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Successful One-Stage Operative Resection of an Intravenous Leiomyomatosis Involving the Right-Sided Heart Chambers and Pulmonary Artery

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ABSTRACT

Intravenous leiomyomatosis (IVL) is a rare neoplastic disorder. It may extend into the right cardiac chambers, which are named after intracardiac leiomyomatosis (ICL). We describe a case of IVL extending into the right heart and pulmonary artery in a 46-year-old woman. The patient successfully underwent a one-stage operation under cardiopulmonary bypass (CPB) with deep hypothermic circulatory arrest. The patient fully recovered without major complications and did not exhibit any recurrence at the 9-month follow-up. The unique feature of this case is its involvement with the pulmonary artery, which is uncommon but potentially lethal.

INTRODUCTION

Intravenous leiomyomatosis (IVL) is rare, and intracardiac leiomyomatosis (ICL) even rarer. The first case of IVL was described by Birch-Hirschfeld in 1896 [Birch-Hirschfeld 1896]. Currently, approximately 300 cases of IVL have been reported and the number of ICL cases, initially reported by Durck in 1907, totals more than 100 [Durck 1907; Zhang 2010]. We describe a case of IVL involving the right heart and pulmonary artery.

CASE REPORT

A 46-year-old female with a history of cesarean section presented complaining of recurrent syncopal attacks for 4 months. She denied any other symptoms related to vascular diseases or nervous system diseases. The physical examination and laboratory examinations showed no abnormal findings. On the work-up for repeated syncope, transthoracic echocardiography exhibited a mobile mass that originated from the inferior vena cava (IVC), and extended into the right atrium, right ventricular and pulmonary artery. Gynecological ultrasound revealed multiple uterine fibroids. A solid mass, approximately 64 × 56 mm in size, was observed in the left adnexa. Lower extremity venous ultrasound showed a space-occupying lesion at the proximal end of right common iliac vein and external iliac vein. A contrast-enhanced CT scan (Figure 1) showed multiple lesions in the uterus and bilateral adnexa. In addition, a continuous filling defect was noted to spread along IVC via bilateral ovarian vein and right renal vein, and then grow into the right heart and pulmonary artery. In an ultrasound of the IVC, a hypoechoic cord-like mass was noticed and measured 0.9 cm at the greatest diameter. Transvaginal ultrasound demonstrated the presence of a pelvic mass, multiple lesions in parauterine vessels, and irregular low echo in the front uterine wall. Computed tomography venography and computed tomographic pulmonary angiography (Figure 2) showed a continuous mass in the pulmonary artery, right heart, IVC, right renal vein, bilateral ovarian veins, and right common iliac vein. These examinations suggested the diagnosis of IVL.

The patient was diagnosed with IVL involving the right-sided heart and pulmonary artery. After a multidisciplinary discussion, surgical resection was deemed mandatory. The patient underwent myomatectomy, hysterectomy, and bilateral adnexectomy under cardiopulmonary bypass (CPB) with deep hypothermic circulatory arrest and laparotomy in a one-stage operation. Intraoperative transesophageal

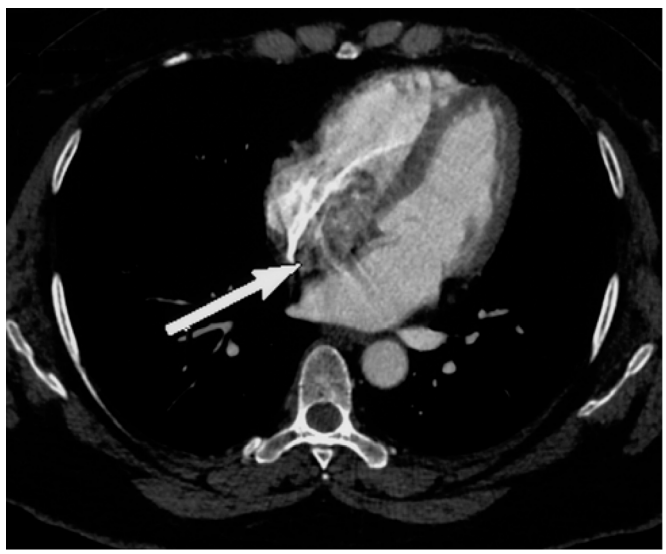

Figure 1. Contrast-enhanced CT scan shows right cardiac chambers had filling defect (arrow).

