

# Intracardiac leiomyomatosis: iliac vein to right-ventricular outflow tract

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## SUMMARY

**Background** A 58-year-old female with a history of hypertension and asthma presented to an internist for a routine physical examination. A grade II/VI systolic ejection murmur and electrocardiogram abnormalities were noted. She was referred to a cardiologist for further assessment.

**Investigations** Transthoracic echocardiography, transesophageal echocardiography, contrast-enhanced CT and MRI, exploratory laparotomy.

**Diagnosis** Intracardiac leiomyomatosis.

**Management** Surgical excision.

**KEYWORDS** cardiac tumor, inferior vena cava, leiomyoma

## CME

This article offers the opportunity to earn one Category 1 credit toward the AMA Physician's Recognition Award.

## THE CASE

A 58-year-old woman with a history of hypertension and asthma presented to an internist for a routine physical examination. The patient had no symptoms, but physical examination revealed a grade II/VI systolic ejection murmur. On an electrocardiogram, a new right-bundle-branch block pattern was noted and she was referred to a cardiologist. A mass was seen in the right atrium on transthoracic echocardiography, and on transesophageal echocardiography a large multilobed serpentine mass was visualized extending from the inferior vena cava into the right ventricle (Figure 1A–C). The mass crossed the tricuspid valve without obstruction and extended into the right-ventricular outflow tract to the level of the pulmonic valve (Figure 1D). Right-ventricular systolic pressure was estimated to be 26 mmHg. The cross-sectional area of the mass ranged from 1.38 to 5.45 cm<sup>2</sup> according to measurement on transesophageal echocardiograms.

To further evaluate the origin and extent of the mass, contrast-enhanced CT of the chest and abdomen was performed. The cardiac mass was confirmed to originate from the inferior vena cava; however, it was still unclear whether the caval component represented a thrombus or tumor. No hilar or mediastinal adenopathy and no pericardial effusion were noted. MRI of the pelvis revealed a 7.2 × 4.0 × 4.3 cm complex right-adnexal mass, contiguous with the uterus, suspected as being neoplastic. The patient was admitted to the hospital for definitive diagnosis.

On admission, the patient remained completely asymptomatic. A team of cardiothoracic and vascular surgeons performed a combined laparotomy and thoracotomy. The mass was excised from the distal part of the inferior vena cava. The tumor could not be safely detached from its origin, however, necessitating transection at the line of the right internal iliac

