated by TTE, which demonstrated a mobile, 3.0 cm×3.2 cm mass extending from the IVC into the RA, with a serpentine projection across the tricuspid valve into the RV (Supplementary material online, *Video S1*). Further evaluation by CMR with contrast and velocity flow mapping (*Figure 2* and *Table 2*) was performed. Consistent with the TTE, a 3.8 × 1.5 cm mobile mass in the RA was observed projecting into the RV (Supplementary material online, *Video S2*). The mass was isointense on T1-weighted images, hyperintense on T2-weighed images, and homogenous on delayed gadolinium enhancement (*Figure 2B–D*). While the tissue characteristics were consistent with rhabdomysarcoma,[4] IVL was considered most likely due to a lack of invasion into the myocardium. A multidisciplinary meeting involving radiology, gynaecological oncology, urological oncology, surgical oncology, and cardiac surgery was held to review the diagnostic imaging and therapeutic options. From this discussion, it was felt that total or at least partial resection was feasible, given the features observed on the TTE, CT, and CMR studies.

Approximately 2 weeks after admission, our patient underwent a single-staged thoracoabdominal resection. The procedure was well tolerated, and the entire mass (*Figure 3A* and *B*) was removed without complication. Resected samples stained strongly for the smooth muscle cell makers desmin and caldesmon, and cytology showed no atypia (*Figure 3C*), confirming the diagnosis of IVL. Consistent with previous reports,[5] samples from our patient strongly expressed oestrogen

Table 1 Laboratory studies on presentation

Tumour marker	Test value	Reference value
Carcinoembryonic antigen	0.9 ng/mL	0.1–5 ng/mL
CA 19-9	16 U/mL	0–37 U/mL
CA 125	70.4 U/mL	≤34.9 U/mL
Inhibin A	7 pg/mL	<98 pg/mL
Inhibin B	77 pg/mL	1–107 pg/mL

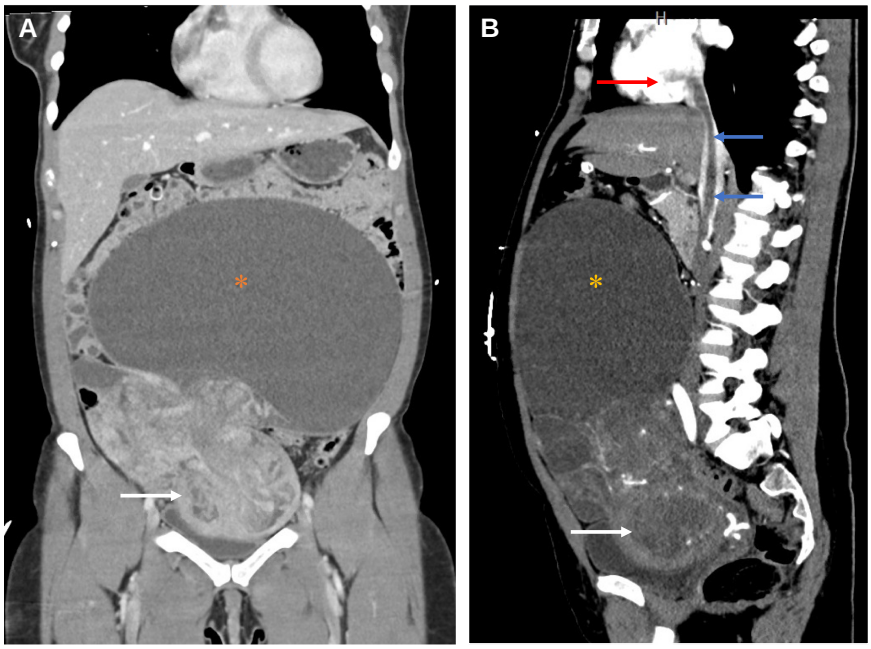


Figure 1 Computed tomography of the chest, abdomen, and pelvis with intravenous contrast. A mixed solid and cystic exophytic mass ~13.7 × 13.6 × 12.1 cm arising from the pelvis (*white arrow* marking the uterus) with a cystic component (*), measuring ~24.8 × 12.0 × 16.4 cm, extending into the epigastric region. (*A*) Coronal view. (*B*) Sagittal view. A non-occlusive filing defect in the inferior vena cava (*blue arrow*) extending into the right atrium (*red arrow*).

