Intravenous Leiomyomatosis with Intracardiac Extension

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Abstract

Whereas uterine leiomyoma is a common woman disease, intravenous leiomyomatosis with intracaval and intracardiac complications is a rare condition. The initial presentation is dependent upon the severity of the intracardiac involvement, although complete surgery is the best treatment. The case of a 39-year-old woman is described here, with an initial presentation of dyspnea and right heart failure. Leiomyomatosis originating from the uterus and extending to the inferior vena cava and right atrium was diagnosed from various preoperative studies. The patient was resuscitated because of respiratory failure and severe right heart failure. However, she was operated successfully through a two-stage approach and remained well postoperatively. This case illustrates an intriguing presentation of intravenous leiomyomatosis and a curative surgical intervention even in critical condition.

Key words: leiomyomatosis, intravenous, intracardiac, heart failure, surgery

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Introduction

Intravascular leiomyomatosis (IVL) is a rarely described smooth muscle cell tumor that grows within venous channels but does not invade the tissues. The tumor primarily extends through the uterine veins, sometimes as far as the inferior vena cava (IVC), reaching the right-side cardiac chambers and the main pulmonary artery, resulting in death (1). IVL has been diagnosed after abnormal uterine bleeding or lower abdominal tenderness has been observed. Even with extensive intravenous tumor extension, patients may remain asymptomatic until intracardiac growth leads to cardiac insufficiency, pulmonary embolism, or even sudden cardiac death. Nevertheless, because of its rarity, the tumor is occasionally misdiagnosed or diagnosed late, and subsequently improperly treated. Correct diagnosis relies on a huge index of suspicion. Here, we present an IVL case in which the intravenous leiomyoma had extended deep into the right ventricle. The patient developed a critical condition with respiratory failure along with severe right heart failure and finally was cured by surgical removal of the tumor. We also review this potentially lethal disease.

Case Report

A 39-year-old multiparous Chinese woman was admitted to our institution following two months of intermittent chest tightness and one week of progressive dyspnea. She had previously undergone a subtotal hysterectomy for uterine myomatosis in 2006 and was admitted in June 2007 after several episodes of dyspnea over the week before her admission. The patient had no significant history of chest pain or alcohol or drug abuse. She denied any significant travel history or prior parasitic infection.

On physical examination the patient had a temperature of 37.0°C with a respiratory rate of 22 breaths/minute, blood pressure of 140/76 mmHg, and a pulse rate of 110 bpm. Except for 2+ bilateral pedal edema and a grade 2/6 systolic murmur at the left lower sternal border, the remainder of the physical examination was unremarkable. The chest radiograph showed mild cardiomegaly and mild pulmonary vas-

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Figure 1. Transthoracic echocardiography demonstrated a mass extending from the right atrium to the right ventricle through the tricuspid valve. RV, right ventricle; RA, right atrium; T, tumor.

Figure 2. Computed tomography showed a hypodense mass emerging from the left iliac vein to the inferior vena cava and then to the right atrium (arrows).

cular congestion. With the exception of sinus tachycardia, the results of the 12-lead electrocardiography (ECG) appeared normal. Her serum electrolytes and hematologic profile were within normal limits. However, the patient had an erythrocyte sedimentation rate of 36 and an alkaline phosphatase level of 128 U/L with an aspartate aminotransferase level of 76 U/L. An echocardiogram showed a mass occupying the right ventricle and right atrium, some of which could be seen extending into the IVC (Fig. 1). A computed tomographic scan showed a hypodense mass emerging from the left iliac vein, which had invaded the IVC (Fig. 2). A lobulated tumor in the pelvis was also noted that extended to the left pelvic wall. During the admission, her congestive symptoms worsened and endotracheal intubation was applied following hypoxic respiratory failure. Even 100% O₂ was supplied for the serious status. Jugular vein engorgement was found physically and a bedside cardiac echocardiography revealed right ventricular dilatation in addition to hypokinesis together with severe tricuspid regurgitation and dilatation of the pulmonary artery. The lab examination revealed disseminated intravascular coagulopathy with severe thrombocytopenia (Table 1). An emergency operation was arranged despite the patient’s serious condition.

First, a median sternotomy was performed. A cardiopulmonary bypass was then instituted between the ascending aorta, the superior vena cava and right femoral vein, followed by a right atriotomy and longitudinal venotomy. The intracardiac mass was clearly free-floating without involvement of the cardiac structure, whereas the intracaval mass was extensively attached to the IVC wall. The tumor was extracted from the right atrium and the IVC as far as the suprarenal level. Tricuspid valve repair and patent foramen ovale closure were also performed. A whitish stromal tumor measured approximately 46 cm in length was removed (Fig. 3). The postoperative recovery was uncomplicated. Her thrombocytopenia turned to normal gradually and extubation was carried out two days after the operation. The patient was discharged 10 days later. In October 2007, a second-stage operation involving a laparotomy was performed. A large, convoluted tumor mass was seen arising from the uterus. It was observed intraoperatively that the tumor growth extended into the left gonadal vein, growing along the left renal vein and finally reaching the IVC. Total hysterectomy and bilateral salpingo-oophorectomy was done. A venotomy and the complete excision of the lobulated, grey-white rubbery tumor were performed. The histology revealed that it consisted of proliferating smooth muscle fibers without any abnormal mitotic activity, consistent with IVL (Fig. 4). The whole course of treatment is summarized in Fig. 5.