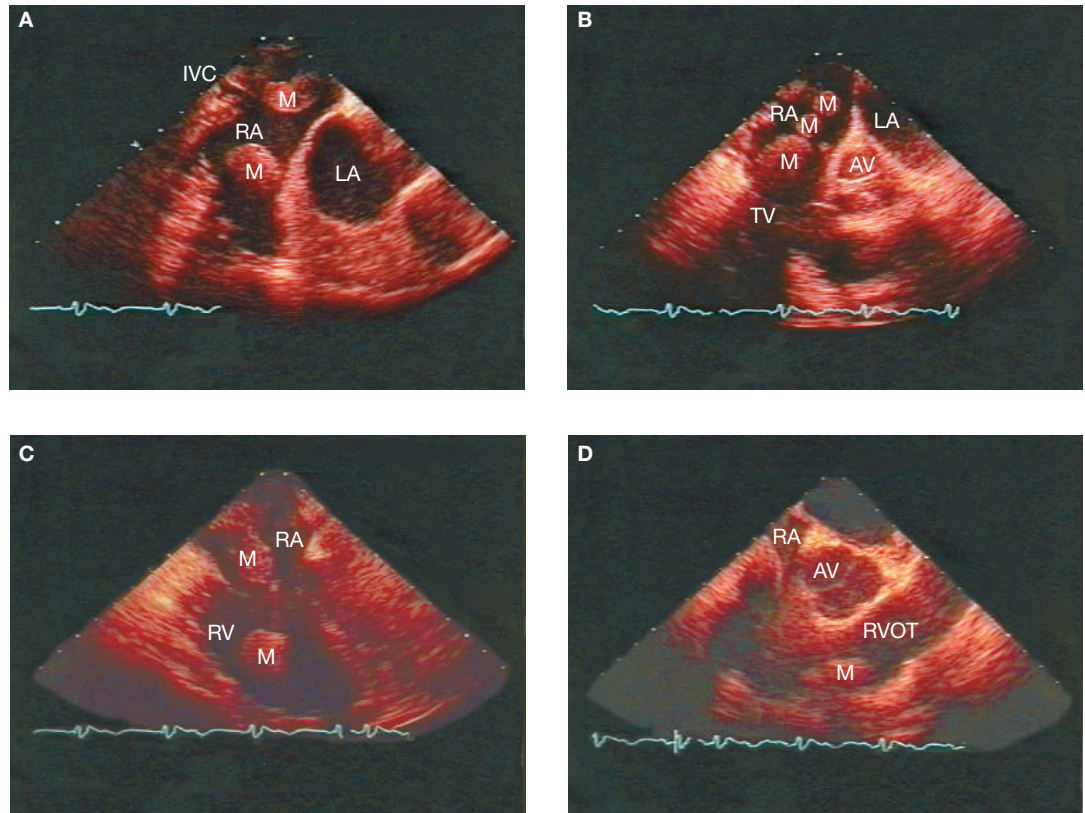
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**Figure 1** Transesophageal echocardiogram. (A) Transesophageal echocardiogram revealed tumor extension from the inferior vena cava into the right atrium. (B) A multilobed mass was seen in the right atrium just proximal to the tricuspid valve, (C) extending into the right ventricle and (D) crossing the tricuspid valve with extension into the right-ventricular outflow tract just proximal to the pulmonic valve. AV, aortic valve; IVC, inferior vena cava; LA, left atrium; M, leiomyoma; RA, right atrium; RV, right ventricle; RVOT, right-ventricular outflow tract; TV, tricuspid valve.

vein. The ovaries appeared bilaterally atrophic, with no adnexal masses present. A small uterine leiomyoma measuring 2.0 cm in diameter was visualized. The suspicious shadow seen in the pelvis on the MRI was not identified at surgery and was thought to have represented a fluid collection.

Pathologic examination of the specimen revealed a cylindrical mass measuring 30.0 cm in length and 1.5–2.5 cm in diameter (Figure 2). A branching segment measuring 4.5 cm in length and 1.2 cm in diameter arose 12.0 cm from the end of the surgical margin. The outer surface was tan to pink, smooth and glistening in appearance. Histologic examination showing fascicles of spindle cells was consistent with a leiomyoma of the inferior vena cava extending into the right heart (Figure 3). The patient had an uneventful postoperative course and was discharged 7 days later.

#### DISCUSSION OF DIAGNOSIS

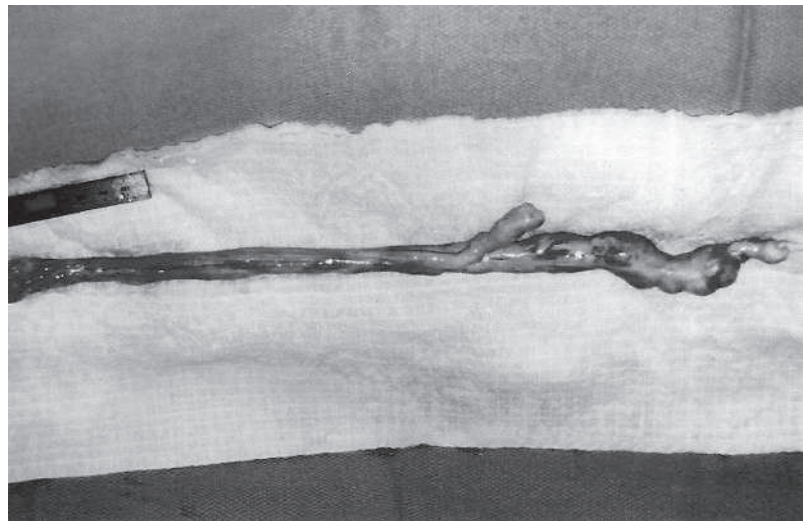
Metastatic tumors that involve the heart, pericardium or both are 20–40 times more common than primary tumors of the heart.<sup>1–3</sup> Although most malignancies can involve the heart, carcinomas of the lung and breast do so most often. Tumors metastasize to the heart via hematogenous and lymphatic spread or by direct invasion, and can involve the pericardium, myocardium, valves and coronary arteries. Manifestations of pericardial involvement usually include pleuritic chest pain and pericardial friction rubs, along with pericardial effusion. Atrial and ventricular arrhythmias are common with myocardial involvement. Intracavitary tumors usually result from extension of the primary tumors; most commonly hepatocellular carcinoma, renal carcinoma or uterine leiomyomatosis via extension into the inferior vena cava.<sup>2–4</sup>

