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One-Stage Surgical Removal of Intravenous Leiomyomatosis with Right Heart Extension is Safe

Qiang Li,[1] Jie Ma,[1] Bin Hao,[2] Xintao Pi,[2] Hao Li[1]

Departments of [1]Cardiothoracic Surgery and [2]Vascular Surgery, Second Hospital of Shanxi Medical University, Taiyuan, People's Republic of China

ABSTRACT

Intravenous leiomyomatosis is a rare smooth muscle tumor. We report the case of a 42-year-old woman with both intravenous and intracardiac extension of leiomyomatosis who underwent 3 operations within 9 years. During the last admission, she underwent a successful single-stage surgical approach while under cardiopulmonary bypass with circulatory arrest. A postoperative histopathologic examination of the resected specimen confirmed the diagnosis.

INTRODUCTION

Intravascular leiomyomatosis (IVL) is a rare benign vascular tumor that grows within venous channels but does not invade tissues. The tumor extends primarily through the uterine veins and sometimes reaches as far as the inferior vena cava (IVC). From there, tumor tissue can enter the right-side cardiac chambers and the pulmonary arteries, a stage of progression that most commonly causes death [Koh 2000]. This disease was first described in 1896 [Birch-Hirschfeld 1896]. We report a case of IVL with intracardiac extension that was successfully treated with a single-stage surgical approach.

CASE REPORT

A 42-year-old multiparous Chinese woman was admitted to our hospital following 9 months of palpitations and hypodynamia on exertion. A hysterectomy had been performed 9 years previously because of the presence of uterine leiomyoma. A right-sided oophorectomy was carried out 2 years after the hysterectomy because of the presence of an ovarian cyst, but the details of these procedures were not available at the time of her third admission. She had previously been in excellent health before the onset of the shortness of breath. She was a nonsmoker with no history of a hypercoagulable state, deep vein thrombosis, weight loss, or cancer.

On examination, the patient's lungs were clear, but a systolic ejection murmur was present on auscultation (Levine 2/6) over the third left sternal border. No edema was present in the extremities. The results of the blood count and other laboratory investigations were normal. An electrocardiogram showed a normal sinus rhythm, and chest x-rays revealed no cardiomegaly or abnormal shadows in the lung fields. No other symptoms were observed.

Magnetic resonance imaging (MRI) (Figure 1) demonstrated extension of the uterine leiomyoma into the right internal iliac and ovarian veins with progression into the IVC to the level of the right atrium (RA).

The patient was taken to the operation theater, and a simultaneous operation was performed, with a median sternotomy

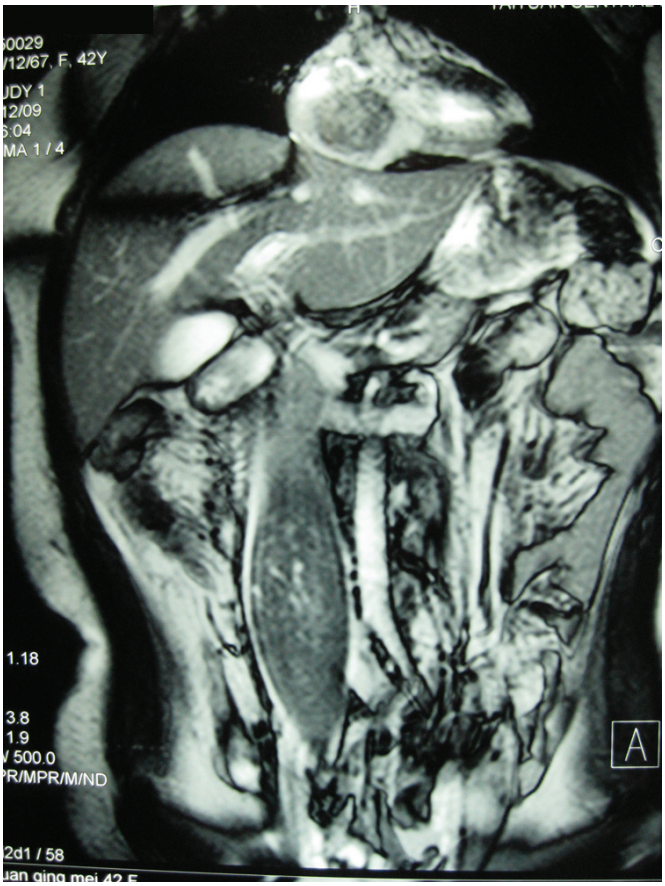

Figure 1. Sagittal and coronal magnetic resonance images. The tumor can be seen to extend from the right atrium to the inferior vena cava.

