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Case Report

Intravenous Leiomyomatosis with Intracardiac Extension as a Rare Cause of Abdominal Pain in an Adult Patient: A Case Report

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Abstract

Intravenous leiomyomatosis (IVL) is a rare and benign smooth muscle tumor that arises from intrauterine venules or the myometrium. We herein describe a 49-year-old woman with a history of myomectomy who developed abdominal pain. An intravascular mass with extension to the right atrium was detected in the inferior vena cava. The mass was surgically resected in a single stage under cardiopulmonary bypass. IVL features were indicated by subsequent histopathology. Postoperatively, the patient was diagnosed with massive pericardial effusion and treated with a pericardial window. At 3 months' outpatient clinical follow-up, she was asymptomatic. This case indicates that the diagnosis of IVL with extension to the heart should be kept in mind in patients presenting with abdominal pain.

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Introduction

Primary cardiac tumors are generally rare and predominantly benign.[1] Primary cardiac leiomyomas are even rarer, and most of them are secondary to tumor metastasis or continuity to the uterine leiomyoma.[1] Intravenous leiomyomatosis (IVL) is defined as a benign smooth muscle tumor arising from the intrauterine venules and the myometrium, with identifiable growth within the lumens of veins.[2] The tumor sometimes reaches as far as the hepatic, cardiac, or pulmonary vasculature, leading to severe complications such as pulmonary embolism or signs of right heart failure.[3] Clinical manifestations include respiratory and cardiac symptoms or rarely abdominal pain

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as a late-onset finding. We describe the single-stage surgical management of a rare IVL late after a uterine myomectomy as an intracardiac mass.

Case Report

A 49-year-old woman came to our emergency room with a 2-month history of right upper abdominal pain. The patient had a past medical history of myomectomy for symptomatic uterine leiomyoma 8 years earlier. Upon physical examination, there was no evidence of congestive heart failure. During admission, her general physical examination and laboratory investigations (eg, liver function tests, renal function tests, and complete blood counts) were within normal limits. She underwent a contrast-enhanced computed tomogram (CCT) of her abdomen to exclude any abdominal pathology. A significant mass extending to the right side of the heart was observed in the CCT of her abdomen (Figure 1A-B). The mass extended from the gonadal vein to the cavoatrial junction. Both renal parenchymas were normal in her abdominal CCT. A mobile mass extending from the inferior vena cava (IVC) to the right atrium (RA) was visualized in her transthoracic echocardiogram. An operative decision was made because the mass was large, mobile, and almost completely occupied the RA, with the impending risk of sudden death due to tricuspid valve occlusion and massive pulmonary embolism. Normal coronary arteries were observed in the preoperative coronary angiography. The tumor was approached via a median sternotomy under cardiopulmonary bypass and moderate hypothermia. The median sternotomy was followed by the cannulation of the aorta, the superior vena cava, and the right femoral vein (conservative length until below the infrahepatic IVC). The patient was cooled down to 22 ºC. After the RA was opened, the intracardiac part of the tumor was seen freely floating. It snugly fitted in the RA–IVC junction, and it was freed by blunt finger dissection (Figure 2). The entire tumor was easily pulled out with little tension from the cardiac to the infrarenal IVC and gonadal vein extension. The RA was closed, and the patient was weaned off the cardiopulmonary bypass. Grossly, the tumor was firm, rubbery, gray-white, and 18×20 mm in diameter (Figure 3A). Spindle to stellate-shaped cells without any abnormal mitotic figures were revealed by histopathology (Figure 3B). The spindle cells actin (+), caldesmon (+), panck (−), myogenin (−), CD-34 (−), and S-100 (−) were shown in immunohistochemistry (Figure 3C–3D). The staining for estrogen and progesterone receptors was positive, suggesting a uterine origin. The patient's in-hospital course was uneventful after surgery, and she was discharged from the hospital on the 10th postoperative day. The patient complained of progressive dyspnea at a 1-month outpatient clinical follow-up. She was diagnosed with massive pericardial effusion, which was subsequently treated with a pericardial window. Afterward, she was asymptomatic at the 3-month outpatient clinical follow-up.

