Inguinal pain and fullness due to an intravascular leiomyoma in the external iliac vein

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ABSTRACT

Intravascular leiomyomatosis (IVL) is a benign smooth muscle tumor that evolves from the pelvic veins and can spread to the central veins and heart. Cardiac involvement is the most commonly reported presentation. Initial diagnosis is difficult, and IVL is commonly misdiagnosed as thrombus or atrial myxoma. Appropriate imaging and a high clinical suspicion are required for accurate diagnosis. We report a rare case of IVL in the external iliac vein that recurred 4 years after hysterectomy. Only four cases have been reported in the literature to involve the external iliac vein as it has no direct connection to pelvic venous drainage. (J Vasc Surg Cases and Innovative Techniques 2017;3:102-4.)

Intravascular leiomyomatosis (IVL), defined by noninvasive intravenous benign smooth muscle neoplasms, is rare and commonly originates from the pelvic veins of the broad ligament, uterus, ovaries, and vagina. Direct intravenous extension occurs in a subset of cases and, if untreated, can result in extension into the inferior vena cava or right atrium in a third of patients. Local disease typically causes symptoms of pelvic pain and fullness (90%), whereas intravenous extension can lead to congestive symptoms and thromboembolic events.12 Treatment involves resection with or without hormonal suppression and is well tolerated. Involvement of the external iliac vein (EIV) is exceedingly rare because of the lack of direct drainage from pelvic organs. Here we report an unusual case involving the EIV by growth through a collateral vein. Consent from the patient was obtained to present this case.

CASE REPORT

The patient is a 49-year-old woman who, 4 years before presentation, underwent a total abdominal hysterectomy for fibroids with IVL demonstrated on pathologic examination. During that operation, there was concern for a possible embolic event to the lungs thought to possibly be a leiomyoma. She was treated with a 6-month course of leuprolide to prevent regrowth, and repeated chest imaging demonstrated no evidence of intravascular filling defects. There was no further follow-up afterward. She subsequently developed pancreatitis, and a computed tomography scan demonstrated an incidental finding of a filling defect in the left EIV. She was referred from her gynecologist with concerns of possible IVL vs deep venous thrombosis. On initial evaluation, she had complaints of left inguinal fullness and discomfort for the past few months. Magnetic resonance imaging and duplex ultrasound demonstrated a nonocclusive intravenous mass within the left EIV deep to the inguinal ligament highly suggestive of IVL. The lesion was hypoechoic on ultrasound examination, causing a luminal flow disturbance without associated vessel wall inflammation or chronic changes (Fig 1). After discussion with her gynecologist, the decision was made to proceed with resection of the intravascular mass, repair of the vein, and bilateral salpingooophorectomy (BSO), given the likelihood of continued growth.

The surgical approach included retroperitoneal and groin incisions to allow proximal and distal control of the vein. A longitudinal venotomy was created in the distal EIV, exposing a spongy tan mass nonadherent to the vein wall. The mass had a stalk originating from a large venous side branch identified between the EIV and the common femoral vein. The involved side branch and portion of the EIV wall were resected en bloc with the mass (Fig 2). The venotomy was repaired using a bovine pericardial patch and 5-0 Prolene sutures. Laparoscopic BSO was then performed to reduce systemic estrogen levels and to decrease the risk of recurrence. Pathologic findings were consistent with IVL with estrogen and progesterone receptor-positive staining, without evidence of extension into surrounding tissue (Fig 3). The patient did well and was discharged on aspirin and a 3-month course of warfarin. At 6-month follow-up, she was asymptomatic, and duplex ultrasound showed a patent repair.

DISCUSSION

IVL is a rare condition found in women, most frequently around the age of 40 years. These women are primarily treated with hysterectomy, with or without BSO, or...
myomectomy. A preoperative diagnosis of IVL is difficult to make, and therefore diagnosis is typically made during hysterectomy, on histopathologic specimens, or during subsequent imaging studies. Presenting symptoms vary but are most commonly related to mass effect and venous obstruction, which are components of other uterine pathologic processes. Symptoms include abdominal pain, pelvic pain, and lower extremity swelling. Rarely, patients present with thromboembolic events involving the lower extremities, pulmonary circulation, or hepatic veins.

Although the leiomyoma is nonmalignant, there is significant danger associated with invasion of the central vessels and heart. Interestingly, cardiac cases (10% incidence) dominate the literature, probably because patients are symptomatic, leading to a reporting bias. In the nearly 300 cases of heart and central vein involvement, cardiopulmonary symptoms commonly include shortness of breath and palpitations. Reported deaths were largely due to late diagnosis or recurrence (30%) to the point of hemodynamic compromise and collapse. IVL is most commonly misdiagnosed as venous thrombosis and atrial myxoma.

Growth of the tumor into the venous system follows the pattern of drainage from the uterus, starting in the ovarian vein and leading into the internal iliac and common iliac veins. The internal iliac vein drains the uterine plexus and vaginal plexus of veins. EIV involvement, however, is exceptionally rare owing to the fact that it lies upstream from a venous drainage perspective and therefore not surprisingly has been reported in only four cases in the literature. In all four cases, there was continuous extension from the common femoral vein to the inferior vena cava with involvement of the right atrium on all occasions. Two cases were misdiagnosed as right atrial myxoma or sarcoma, and two were misdiagnosed as acute thrombus. Half of the patients were symptomatic with either dyspnea and syncope or lower extremity swelling. It is especially rare to discover IVL in the EIV and at such an early stage, as in our case, because of a lack of formal direct drainage from the pelvic venous system. Significant in this case is the presence of a side branch that likely connected the pelvic drainage with the EIV.

In this case, ultrasound was the most helpful in differentiating IVL from thrombus as it appeared nonocclusive, slightly compressible, echogenic, and without chronic vessel wall changes. Venous-phase computed tomography and magnetic resonance imaging are necessary adjuncts for accurate diagnosis and operative planning. IVL appears as a heterogeneous intravascular mass with extension to the iliac veins or inferior vena cava.
CONCLUSIONS
IVL is a rare entity that can often be misdiagnosed. Heightened clinical awareness coupled with appropriate diagnostic imaging allows timely intervention. Misdiagnosed or undiagnosed IVL is associated with mortality, highlighting the importance of appropriate assessment and identification.

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