SHORT REPORT

Recurrent Pelvic Intravenous Leiomyomatosis with Extension up the Inferior Vena Cava


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Abstract We present a case of a 43-year-old woman with recurrent pelvic intravenous leio-
myomatosis with extension up the inferior vena cava, who underwent three operations within
three years. In the last admission, she underwent surgery by a two-stage surgical approach and
postoperatively pathologic examination confirmed the diagnosis. We prefer to a two-stage
approach because of safe resection of the intracardiac tumor mass, shorter operative time
and less postoperative complication. Patients with intravenous leiomyomatosis need long-term
observation because of the high recurrence rate of the tumor.

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Introduction

Intravenous leiomyomatosis (IVL) is a rare uterine tumor
defined as "an intraluminal growth of benign smooth
muscle cells in either venous or lymphatic vessels outside
the confines of, or even in absence of, leiomyomas". Although histologically benign, IVL may be malignant in its
mode of behavior. IVL may extend into the right side of the
heart through the inferior vena cava (IVC) in 10% of

patients, which may mechanically obstruct the IVC, right
heart or the pulmonary artery, resulting occasionally in
death. Recurrence of IVL which extended into the IVC has
been described previously in only a few reports in the
English literature.1–3 We report a case of a 43-year-old
woman with recurrent pelvic IVL with extension up the IVC.

Case Report

A 43-year-old female with menorrhagia was diagnosed as
"uterine fibroids" by pelvic ultrasound and underwent
a subtotal hysterectomy in July 2005 for in local hospital.
Her postoperative course was uneventful. However, in
September 2005, she presented with complaints of
palpitation, dyspnea and short breath. Transthoracic

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echocardiography displayed that in the dilated right atrium and IVC, there was a large mass lesion, so she underwent an operation for a right atrial and the IVC mass in department of cardial surgery in our hospital and the histology showed IVL at that time.

In March 2008, after 2.5 years of symptom-free period, she was readmitted to our hospital with a mass in lower abdomen, dyspnea and short breath, and the symptoms mentioned above were aggravated after movement. She denied abdominal pain and precordialgia and bilateral leg edema. A CT venography for the heart and the major vessels displayed that in the dilated right atrium, there was a large mass lesion which extended through the IVC and almost completely filled it, and also reached the iliac veins and that a mass was also seen in the cavitas pelvis (Fig. 1).

As a result of these findings, in April 2008, she underwent surgery with the presumed diagnosis of IVL by a two-stage surgical approach. She underwent an operation for a right atrial mass at first and a mass in the IVC excision and bilateral salpingo-oophorectomy and mass in the pelvic sidewall excision were done 20 days later (Fig. 2). Post-operatively pathologic examination of the specimens obtained from the mass lesion in the inferior vena cava was reported as IVL. The masses were composed of benign proliferations of smooth muscle without significant cytologic atypia, coagulative necrosis, or excessive mitotic activity. The smooth muscle nature of these lesions was supported by positive desmin and a-SMA immunoreactivity. Focally there was some intrusion of smooth muscle into the vascular spaces, supported by CD34 immunostains. After the operation, the echocardiography and 64 multi-detector CT venography did not detect any residue. The patient has remained free of disease based on short-term followup.

Discussion

Intravenous leiomyomatosis generally affects women of reproductive age but may affect menopausal women.
Typical patients are middle-aged women and the majority of them have a history of hysterectomy or uterine myoma. Patients with IVL may present with the symptoms of uterine leiomyoma, i.e. pelvic pain and vaginal bleeding, respiratory symptoms (dyspnea, cough) in case of pulmonary metastasis, swelling of legs in case of inferior vena cava occlusion by the tumor, and symptoms of cardiac failure due to the tumor extension in the right cardiac cavities. It might provoke serious complications in cases of intracaval or intracardiac extension and can lead to sudden death. It must be emphasized that intravenous leiomyomatosis reaching from the pelvic veins to the right-sided heart, thus life-threatening, can recur within a relatively short time span and must therefore be closely observed following surgical resection.

This report also raises a question about the risk of recurrent IVL. Although extremely rare, leiomyomatosis may present recurrences are described in our case. According to similar reports on IVL with extension up the IVC, the rate of recurrences is possibly 30% and might be underestimated by most vascular surgeons. When recurrence is seen, reintervention is actually universally recommended to achieve long-term disease-free survival. So completely resection of the tumor is the key of the prevention of recurrence. Thus patients should be followed regularly because of the high recurrence rate of the tumor.

Surgery remains the gold standard of treatment; complete removal of the tumor, even without clinical signs of venous hypertension, is mandatory to avoid recurrence. Surgery could be performed with single- or double-staged procedures. We prefer to the latter. Because cardiopulmonary bypass (CPB) has been necessarily used to remove tumors and thrombus in the IVC and right heart cavities with success; both procedures have advantages and disadvantages: the one-stage technique involves sternotomy with CPB and exploratory laparotomy that could result in a longer intervention with potential for more complications (e.g., bleeding). In addition, mobilization of the IVC and occlusion may cause hemodynamic compromise that may require active or passive veno—veno bypass. Although the choice of the two surgical approaches remains controversial, nowadays, single-stage caval and cardiac resection of the tumor has been abandoned due to the high risks. We adopted a two-stage approach because the initial thoracic approach should allow safe resection of the intracardiac tumor mass; moreover, operative time was shorter and, despite the risks of a second general anesthesia, we potentially reduced the risk of bleeding following a systemic heparinization required for CPB. Furthermore, there is no "all in one operation" for this condition involving two anatomical regions, the heart on one hand, the vena cava, its tributaries and other peritoneal locations on the other. Surgical resection, the only logical form of treatment, should be a two-stage procedure and well-coordinated. Thus it is necessary that surgeons from multi-department make efforts to design an appropriate approach to accomplish the surgery.

References