Qiang Li and Jie Ma contributed equally to this work.

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Correspondence: Jie Ma, Department of Cardiothoracic Surgery, Second Affiliated Hospital, Shanxi Medical University, Taiyuan 030001, People's Republic of China; 86-351-3365106 (e-mail: lee.q@126.com).

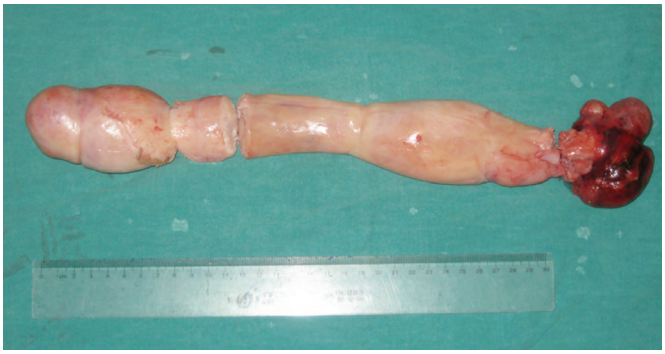

Figure 2. Tumor tissue excised en bloc from the right atrium and intravenous tumor specimen.

and a median abdominal approach used to access the right common iliac vein. Cardiopulmonary bypass (CPB) was instituted with cannulae through the superior vena cava and the ascending aorta. Then, the RA was opened during core cooling to 28°C with circulatory arrest. There was a single yellowish tumor in the RA that measured approximately 5 cm in diameter. No visible adhesions connected the tumor to the atrial septum or the tricuspid valve. With the patient under total circulatory arrest, the intracardiac tumor was removed from the RA, and a cannula was inserted into the IVC through an incision in the RA. Flow resumed through the ascending aorta, and the heart was perfused again, after which it recovered a normal sinus rhythm.

The right common iliac vein was then incised. The main body of tumor within the IVC was rubbery in consistency, and although it was adherent to the intimal layer of the vessel, it did not infiltrate the venous wall. The tumor was easily removed with the use of gentle traction. Despite these efforts, the portion of the tumor that originated from the right internal iliac vein remained adherent to the vein by a loose adhesion, which was excised (Figure 2). The internal iliac venotomy was closed with a 5-0 polypropylene suture. The CPB machine was stopped, and the extracorporeal circulation tubing set was removed according to standard practice.

The patient tolerated the procedure well and was discharged home on the sixth postoperative day. Hormonal therapy was not selected because of the patient's low levels of serum estrogen. She continues to do well after a 6-month follow-up period, with no residual tumor identified and no recurrent cardiac symptoms.

A pathology analysis confirmed the diagnosis of benign leiomyomatosis. Microscopically, IVL was characterized by endothelium-coated plugs of benign smooth muscle cells within myometrial vessels or the lymphatic system (Figure 3).

DISCUSSION

IVL is a rare, benign, well-differentiated smooth muscle tumor that originates from the uterus and infiltrates through the lumen of veins. Tumor extension mainly occurs via 2 different routes of venous channels [Lam 2004]. In the first route, the tumor extends into the uterine vein, the internal iliac vein,

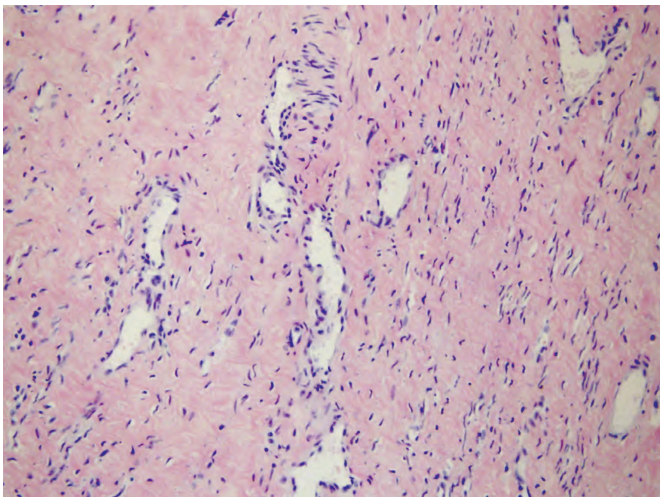

Figure 3. Pathology specimen from the tumor in the right atrium (hematoxylin and eosin staining, original magnification ×200).

the common iliac vein, and then the IVC. In the second route, the tumor extends into the ovarian vein, the renal vein, and then the IVC. The complexity of achieving a cure in cases of this complex disease emphasizes the need for accurate preoperative delineation of the tumor's extension and a planned systemic approach by a multidisciplinary surgical team.

