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CASE REPORT
Other

Intravascular leiomyomatosis with cardiac extension: a case report

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Background Intravascular leiomyomatosis (IVL) with intracardiac extension is a rare benign tumour seen exclusively in women, characterized by proliferation of uterine smooth muscle cells through the venous circulation into the inferior vena cava (IVC) and the right heart chambers.

Case summary A 47 years old women with history of previous hysterectomy due to myomatosis, presented with nausea, anorexia, and bilateral lower limb swelling over the preceding 2 months. An outpatient abdominal ultrasound discovered a mass in the IVC. Echocardiogram and computed tomography demonstrated a large intravascular mass extending from the pelvis to the right heart chambers. The tumour was completely removed in a concomitant open-heart surgery and laparotomy. Post-operative course was uncomplicated. A month later, the patient was feeling well and in good clinical condition. The histological analysis consisted with IVL.

Discussion Intracardiac leiomyomatosis is a rare clinical condition which requires high index of suspicion. Multimodality imaging is usually required to establish the preoperative diagnosis, although the final diagnosis is achieved with tissue investigation. Complete surgical resection of the tumour is curative and associated with good long-term prognosis.

Keywords Case report • Leiomyomatosis • Hysterectomy • Intravascular extension • Intracardiac

ESC Curriculum 6.8 Cardiac tumours • 2.1 Imaging modalities

Learning points

- Intracardiac leiomyomatosis should be suspected when a right heart chamber mass extending from the inferior vena cava is detected in middle-aged women, especially when a history of hysterectomy is present.
- Complete surgical resection of the tumour is curative and associated with good long-term prognosis.

Introduction

Intravascular leiomyomatosis (IVL) is a rare condition, characterized by intravascular proliferation of uterine leiomyoma through the venous circulation.[1–3] When the extension continues further through the inferior vena cava (IVC) and penetrates into the right heart chambers, the tumour is called intracardiac leiomyomatosis. The clinical presentation is variable and mostly related to the severity of cardiac involvement.[1–5]

As the risk of death is due mainly to obstruction of the right outflow tract, early surgical treatment is recommended. Complete

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surgical resection is curative,[1,4–6] while incomplete resection is associated with high recurrence rate.

Timeline

Timeline	Event
2018	The patient had partial hysterectomy and right adnexectomy due to menometrorrhagia and a space occupying lesion involving the uterus and right ovary with histological evidence of uterine leiomyomas.
2 months preceding to admission	Patient has been complaining nausea, anorexia, and worsening bilateral lower limb swelling.
On arrival	Echocardiogram demonstrated a large mass extending from inferior vena cava to the right heart chambers.
Day 7	Complete resection of the tumour in concomitant open-heart surgery and laparotomy.
Day 12	Discharge from admission after uncomplicated post-operative course.
Day 39	Histopathology final analysis confirmed intravascular leiomyomatosis.
Day 45	Follow-up visit documented that the patient was feeling well and was found in good clinical condition.

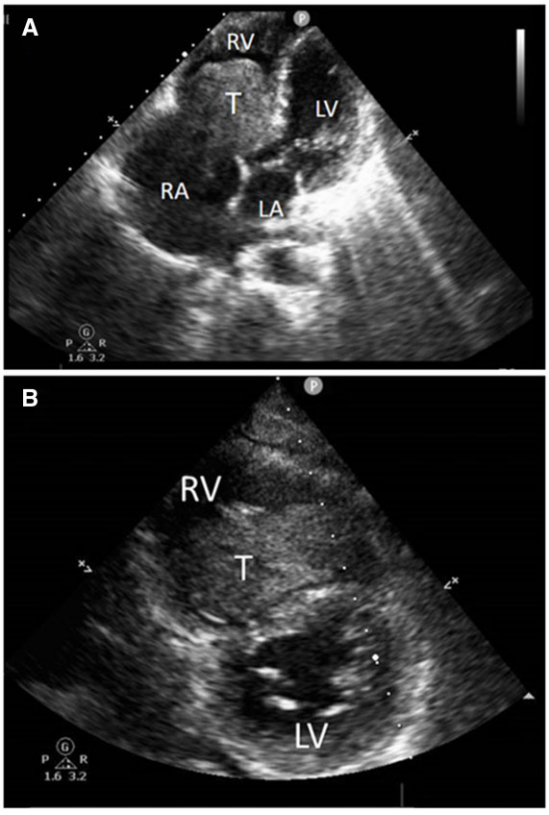


Figure 1 Transthoracic echocardiogram showing a highly mobile tumour within the right atrium and right ventricle in four-chamber view (*A*) and in parasternal short axis (*B*). LV, left ventricle; RA, right atrium; RV, right ventricle; T, tumour.