Discussion

IVL is a rare uterine tumor characterized by a nodular mass of benign smooth muscle growing within the lumen of veins. At least 77 of the IVL cases reported between 1980 and 2008 had cardiac complications, including the present case (Table 2) (1-5). Out of these, 44 had undergone hysterectomies. We presume that in those cases, IVL had simply not been diagnosed in the period of time immediately following the hysterectomy. A tumor left inside the pelvic veins at the time of hysterectomy could have extended towards the right heart, leading to obstruction or other adverse events later on in life. The median interval between hysterectomy to the diagnosis of IVL with cardiac involvement was 4 years (6). However, the time lag in the present case was only one year. It has been reported that differences in cytological features only correlate with the time period of recurrence, and not with the IVL clinical outcome (7). We

Table 1. Lab Examination Revealed Disseminated Intravascular Coagulopathy with Severe Thrombocytopenia

<table>
<thead>
<tr>
<th>Peripheral blood</th>
<th>Coagulation studies</th>
<th>(normal values)</th>
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<tbody>
<tr>
<td>WBC 10,730 (μL⁻¹)</td>
<td>PT 20.3 (s)</td>
<td>(11.5-14.0)</td>
</tr>
<tr>
<td>RBC 404 × 10⁴ (μL⁻¹)</td>
<td>APTT 49.5 (s)</td>
<td>(30.0-42.0)</td>
</tr>
<tr>
<td>Fb 12.1 g/dL</td>
<td>Fibrinogen 67 (mg/dL⁻¹)</td>
<td>(180-400)</td>
</tr>
<tr>
<td>PLT 1.5 × 10⁴ (μL⁻¹)</td>
<td>FDP 394 (μg/dL⁻¹)</td>
<td>(&lt;5.0)</td>
</tr>
<tr>
<td>D-dimer</td>
<td>389 (μg/dL⁻¹)</td>
<td>(&lt;1.0)</td>
</tr>
<tr>
<td>ATIII 74%</td>
<td>&gt;82%</td>
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Figure 3. The 46-cm-long grey-white stromal tumor mass removed from the inferior vena cava.

believe that there should be more vigilance against recurrence in such rapidly growing cases.

In our review, the most common presenting symptom is heart failure. Most patients with IVL remain asymptomatic until direct intracardiac extension causes valve obstruction or cavity compromise, leading to cardiac insufficiency. Death due to heart failure caused by mechanical obstruction and/or intracardiac extension of these lesions has also been reported (8). Most cases did not experience serious complications at the time of diagnosis. However, the present patient developed both acute hypoxemia and disseminated intravascular coagulopathy. Tumor-induced venous thromboembolism was suspected. We supposed that once imaged, rapid tumor excision may decrease the incidence of this serious complication and further improve survival rates in IVL patients.

Various imaging modalities are available for the detection and diagnosis of IVL. By 2-dimensional echocardiography, the presence of a long, serpentine, polypoidal mass extending from the IVC into the right atrium should raise the suspicion of IVL (9). The IVL usually also presents echocardiographic features of elongated mobile masses that extend from the veins of the lower body (10). Transesophageal echocardiography (TEE) provides better spatial resolution as to where the tumor is attached to the atrial wall or interatrial septum, and also the relationship with the IVC and SVC (11). Identifying the site of attachment also helps in differentiating IVL from myxoma, because right atrial myxoma is characteristically highly mobile and frequently attached to the interatrial septum, although it may also adhere to the wall of the right atrium (12). Contrast-enhanced CT can reveal the tumoral thrombus within the IVC and its extension to renal, iliac or adnexal veins (11). MRI is superior to CT and conventional venography because, not only does it avoid irradiation, the contrast agent used (gadolinium chelate) is much safer than the iodinated contrast used in CT scanning and conventional venography. MRI also has better soft tissue contrast resolution and can assess blood flow without contrast injection (13). The MRI features of IVL are nonspecific, and appear iso- or mildly hyperintense with respect to the muscle on the T1-weighted images, although the signal intensity is much higher on T2-weighted images (especially after the intravenous injection of gadolinium) (13, 14). Surgery is the treatment of choice for IVL, although it does require the complete removal of the tumor, since incomplete excision may result in a recurrence and hence further surgery or even death. Bilateral oophorectomy is also suggested because the tumor is estrogen dependent (15). In our review, a higher percentage of patients achieved a complete removal...
of tumor, and of those, most were operated on by a two-stage approach. In the present case, we adopted a two-stage approach because single-stage surgery requires a longer operative time which is not appropriate for such a critical condition. In addition, the correct diagnosis was made after the first stage operation. Fast recovery was achieved in our case although multi-organ failure was diagnosed preoperatively. This further confirmed that immediate surgery is still the first choice for treating critically ill patients, although anti-estrogen therapy had also been proposed.

In conclusion, intravenous leiomyomatosis should be suspected in young women who have cardiac symptoms and are carrying a right-sided intracardiac mass. Surgical resection is the best treatment even in the most serious conditions and should be performed as soon as possible, in order to reduce the risk of sudden death and concomitant venous thromboembolism.
References