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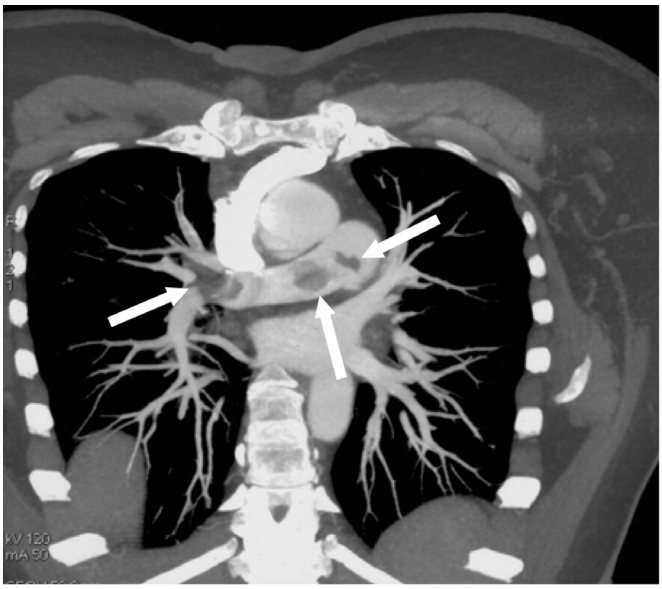

Figure 2. CT pulmonary angiogram shows tumor embolus in the pulmonary artery (arrow).

echocardiography confirmed the existence of a mass arising from the IVC migrating into the right heart and pulmonary artery. First, the abdomen was incised. The mass was found to have extended into the IVC via the right ovarian vein. The mass was divided into two parts by a surgical procedure at the site where it invaded the right ovarian vein. Second, total hysterectomy with bilateral adnexectomy, including the excision of the tumor, was performed. Subsequently, infrahrenic vena cava was successfully isolated and a median sternotomy was performed. After the heparinization (3 mg/kg), CPB was set up with a venous cannula in the superior and inferior vena cava and an aortic cannula in the ascending aorta. The right atrium was opened under deep hypothermic circulatory arrest. The mass was detected to have well-demarcated borders with cardiac structure and IVC. Immediately, the IVC was incised via a venotomy. Once the tumor was exposed, it was then resected. Intracardiac and intracaval masses were separately extracted and removed. During this course, no adhesion was discovered. Next, CPB was reestablished and rewarming was initiated. Finally, we excised the tumor in the main pulmonary artery and right pulmonary artery. The left pulmonary artery was carefully explored but no mass was found. The total CPB time was 63 minutes, aortic cross clamp time was 40 minutes, and circulatory arrest time was 15 minutes.

The resected specimen (Figure 3) showed a 40-cm-long gray-white tumor that was solid and elastic, without any tumor thrombi or areas of friable tissue. IVL was definitively diagnosed by the pathological examination, which suggested smooth muscle cell proliferation. The patient fully recovered without major complications and was discharged within a week. On 9-month follow-up, no symptomatic discomfort was experienced and the patient did not exhibit any recurrence.