The diagnosis of IVL is relatively easy to make when the connection between the intravenous mass and the uterus is visualized. Given that most of the symptoms relate to the cardiovascular system, echocardiography and electrocardiography evaluations are frequently performed as the initial studies. When an RA mass is identified, diagnostic studies should also include MRI, venography, and computed tomography scans of the chest, abdomen, and pelvis in order to delineate the extent and attachments of the tumor. MRI is a useful tool to assure the diagnosis of IVL. MRI is particularly helpful for the assessment of patients with suspected intravascular lesions because of its multiplanar capabilities, better soft-tissue contrast resolution, and its ability to assess blood flow without the injection of contrast. The MRI features of IVL are nonspecific, and the tumor can appear iso- or mildly hyperintense with respect to smooth muscle on T1-weighted images, although the signal intensity is much higher on T2-weighted images. This difference is particularly true after the intravenous injection of gadolinium [Hayasaka 2000]. MRI has been shown to be superior to computed tomography imaging as a tool to confirm the diagnosis of IVL [Ahmed 2004].

To date, there have been at least 200 case reports of IVL with intracardiac extension, although the tumor is usually confined to pelvic veins. Almost 60% of the cases have been reported in the last 20 years [Lam 2004]. This situation may be because the diagnosis has been made easier with the increasing knowledge of the disease entity; however, the number of IVL cases most likely remains underestimated because the diagnosis can still be easily missed. When tumor extension remains inside the small vessels of the myometrium, it cannot be detected, even with preoperative imaging. The correct

diagnosis therefore depends on a high index of suspicion, particularly in the early stage of the diagnostic process.

Surgery is the treatment of choice, and complete removal of the tumor is mandatory to avoid recurrence, even without clinical signs of venous hypertension. The first successful total resection was reported in 1982 [Ariza 1982]. Surgery can be performed with a single- or double-stage procedure. Some surgeons prefer the latter because single-stage surgery requires a long operative time and carries an increased risk of bleeding because of the systemic heparinization required for CPB [Gaudino 2002]. In addition, the frequent attachment of tumor to the IVC wall precludes tumor extraction through the RA, which can cause fatal retroperitoneal hemorrhage, and any use of a single abdominal approach to resect a tumor that generally hugs the cardiac cavities would also be dangerous. In addition, mobilization and occlusion of the IVC may cause a hemodynamic compromise that may require active or passive veno-veno bypass to be established [Lam 2004].

Many surgeons, however, have supported the single-stage procedure [Ma 2007]. With the development of the realization of IVC bionomics and operative and CPB techniques, hemorrhage and IVC occlusion are no longer the main difficulties with this type of procedure. At the origin of the IVC, adhesions can occur between the tumor and the vessel wall. At the distal end of the IVC, adhesions can occur at the inlet of the IVC to the RA because of the impact of turbulent blood flow on the tumor. Adhesions have been found to occur only at the proximal and distal ends of the IVC, which means that no further incisions are required along the length of the IVC. Therefore, the operation time becomes much shorter, and a single-stage surgery can avoid a second general anesthesia. There are 2 ways to shorten the CPB and total operation times. One is that used with the patient we have described. In the second way, the origin of the tumor is resected initially from the right internal iliac vein through a lower-abdominal incision. CPB is then instituted, and the main body and distal end of the tumor can then be easily removed via an incision in the RA.

In conclusion, IVL is a smooth muscle cell tumor that may grow within veins along several routes. An accurate preoperative description of the tumor's extension is vital for the success of any attempt at complete tumor excision. A planned systemic approach by a multidisciplinary surgical team is essential to improve the chances of curative surgery for IVL. It is our view that a single-stage procedure is safe in these patients, because no adhesions form in the middle section of the IVC.

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