Primary cardiac tumors are rare, with an autopsy incidence between 0.001% and 0.33% in the general population. The risk would be even smaller in a female more than 50 years old. There are no particular factors that predispose people to developing cardiac tumors.<sup>1</sup> The differential diagnosis in our patient included tumor, thrombus, cyst, vegetation, enlarged eustachian valve or artefact. An echocardiogram with subsequent CT was required to confirm our suspicions of a mass rather than the other diagnoses. The MRI was then helpful in localizing the origin as contiguous with the uterus. The final diagnosis was made at surgery.

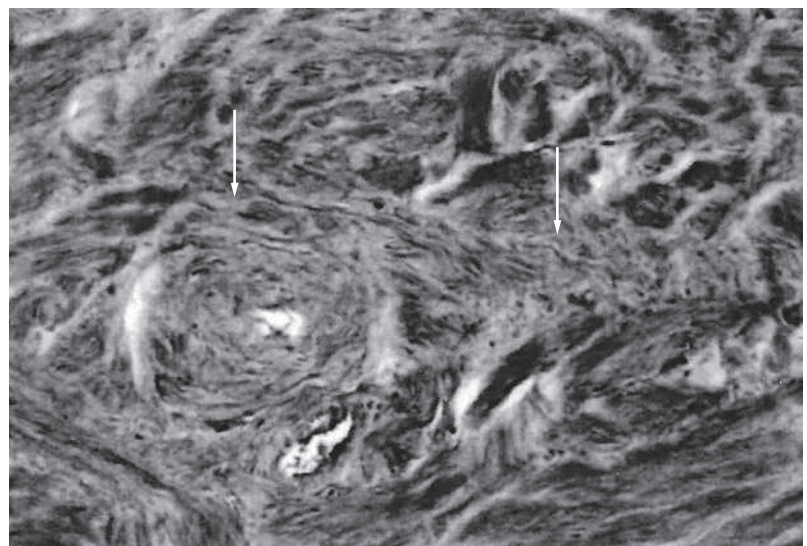
Intravenous leiomyomatosis was first defined by Norris and Parmley as a leiomyoma arising from the wall of a uterine or ovarian vein, or from a uterine leiomyoma that infiltrates uterine veins and extends into the vessel lumen. In most cases, intravenous leiomyomatosis remains localized within the uterine vessels, but extension into the central vessels and right heart can occur, as seen in our patient. The frequency of retained intravenous leiomyomas in uterine vessels is not known. At least 72 cases of intravenous leiomyomatosis with extension into the right heart chambers have been reported (Table 1).<sup>5,6</sup> Intravenous leiomyomatosis is an insidious process and extension into the right heart, leading to obstruction or other averse events, can occur slowly over many years.<sup>7,8</sup> Many cases are, therefore, discovered incidentally. Patients may remain asymptomatic until direct intracardiac extension causes valve obstruction or cavity compromise, leading to cardiac insufficiency. Death secondary to heart failure caused by mechanical obstruction and/or intracardiac extension of these lesions has been reported.<sup>9</sup> As the case we report demonstrates, however, cardiac function can remain completely uncompromised.