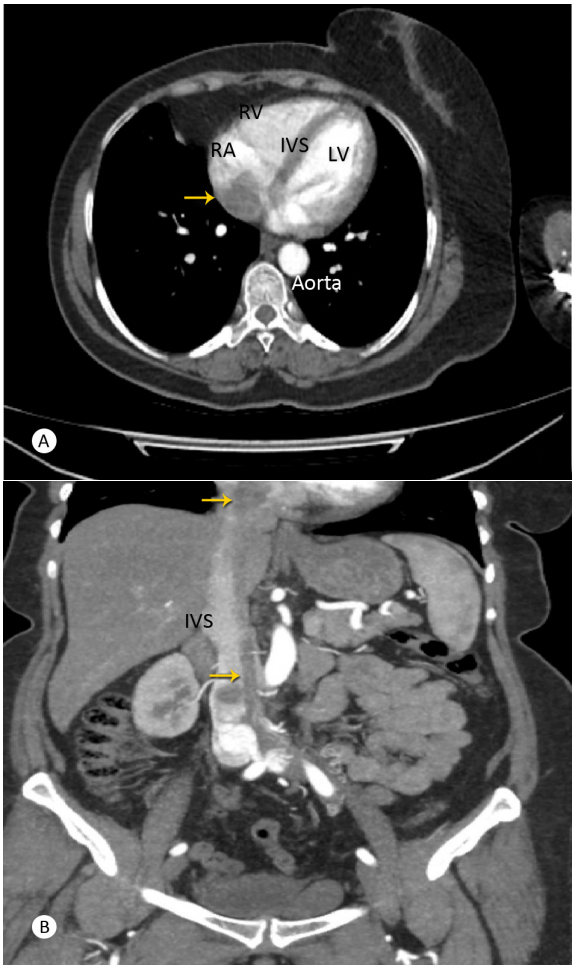

Figure 1. A) Contrast-enhanced computed tomogram of the abdomen image shows an intravenous leiomyomatosis extending into the RA.
B) Contrast-enhanced computed tomogram of the abdomen image shows an intravenous leiomyomatosis extending from the IVC into RA.
RA, Right atrium; RV, Right ventricle; IVS, Interventricular septum; LV, Left atrium; IVC, Inferior vena cava

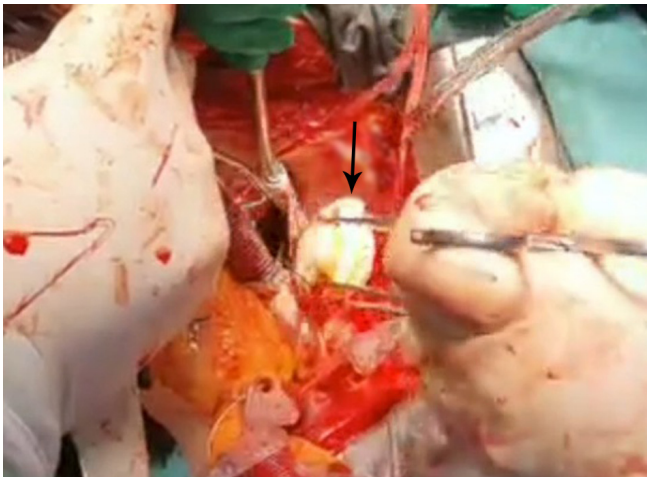

Figure 2. The intravenous leiomyomatosis, seen freely floating here, was subsequently freed by blunt finger dissection.

