

Intracardiac Leiomyomatosis

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Abstract

Intravenous leiomyomatosis is a histologically benign smooth-muscle tumor arising from either a uterine myoma or the walls of a uterine vessel with extension into veins. We describe echocardiographic features of intravenous leiomyomatosis with spread into the right-sided cardiac chambers. The patient was a middle-aged woman, with prior history of hysterectomy 2 years earlier who presented with cardiac symptoms and signs. Echocardiographic features included: 1) elongated mobile mass extending from the inferior vena cava, and 2) multiple masses in the right-heart chambers (right atrium and ventricle).

Intracardiac leiomyomatosis should be considered in a female patient presenting with an extensive mass in the right-sided cardiac chambers (*Iranian Heart Journal 2006; 7 (4): 61-66*).

Key words: echocardiography ■ leiomyomatosis ■ cardiac tumor

Leiomyomatosis is a rare uterine tumor defined as the extension into venous channels of a histologically benign smooth-muscle tumor arising either from a uterine myxoma or from the walls of a uterine vessel.¹

Cardiac involvement is present in up to 10% of cases and may be misdiagnosed as a primary cardiac tumor or venous thrombus-in-transit.²

The first case was reported by Birch-Hirschfield in 1897,³ and the first case report in the English literature was described by Marshall and Morris in 1959.⁴

We describe a case of intracardiac leiomyomatosis (intravenous leiomyomatosis with extension into the right-sided heart chambers) and emphasize the echocardiographic features of this rare tumor.

Case report

A 46-year-old woman presented with worsening dyspnea on exertion of several weeks' duration. Past medical history included a hysterectomy with salpingo-oophorectomy 26 months before due to fibromatoid uterus, leiomyomatous mass of the left broad ligament with hydropic and cystic degeneration, and cardiac surgery 8 months before for a right atrial mass. The resected cardiac tumor consisted of an elongated banana-shaped mass, measuring 15 cm in length and 3.5 cm in greatest diameter. Histologic examination showed hypocellular as well as hypercellular areas composed of isolated and bundles of spindle cells with oval nuclei and long, slender cytoplasmic processes.

Received Dec. 14, 2005; Accepted for publication Nov. 26, 2006.

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No pleomorphism or mitosis was seen. These findings were consistent with leiomyoma (Fig. 1).

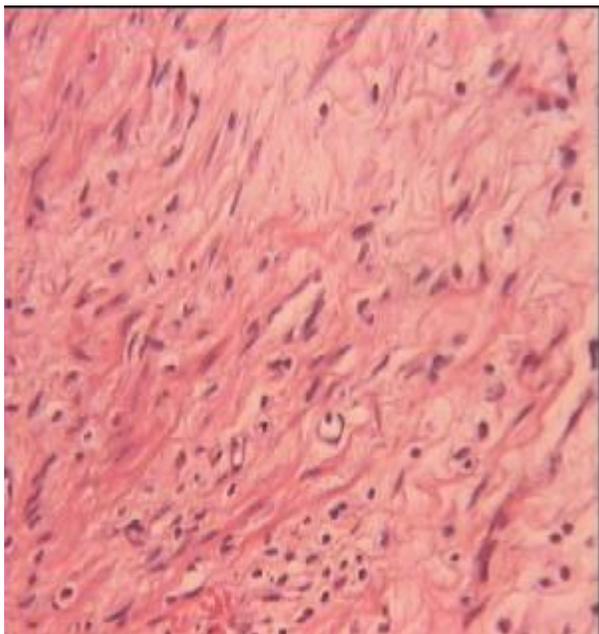
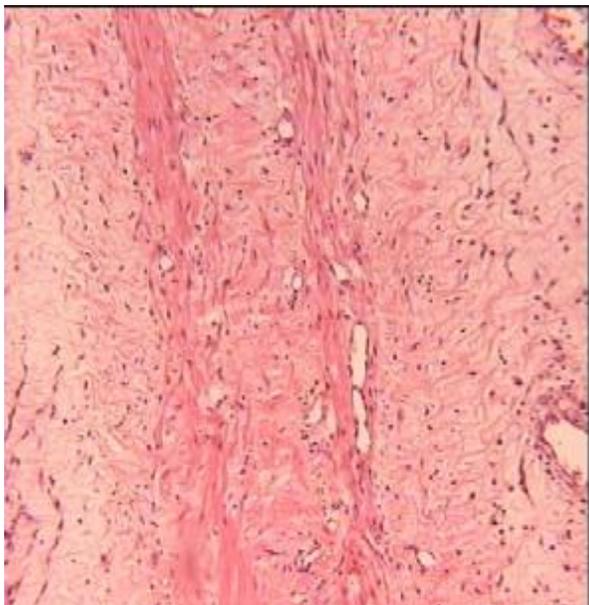


Fig. 1. Histopathologic appearance of the resected cardiac tumor shows bundles of spindle cells with oval and long, eosinophilic cytoplasmic processes.