Case presentation

A 47 years old Caucasian women (multiparous) was referred to Assuta Ashdod Medical Centre for the investigation of a mass in her IVC discovered on an abdominal ultrasound. She had been complaining of nausea, anorexia, and bilateral lower limb swelling over the preceding 2 months.

Her past medical history was remarkable for partial hysterectomy and right adnexectomy 3 years earlier due to menometrorrhagia and a space occupying lesion involving the uterus and right ovary with histological evidence of uterine leiomyomas and simple ovarian cyst. The patient did not continue with any gynaecological surveillance. No other comorbidities nor prior use of medication.

On presentation, the patient was comfortable with vital signs within normal limits. Prominent jugular vein distention and bilateral lower extremities oedema was noted.

Laboratory examination and coagulation study results were unremarkable. D-Dimer was within normal limits.

Transthoracic echocardiogram demonstrated (*Figure 1*, *Videos 1–3*) a large mass extending from the IVC into the right atrium and the right ventricle through the tricuspid valve, causing severe functional tricuspid stenosis. The mass was highly mobile, stalk free, without any adhesion to the right chamber walls. The right ventricle was severely dilated, with impaired contractility small to moderate pericardial effusion was observed.

A thoraco-abdominal computed tomography (CT) demonstrated a mass extending from the right ovary veins through the IVC and entering into the right atrium, ventricle, and right ventricular outflow tract. No evidence of pulmonary embolism was observed. A heterogeneous mass with lobular boundaries was demonstrated in the uterine bed.

A cardiac CT (*Figure 2*) demonstrated a homogenous mass, extending from the IVC through the right heart chambers and the right ventricular outflow tract, without any adhesion to the cardiac wall.

Treatment with IV heparin was initiated on the patient's admission considering the Differential Diagnosis of a large thrombus. Due to the large intravascular mass extending into the heart, accompanied with right heart obstructive features, the patient was transferred to the Cardiothoracic surgery department for an urgent surgical intervention.

A concomitant operation by multidisciplinary teams was performed by a dual approach, sternotomy and laparotomy (*Figure 3A*).

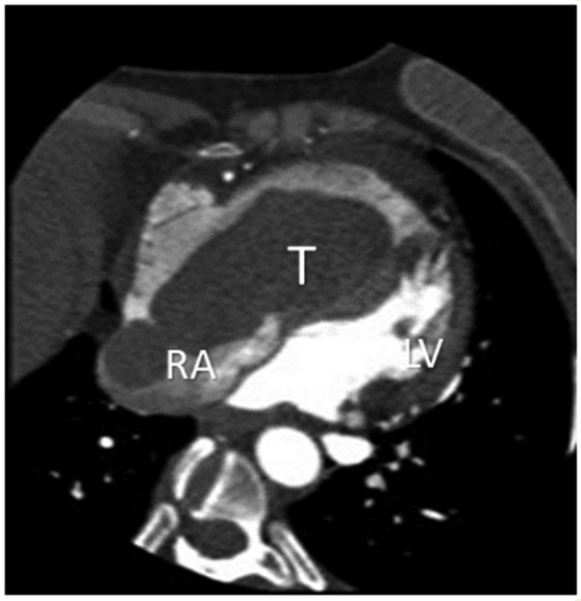


Figure 2 A cardiac computed tomography showing a homogenous mass, extending from the inferior vena cava through the right heart chambers. LV, left ventricle; RA, right atrium; RV, right ventricle; T, tumour.

The IVC and the right gonadal vein were opened simultaneously and the tumour was separated completely from the vessels. The tumour was excised from IVC to right atrium junction in one block (*Figure 3B*). The patient had an uncomplicated post-operative course and was discharged home on post-operative Day 5. On a follow-up visit a month later, the patient was feeling well and was found in good clinical condition.

Microscopic examination showed spindle-shaped cells arranged in bundles, without evidence of atypia, necrosis, or excess mitoses. Immune-histochemical staining was positive for smooth muscle cell markers (caldesmon, desmin, and smooth muscle actin). The histological findings consisted with the IVL.