(*Figure 3D*) and progesterone receptors. Since discharge, the patient has had an uneventful course with regular follow-up.

Discussion

Intravenous leiomyomatosis is a rare condition characterized by the growth of a benign smooth muscle tumour within vascular channels. It is most commonly reported in premenopausal women and may present in patients with a history of hysterectomy.[1] While the exact incidence of IVL is unknown, fewer than 300 cases were reported between 1959 and 2010.[1] It is estimated that extrauterine involvement is seen in 30% of cases.[2] Extension into the systemic vascular system typically begins with growth into the ovarian vein, with direct access to the IVC from the right ovarian vein,[6] as seen in our patient. In the most advanced cases, IVL presents with cephalad growth along the IVC and entrance into the right cardiac chambers.

Clinically, the presentation of IVL is variable. Approximately 10% of patients with intracardiac leiomyomatosis are asymptomatic,[7] as in our case. Occult growth may not manifest until cardiac insufficiency from obstructive mass effect results in dyspnoea, syncope, and sudden death. While surgical resection is indicated for the definitive management for IVL, recurrence rates may be as high as 30%.[8] Thus, close monitoring for recurrence following resection is required; however, no definitive guidelines for post-operative follow-up have been established. Moreover, adjuvant medical therapy with tamoxifen and aromatase inhibitors has been proposed as a strategy to reduce recurrence. Given the rarity of IVL, no studies evaluating the efficacy of these therapies have been performed.[9] Our institutional tumour board recommended post-operative treatment with an aromatase inhibitor for our patient due to the extent of her disease on presentation. Additionally, our patient will be surveyed by CT every 6 months for the first post-operative year.

Cardiac imaging plays a critical role in the evaluation of IVL, and several features suggest the diagnosis. Echocardiogram revealing a freely a mobile mass, without attachment to the endocardium, and well-demarcated borders in the right heart chambers are consistent with IVL.[3] Moreover, TTE of the IVC may show a mobile, serpentine venous cast. These echocardiographic features of IVL are distinct from the fixed lesions more commonly observed in metastatic disease.[3] Transthoracic echocardiogram alone, however, may lead to misdiagnosis of a primary cardiac mass and CT imaging is important for identifying the origin, size, and route of invasion.[10] The utility of CMR in the evaluation of cardiac masses is well established.[4,10] In particular, the ability to discriminate differences in tissue densities and the resulting signal patterns on T1- and T2-weighted techniques can help narrow the differential. The tissue characteristics of IVL on CMR, however, remain poorly described. The tumour in our patient was isointense on T1-weighted images, hyperintense on T2-weighed images, and homogenous on delayed gadolinium enhancement, characteristics which are also seen in rhabdomyosarcomas. Importantly, CMR is also highly sensitive and specific for detecting tumour invasion into vessel walls,[11] and can help differentiate between malignant and benign growths. While imaging features may suggest a diagnosis of IVL, definitive diagnosis, for now, requires analysis of tumour samples by pathology.