On physical examination, a grade 2/6 systolic murmur increasing with inspiration was audible at the left lower sternal border; other physical findings were normal. On transthoracic echocardiography, the right atrium (RA) and ventricle (RV) were markedly enlarged, and moderate tricuspid regurgitation was noted. Doppler calculated RV systolic pressure was increased at 40-45mmHg. A large (6 by 3.5 cm) mobile non-homogenous prolapsing mass was seen in the right atrium which extended from the inferior vena cava into the RA. Another smaller mass also was seen in the RVOT.

The inferior vena cava was severely dilated (3 cm) and was nearly filled by the tumor at least 10 cm below its entrance to the right atrium (Figs. 2, 3).



Fig. 2. Large prolapsing mass in RA and a smaller mass in RVOT.



a

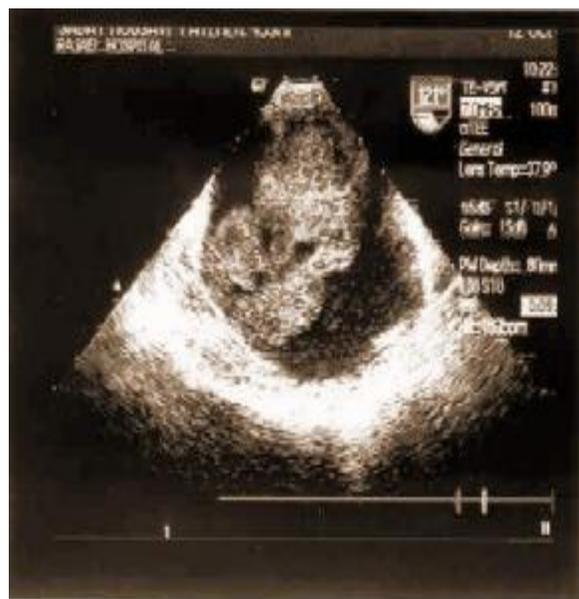


b

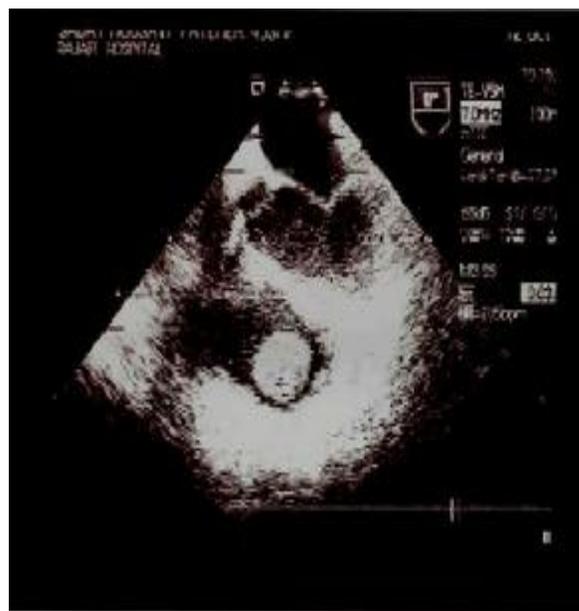
Fig. 3. Transthoracic subcostal view (a) and transesophageal (b) views show the IVC is nearly filled by a tumor which extends into the RA.

Transesophageal echocardiography revealed a large, serpiginous, freely mobile mass extending into the right atrium from the dilated inferior vena cava. The mass extended from the inferior vena cava and crossed the tricuspid valve into the right ventricle. Another smaller semi-mobile mass was attached to the right ventricular outflow tract (Fig. 4). Main pulmonary artery and left and

right pulmonary arteries were dilated but no mass could be seen within the vessel lumens.



a



b

Fig. 4. Transesophageal views of RA (a) and RVOT (b) masses.

A CT scan of the thorax and pelvis showed a filling defect in the right atrium due to a large mass with extension into the IVC, a small filling defect in the right ventricle, thrombus formation in the IVC and iliac veins, and multiple small pelvic masses in the left

paravesical region in favor of uterine fibroma (Fig. 5).