### TREATMENT AND MANAGEMENT

The treatment of intravenous leiomyomatosis is surgical removal of the tumor, because it can cause obstruction, valvular regurgitation or decrease cardiac performance to the point of overt heart failure. In our patient, exploratory laparotomy was performed without evidence of intra-abdominal pathology. The pelvis was inspected and both ovaries appeared to be atrophic. A uterine leiomyoma measuring 2.0 cm was seen that had no malignant features and required no further intervention. The retroperitoneum was explored and the



**Figure 2** Gross pathologic specimen of leiomyoma measuring 30.0 cm in length, with a diameter ranging from 1.5 to 2.5 cm.



**Figure 3** Microscopic examination of leiomyoma with focal hyalinization and myxoid degeneration. The left-hand arrow indicates focal hyalinization and the right-hand arrow indicates myxoid degeneration.

inferior vena cava identified. The next step was a midline sternotomy. Cardiopulmonary bypass was established and the right atrium was opened with an incision between the right atrial appendage and the inferior vena cava. The mass was easily identified, freely floating within the right atrium. Simultaneously, the inferior vena cava was opened intra-abdominally, just above the iliac bifurcation. The tumor was observed in the inferior vena cava, carefully dissected, and removed through the venotomy in one piece. It was superiorly detached from the right atrium.

**Table 1** Summary of a total of 72 cases of intracardiac leiomyomatosis reported between 1900 and 2005.

Characteristics	Number of patients (%)
History of hysterectomy	40 (55.6%)
<b>Presenting symptoms</b>	
Congestive heart failure	47 (65.3%)
Venous obstruction	4 (5.56%)
Abdominal distension	11 (15.2%)
Sudden death	1 (1.39%)
Others	9 (12.5%)
<b>Extent of involvement</b>	
Right atrium	32 (44.4%)
Right ventricle	32 (44.4%)
Pulmonary vessel	7 (9.72%)
Tricuspid valve	1 (1.39%)
<b>Route of extension</b>	
Unknown	15 (20.8%)
Unilateral	53 (73.6%)
Bilateral	4 (5.56%)
Iliac only	35 (48.6%)
Ovarian only	17 (23.6%)
Both iliac and ovarian	5 (6.94%)

The median age at diagnosis was 47 years (range 26–76).

**Competing interests**

The authors declared they have no competing interests.

It was not possible to detach the tumor from its origin as it was embedded in the base of a vascular structure, so it was transected, almost at the line of the internal iliac vein. The tumor was sent for pathology. The inferior vena cava incision was closed and the patient was gradually weaned off bypass without difficulty. The chest and laparotomy incisions were then closed in a routine fashion. The patient was observed, remained stable and was transferred to the recovery unit in satisfactory condition. She was discharged 1 week later.

Currently, the patient remains asymptomatic. She no longer has an audible systolic ejection murmur and there is resolution of the right bundle-branch-block pattern on electrocardiogram. Follow-up monitoring has been undertaken with ultrasonography and echocardiography for 2 years without evidence of recurrence; annual follow-ups will continue.

**CONCLUSION**

Intravenous leiomyomatosis should be considered in the differential diagnosis of a middle-aged woman with a right-heart tumor. Of note, this process can occur even in the absence of uterine tissue, for example in women with a history of hysterectomy, presumably by intravenous spread.<sup>5,6,8,10</sup> Large vascular leiomyomas usually arise from one site, most commonly the iliac vein.<sup>6</sup> CT and MRI can be used to identify the site of origin and to assess the extent of vascular involvement, as was done in our patient. This possibility warrants long-term follow-up with vascular and cardiac ultrasonography as well as observation by the physician. The management of leiomyomas that remain localized within uterine vessels is observation and annual follow-up.

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