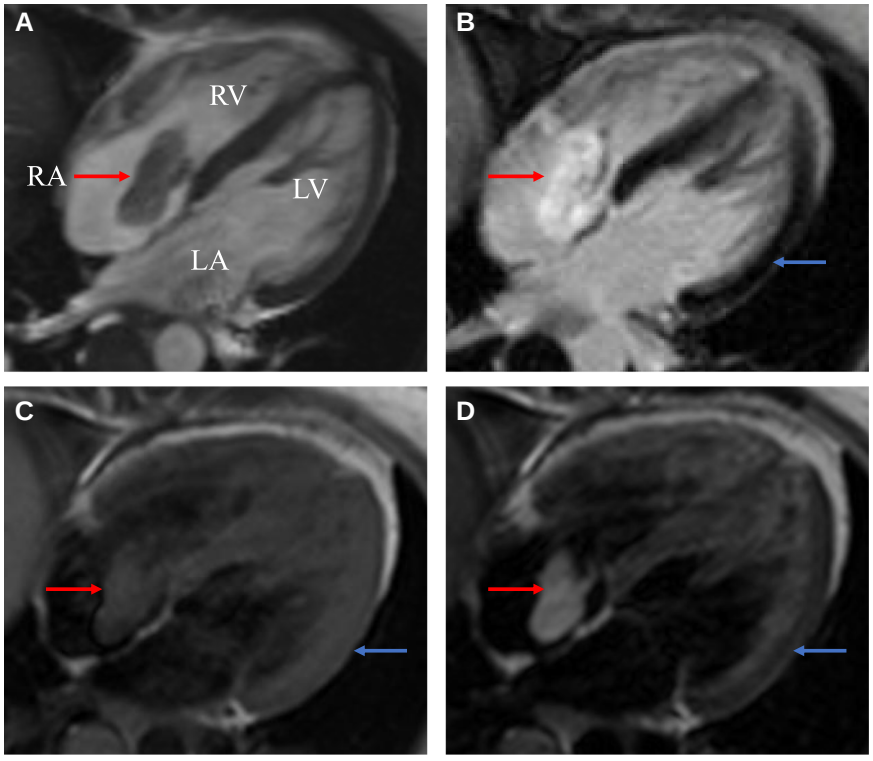
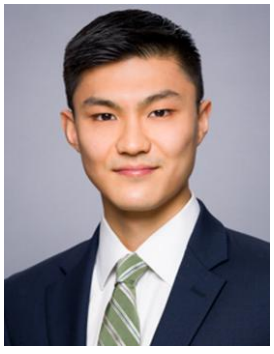


Figure 2 Cardiac magnetic resonance of right-sided heart mass. Cardiac magnetic resonance with contrast and velocity flow mapping. (*A*) Four-chamber view showing a 3.5 × 1.8 cm mass (*red arrow*) in the right atrium. (*B*). Delayed gadolinium imaging showing homogeneous late enhancement of right atrium mass. (*C*) T1-weighted imaging showing isointense right atrium mass compared with myocardium (*blue arrow*). (*D*) T2-weighted imaging showing hyperintense right atrium mass when compared with myocardium.

Table 2 Cardiac magnetic resonance with contrast and velocity flow mapping

Cardiac findings	
Left ventricular ejection fraction	59%
Right ventricular ejection fraction	50%
Velocity flow mapping	
Aortic forward volume	74 mL
Aortic regurgitant volume	2 mL
Peak aortic velocity	1.2 m/s
Pulmonary forward volume	70 mL
Pulmonary regurgitant volume	1 mL
Peak pulmonary velocity	1.2 m/s
Tissue characterization of mass	
T1-weighted images	Isointense
T2-weighted images	Hyperintense
Delayed gadolinium enhancement	Homogenous

Lead author biography



Daniel S. Kikuchi is a first-year resident in the Osler Medical Residency at The Johns Hopkins Hospital. He received his medical degree from Emory University School of Medicine, where he developed an interest in cardiovascular disease. Following residency training, he hopes to obtain a cardiology fellowship. His main interests are in arrhythmias, cardio-oncology, and health disparities.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