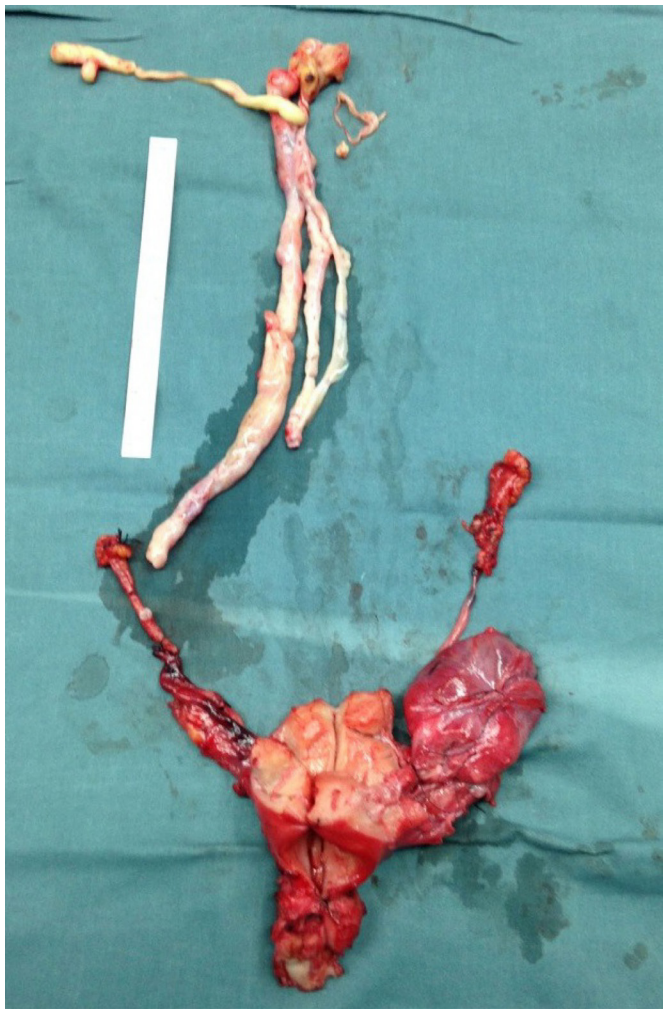

Figure 3. Resected specimen of the uterus, bilateral adnexa, tumor embolus in bilateral ovarian vein, IVC, right cardiac chambers, and pulmonary artery.

DISCUSSION

IVL is an uncommon neoplasm, which is frequently ignored, misdiagnosed, or improperly treated. It is characterized by overgrowth of benign smooth muscle that may behave in a malignant way, with not only potential involvement of pelvic veins, systemic veins, IVC, right heart chamber and pulmonary artery, but also potential risk of distant metastasis [Xu 2013]. As for the pathogenesis of IVL, two main hypotheses have been proposed: the first is that it might arise from uterine leiomyomas, while the other indicates that the vein wall, as the primary tumor, causes intravascular projections into a neighboring venous lumen [Gaudino 2002]. In our case, we observed that IVL stemmed from the uterine leiomyoma and extended into vascular lumen, which may support the first hypothesis. In most cases, the usual extension pathway is that the tumor enters via the iliac vein and migrates into the IVC. Nonetheless, in this case, the right ovarian vein functioned as a specific route to the IVC.

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IVL predominantly occurs in women of childbearing age after hysterectomy or concurrently with uterine leiomyoma. Clinical manifestations of this tumor are atypical and mainly depend on its location and scale. Even though IVL may reach as far as the IVC, the patient might still be asymptomatic unless cardiac dysfunction develops due to intracardiac extension. The distal intravascular extension of IVL could lead to various symptoms, such as shortness of breath, syncope, abdominal pain, palpitation, edema of the lower extremities, and so on [Li 2013]. Sometimes it can cause pulmonary embolism, Budd-Chiari syndrome, or even sudden death. In this case, women mainly complained of recurrent syncopal attacks. The reason for misdiagnosis may be because it is an atypical symptom for IVL diagnosis.

To date, although the choice between one-stage operation and two-stage operation is controversial, surgical operation is still the optimal treatment for IVL and ICL [Wang 2012]. The surgical strategy should be carefully considered by a multidisciplinary discussion in order to completely resect the neoplasm, because incomplete excision seems to cause tumor recurrence. A tumor may be completely removed during a one-stage operation; however, it may bring a much higher risk to the patient. So we conclude that a one-stage operation should be performed only when the patient's clinical condition is good. Otherwise, a two-stage operation should be considered. In our case, the patient's clinical condition was relatively good, and, therefore, a one-stage operation was performed and a positive result was obtained.

In conclusion, we performed a one-stage operation in a patient with IVL involving the right heart chambers and pulmonary artery, the postoperative course was uneventful, and the patient did not exhibit any recurrence at the 9-month follow-up.

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