Discussion

Intravascular leiomyomatosis is a rare, benign, smooth muscle tumour, originating from the uterine or pelvic veins, and invading through the venous circulation including the IVC. Rarely, these tumours can further extend to the right heart and pulmonary artery. Intravascular leiomyomatosis was first reported by Birch-Hirschfeld in 1896[7] and until 2013, <200 cases with cardiac extension have been described in the English literature.[1]

Similar to our patient, most patients are middle-aged women (~90% between 40 and 50 years), and multiparous (~75%).

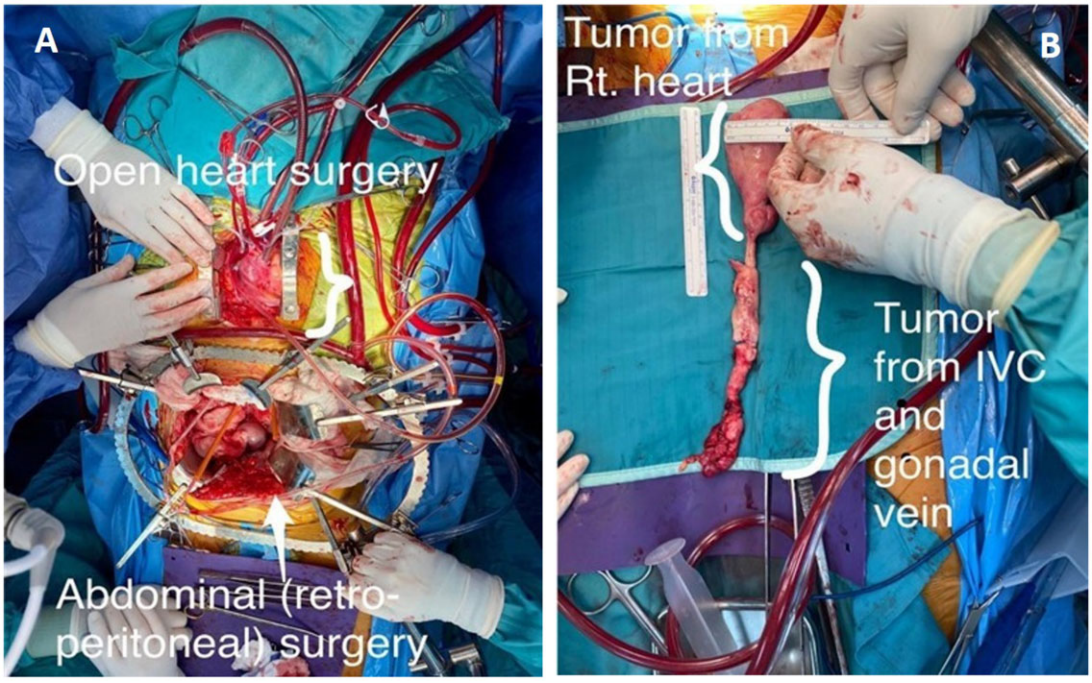
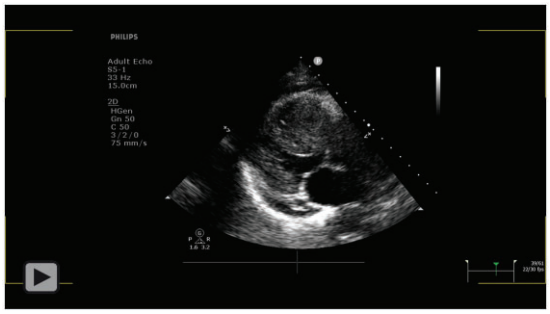
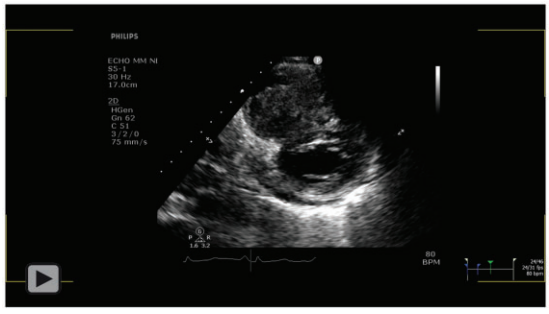


