

## Cardiac Imaging

## Heart Leiomyoma Extending from the Inferior Vena Cava to the Right Heart

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 heart failure.**

**A** 60-year-old woman, with a history of hysterectomy for leiomyoma two years earlier, was admitted to our department because of deteriorating fatigue and dyspnoea on exertion, pedal oedema, and a recent episode of syncope. Physical examination revealed an early diastolic plop associated with a rumbling murmur in the left parasternal region, hepatomegaly, ascites and severe oedema of the lower extremities.

Transthoracic echocardiography revealed mild right ventricular dilatation with preserved systolic function (S-wave velocity measured with pulsed tissue Doppler in the basal free wall of the right ventricle >13 cm/s). In addition, a mass of moderate echogenicity was identified inside the right ventricle in the long left parasternal view (Figure 1A, arrow), as well as in a modified apical 4-chamber view (Figure 1B, arrow). Partial obliteration of the flow of the inferior vena cava due to another mass was also revealed (Figure 2, arrow). The patient then underwent transoesophageal echocardiography. In the bicaval 110° view an elongated multilobular mass could be seen inside the right atrium, originating from the inferior vena cava (Figures 3A, 3B, arrow). In the mid-oesophageal 60° view, as well as in the 4-chamber 0° view, the mass was shown to extend inside the right ventricle after crossing the tricuspid valve, reaching up to the outflow tract and partially obliterating it (Figures 3C

and 3D, respectively, arrow). The patient underwent a ventilation-perfusion lung scan which was interpreted as low probability for pulmonary embolism.

Taking into consideration the history of the previous hysterectomy for a leiomyoma, and the apparent extra-cardiac origin of the tumour, we suspected that a probable diagnosis could be leiomyoma. The symptoms of the patient were mainly attributed to the significant obstruction of the inferior vena cava and the subsequent low preload of the right heart. In addition, the partial, though limited, obstruction of the right outflow tract may have also contributed to the overall pathophysiology.

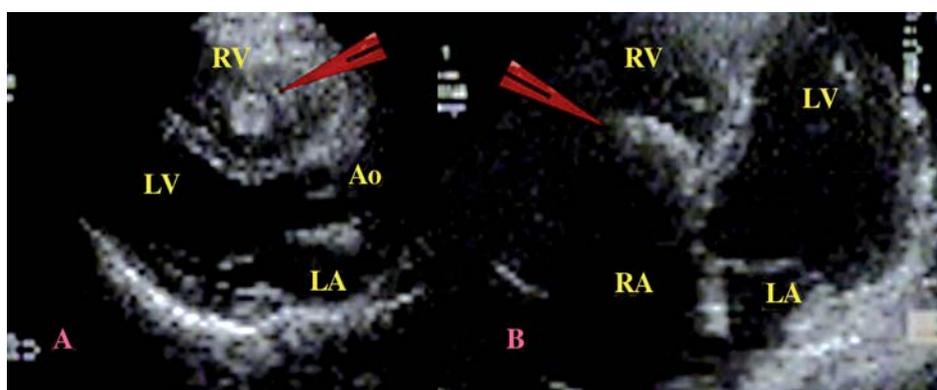
The patient subsequently received open heart surgery. Histological examination of the resected mass confirmed intravenous leiomyomatosis. The mass actually originated from the wall of the inferior vena cava, extending inside the right cavities as far as the right ventricular outflow tract.

Primary tumours of the heart are rare — necropsy studies show incidences between 0.0017-0.28% — and most of them are benign. However, 10-20% of all primary cardiac tumours are malignant.<sup>1</sup> The specific signs and symptoms produced by tumours are more closely related to their precise anatomical location than to their histological type. The most common clinical manifestations of right-sided tumours are those of right heart failure, which are attributed to obstruction of tricuspid valve

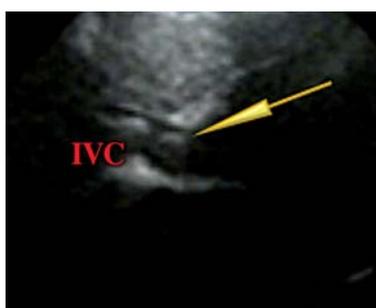
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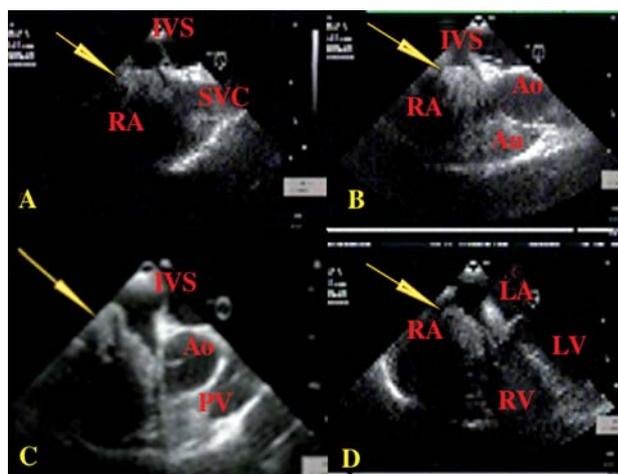
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**Figure 1.** Transthoracic echocardiography. A: Long left parasternal view. B: Modified 4-chamber view. A mass of moderate echogenicity is visible in the right ventricle (arrow).  
Ao – aorta; LA – left atrium; LV – left ventricle; RA – right atrium; RV – right ventricle.



**Figure 2.** Subxiphoid view. Partial obliteration of the flow of the inferior vena cava (IVC) due to a mass (arrow).



**Figure 3.** Transoesophageal echocardiography. A. Bicaaval 110° view. B. Mid-oesophageal 90° view at the level of the auricle of the right atrium. Elongated multilobular mass inside the right atrium, originating from the inferior vena cava (arrow). C. Mid-oesophageal 60° view. D. mid-oesophageal 0° 4-chamber view. The mass extends inside the right ventricle, reaching up to the outflow tract, and partially obliterating it (arrow).  
Ao – aortic valve; IVS – intraventricular septum; PV – pulmonic valve; SVC – superior vena cava.

flow, and/or incomplete filling of the right ventricle, and/or obstruction of outflow tract flow. Pulmonary embolism and pulmonary hypertension can sometimes occur (though not in this case). Intravenous leiomyomatosis is an uncommon non-malignant tumour which originates from the smooth muscle cells and is usually confined to the pelvic venous system. Rarely, intracaval and intracardiac extension has been described: the literature includes only 100 cases with cardiac involvement.<sup>2-5</sup>

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