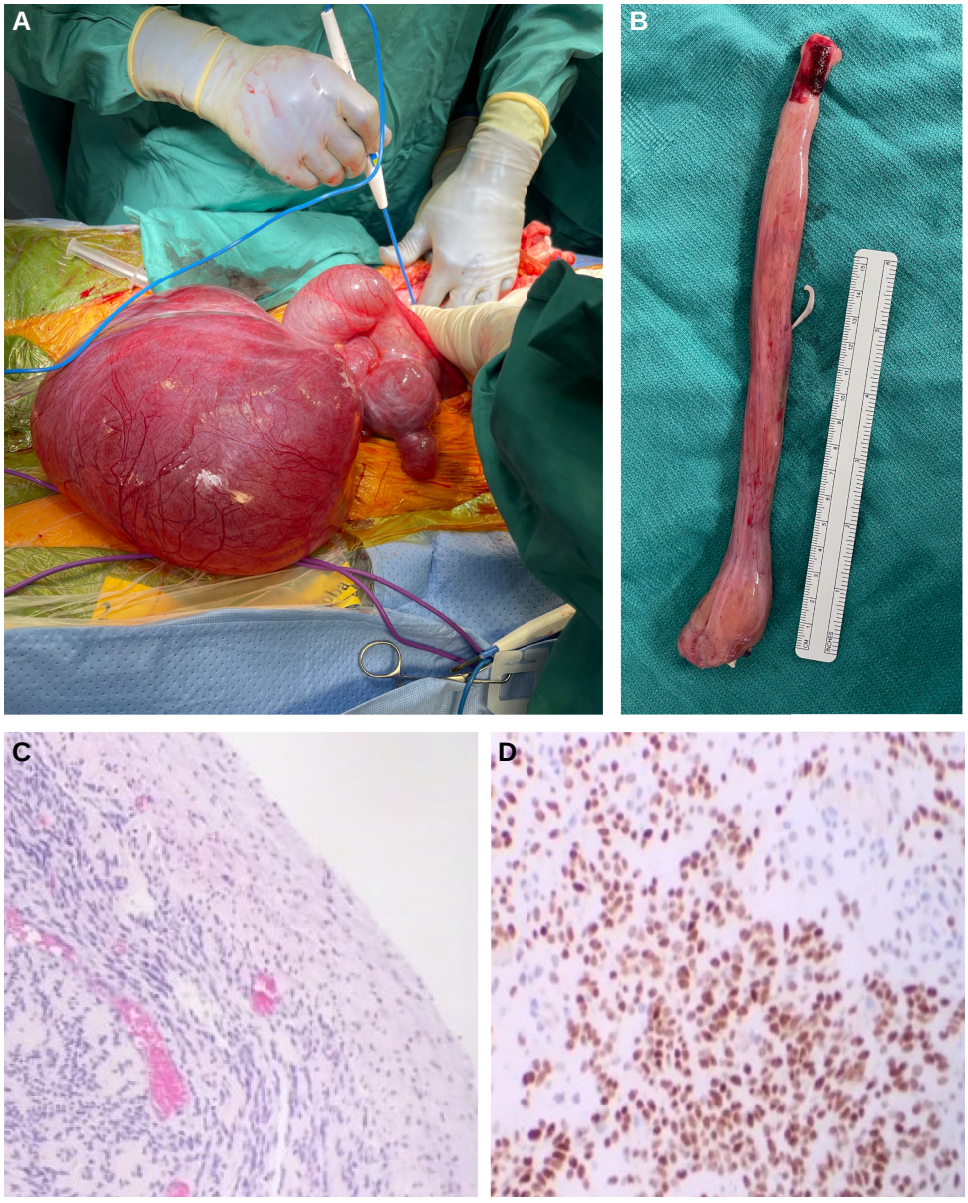


Figure 3 Gross surgical specimens and histology. The entire mass was resected and sent to pathology for analysis. (*A*) Intraoperative image showing mixed solid and cystic mass in the pelvic region. (*B*) Tumour cast excised from the inferior vena cava and right heart chambers. (*C*) Representative H&E slide showing smooth muscle cells without atypia. (*D*) Resected samples strongly expressed oestrogen receptor.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: Written consent for submission and publication of the images and associated text has been obtained from the patient in accordance with COPE guidelines.

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