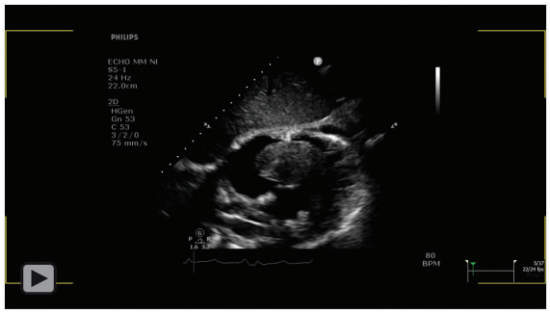
Figure 3 (*A*) An overhead view of the surgical field demonstrating the concomitant surgery. (*B*) An overview of the tumour, after its complete removal in one piece.



Video 1 Transthoracic echocardiogram showing the tumor in RA and RV in parasternal long view.



Video 2 Parasternal short view.



Video 3 Subcostal view.

Approximately 85% have either undergone a previous hysterectomy/myomectomy or had a coexisting uterine leiomyoma.[1]

The clinical presentation is variable and mostly related to the severity of cardiac involvement.[1–4,6] Symptomatic patients present with dyspnoea, syncope, and right heart failure.

Depending on the size and the extension of the tumour, valvular dysfunction (either stenosis or regurgitation) may be observed.

Imaging is required for preoperative diagnosis and plan, and multimodality approach is recommended. Echocardiography is useful in assessment of the tumour size and mobility and valvular involvement. Echocardiographic features include a stalkless, mobile mass, non-adherent to the right heart chamber walls, encroaching the tricuspid orifice.[3]

Transoesophageal echocardiogram (TOE) is superior to transthoracic echo in the evaluation of atrial masses. Also, intraoperative TOE is recommended for localization of the mass and to minimize vascular injury. Computed tomography and magnetic resonance imaging are also useful in evaluating the size, origin and extent of the tumour.

The differential diagnosis for intravascular mass extending into the right heart includes, thrombus, atrial myxoma, abdominal tumours, which can be accompanied with intravascular tumour thrombus such as: renal cell carcinoma, hepatocellular carcinoma, Wilms tumour, adrenal carcinoma, leiomyosarcoma of the uterus, or metastatic tumours.[1,4,6] Atrial myxoma generally attached to the atrial septum through a stalk, with left-sided predominance.[1,3] Renal cell carcinoma or Wilms tumour should be suspected when a kidney mass is demonstrated by imaging.[1] A thrombus has homogenous, avascular appearance, and is usually adjacent to an asynergic myocardium. It is more common post-operatively, in the presence of indwelling catheters and especially at the junction of the superior vena cava and right atrium.[3] In our patient, a thrombus was less likely due to the well-demarcated borders, lack of attachment to the cardiac wall, and the negative D-dimer. Leiomyosarcoma often shares many clinical characteristics with IVL and should be differentiated by pathology workup.

Considering the risk of sudden death due to total obstruction of the right ventricular outflow tract, early surgical treatment is recommended. Complete surgical resection is curative,[1,2,4–6] while incomplete resection is associated with a high recurrence rate (~33%).[1]

Surgery can be performed by a dual approach (thoracic and abdominal), using one- or two-stage procedure.[1,2,4–6] The two-stage procedure is recommended for patients in poor condition who cannot tolerate an extensive one-stage procedure, or when the tumour is extensive or adheres to the vascular wall. A total hysterectomy and bilateral salpingo-oophorectomy should be performed.

Owing to the oestrogen-dependent growth of the tumour, adjunctive therapy with tamoxifen has been proposed; however, it has been incompletely studied and its efficacy is controversial.[2]

Histological examination is characterized by bundled arrangement of spindle-shaped cells and fibromuscular tissue, without atypia or increased mitotic index. Immunohistochemical staining is positive for smooth muscle markers (desmin and actin).[1]

Conclusions

Intravascular leiomyomatosis is a rare condition that should be suspected when a right heart chamber mass extending from the IVC is detected in middle-aged women, especially when a history of myomectomy or hysterectomy is present. Multimodality imaging is very helpful in the preoperative evaluation. Complete surgical resection is potentially curative and holds a good long-term prognosis.

Lead author biography

Einat Shaked is an internal medicine specialist and a senior resident at the Cardiology Division, Assuta Ashdod University Hospital, Ashdod, Israel.

Supplementary material

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

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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: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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