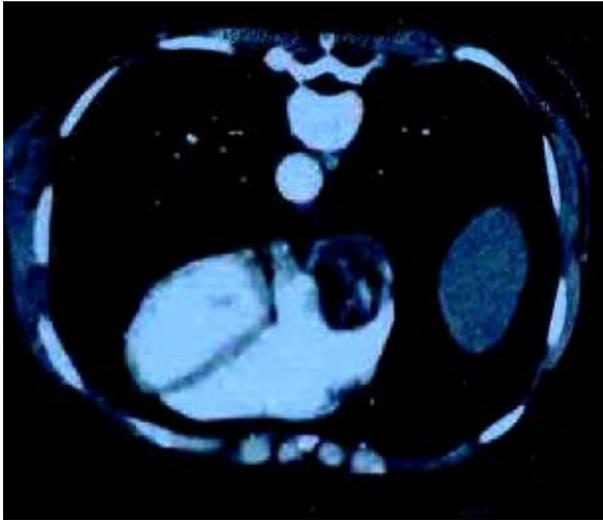


Fig. 5: Thoracic CT scan shows filling defect within RA cavity.

Treatment with tamoxifen (150 mg bid) and decapeptide (6 injections every 2 weeks) was prescribed. The patient was followed after 6 months. Transthoracic and transesophageal echocardiography showed mild decrease in right atrial mass but no change in the right ventricular mass. The patient was referred for redo operation.

During surgery, both cardiac masses and IVC tumor were resected (Figs. 6, 7).

Discussion

Intracardiac leiomyomatosis is a rare condition that generally occurs in women aged 28 to 80 years; most patients are middle-aged (median age: 44 years) women.¹

The patients often have a history of hysterectomy, as in our case, or may have symptoms due to uterine fibroids. Cardiac involvement presents typically with right-sided congestive symptoms. However, other presentations include syncope due to obstruction at the tricuspid valve^{5,6} and right-sided murmurs and clicks suggesting "tricuspid valve disease".^{7,8}

Rarer manifestations that have been reported include a high-output state,⁹ secondary thrombosis with Budd-Chiari syndrome,¹⁰ massive ascites,¹¹ sudden death,¹² and systemic embolism.¹³



a



b

Fig. 6. Post-op TTE (a) and TEE (b) show complete resection of RA and RV masses.



Fig. 7. TTE subcostal view shows complete resection of IVC mass.

Classically, the gross appearance of intravenous leiomyomatosis is a complex coiled or nodular growth within the myometrium with convoluted worm-like extensions into the uterine veins, broad ligament, or other pelvic organs.¹ Microscopically, most cases of intravenous leiomyomatosis are similar to leiomyomatosis but often have prominent zones of hyalinization obscuring the underlying smooth muscle nature of the neoplasm. Any smooth muscle differentiation that occurs in leiomyomatosis may be present in intravenous leiomyomatosis, including epitheloid differentiation.¹⁴⁻¹⁶ Metastasis to lungs and lymph nodes has been reported, and pulmonary nodules have been described.¹⁶ The reported two-dimensional¹⁷ and transesophageal¹⁸ echocardiographic features of intracardiac leiomyomatosis are limited. Although intravenous leiomyomatosis is considered "benign", encroachment of the cardiac chambers is almost malignant, given its potential for damaging valve apparatus, spreading into the pulmonary vasculature, and embolization. The most important condition in the differential diagnosis is right-sided heart thrombus-in-transit, which appears as

elongated mobile masses of venous casts, giving a "popcorn" appearance within the cardiac chambers.² Intracardiac leiomyomatosis is typified by a subacute presentation in a woman who has symptomatic uterine fibroids, often requiring hysterectomy. As opposed to thrombus, intracardiac leiomyomatosis is associated with multiple points of attachment in the setting of a postoperative state, indwelling central lines, or chronic debility. Other tumors such as renal cell carcinoma and hepatomas may extend into the right-sided cardiac chambers via the inferior vena cava. However, those tumors rarely have multiple endocardial attachments and have a mass-like appearance rather than that of a venous cast of long, thin, mobile structures.

Tumor removal may necessitate sternotomy as well as laparotomy.^{5,19} If the tumor is extensive, a two-stage operation may be needed.²⁰ Tamoxifen or megestrol acetate has been used to suppress tumor growth and promote tumor involution.²¹ Radiation and chemotherapy, including oral etoposide, have been used as adjunctive treatments.²² Recurrence after the surgical removal of the cardiac tumor, as in our patient, is not unusual and can occur up to 15 years after operation.¹⁸ Echocardiography may be useful for detecting cardiac recurrence and monitoring tumor growth.¹⁹

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