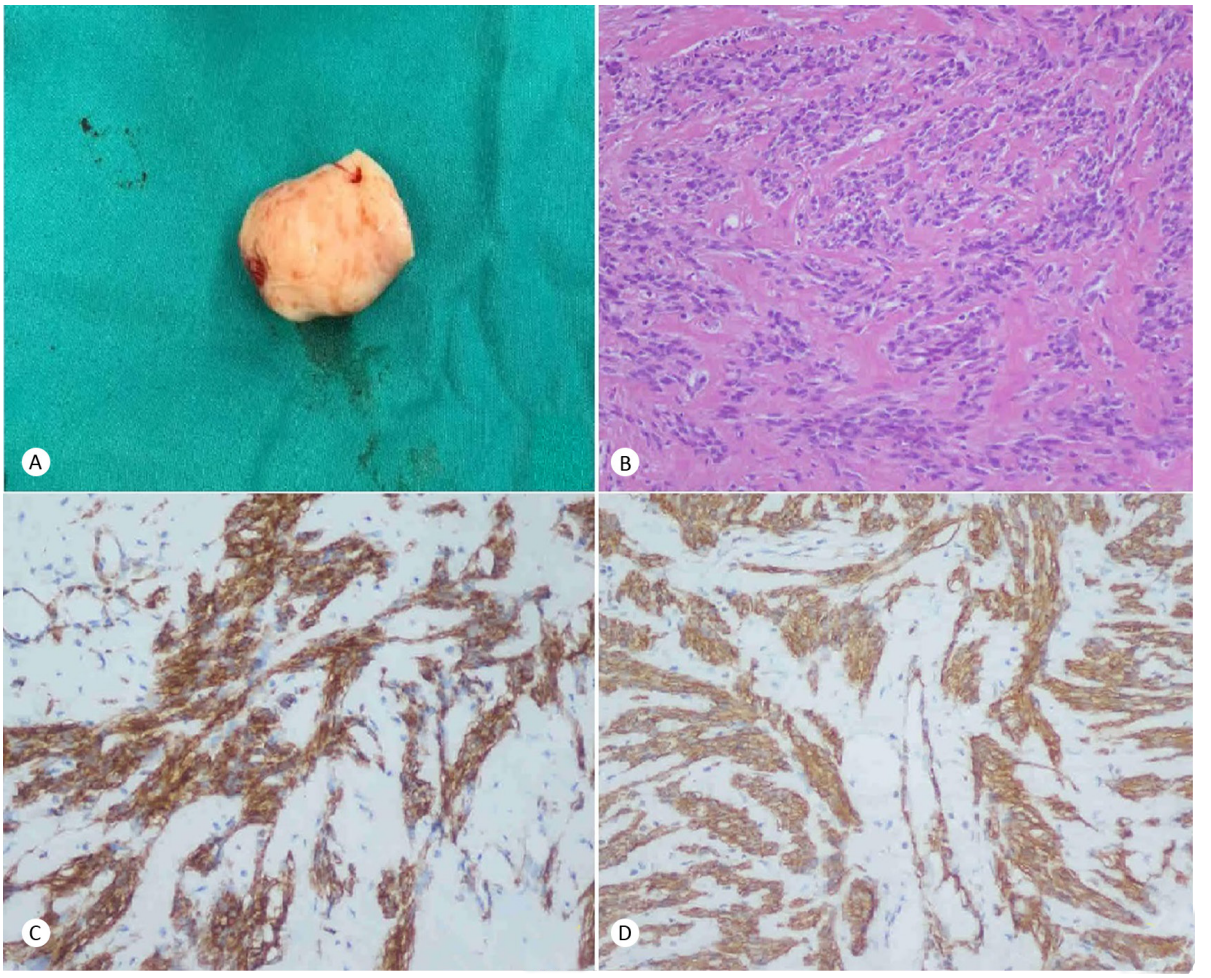

Figure 3. A) The gross pathology of the tumor is firm, rubbery, and gray-white, which is compatible with intravenous leiomyoma.
B) The hematoxylin stain (X200) of the tumor shows spindle to stellate-shaped cells without any abnormal mitotic figures, which is compatible with intravenous leiomyoma.
C) The immunohistochemistry staining (X200) of the tumor shows the presence of caldesmon, which is compatible with intravenous leiomyoma.
D) The immunohistochemistry staining (X200) of the intravenous leiomyoma shows the presence of actin, which is compatible with intravenous leiomyoma.

Discussion

The neoplastic smooth muscles of IVL are histologically and cytogenetically similar to benign leiomyomata. This tumor may behave in a "malignant" fashion in that not only might it involve the pelvic veins, the IVC, the renal veins, the pulmonary artery, and the right cardiac chamber but also it might have distant metastasis to the lung, brain, and lymph nodes. Cardiac involvement is seen in approximately 10% to 40% of cases.[1, 4] According to imaging features, the differential diagnosis of IVL mainly includes intravenous thrombi, leiomyosarcoma, right atrial myxoma, and malignant carcinoma of the renal, hepatic, and adrenal tissues.

The etiology of IVL is unclear; however, 2 theories have been proposed. The first theory suggests that IVL originates *de novo* from the vessel wall, whereas the second propounds intravenous extension from a preexisting uterine leiomyoma.[5] Our patient had a history of myomectomy 8 years previously. CCT imaging revealed an irregular enhancing tumor in the common iliac vein and within the IVC, extending to the RA. The mass had not adhered to the wall of the vascular structure and the heart, and it was resected completely. Therefore, these findings are inclined to support the second theory of pathogenesis.

The age at presentation of IVL is highly variable. Nonetheless, it most commonly affects women in the fifth decade of life. Clinical IVL characteristics mainly depend on the location and scale of the lesion. More distal intravascular extension of the tumor can result in various cardiorespiratory symptoms. Shortness of breath, palpitations, and lower extremity edema comprise the most common symptoms. Our patient's prominent clinical manifestation was abdominal pain. According to Kloska et al,[6] IVL may occasionally occur with acute pulmonary embolism. Moreover, it may be an unusual cause of a right ventricular mass in an adult patient.[7]

Based on our case findings, we can summarize IVL diagnostic points as follows:

1) Middle-aged women usually have a history of myomectomy and ovarian cyst surgery. 2) There may be an inhomogeneously enhanced irregular pelvic mass that invades the pelvic vein and extends to the IVC and sometimes the RA. 3) The lesion widely infringes the venous system is an essential diagnostic feature. 4) The mobile mass within the RA is always accompanied by a large mass in the IVC. 5) The mass in the heart and vein structures has no adhesion to the wall of the heart and the veins.

As is shown in our case, surgical excision is still the best treatment of choice for IVL treatment, and the complete removal of the tumor is considered essential to prevent a recurrence.[8] IVL has a high recurrence rate because a complete resection is difficult to achieve.[8]

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