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Case Report

Intravenous leiomyomatosis with diffuse dissemination on the right side of the pelvic cavity: a case report

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Abstract
Intravenous leiomyomatosis (IVL) is a rare smooth muscle tumor. It is characterized by nodular masses of histologically benign-looking smooth muscle cells growing within the uterine and extrauterine venous system. There are no clear guidelines regarding surgical approach and management of IVL in the literature. We would like to present an IVL case with extension to the right pelvic cavity and discuss the case in accordance with the current literature.

Key words:
Intravenous uterine leiomyomatosis, leiomyoma, uterus

Introduction

Intravenous leiomyomatosis (IVL) of the uterus is a rare benign smooth muscle proliferation originating from the smooth muscles of the uterus. The tumor is characterized by nodular masses. Nodular mass is composed of histologically benign-looking smooth muscle cells. Smooth muscle cells show unusual growth patterns in either veins or lymphatic vessels [1]. Despite IVL is histologically benign, it may also become aggressive through the following mechanisms: by involving the inferior vena cava (IVC), reaching the right-side cardiac chambers, or even invading the right pulmonary artery; therefore, the disease could prove fatal within a short time [2,3]. There are no clear guidelines regarding surgical approach and management of IVL in the literature. We would like to present an IVL case with extension to the right pelvic cavity and discuss the case in accordance with the current literature.

Case presentation

A 49-year-old multiparous woman was admitted to the gynecology clinic because of menometrorrhagia and pelvic pain. Pelvic examination revealed a 20 week gestational size, enlarged, asymmetrically shaped, mobile uterus. Medical and family histories were not remarkable. Transabdominal ultrasound of the pelvis showed a homogeneous hypoechoic soft tissue mass arising from uterus, measuring 23 x 16 x 9 cm in dimension, without necrosis or calcification. Abdominopelvic computerized tomography showed an intramural irregular uterine mass measuring 21 x 15 x 10 cm extending to the right adnexal region.

Laboratory tests, including complete blood count, serum electrolytes, coagulation studies, and tumor markers were normal. Pap test was negative and endometrial sampling showed proliferative changes without any evidence of malignancy. The clinical and radiological findings were suggestive of a uterine fibroid but there were concerns that a pelvic malignancy might be present as well.

Laparotomy was planned after an informed discussion with the patient. During laparotomy, the mass was noted to extend into the retroperitoneum in the right parametrial region, and exploration revealed white or creamy brownish serpiginous, worm like extensions from the mass insinuating to the hypogastric artery (Figure 1a). A total abdominal hysterectomy and bilateral salphingo-oophorectomy was done. Immediate frozen section of the specimen suggested presence of an IVL. A gynecological oncologist was invited to the operation for total removal of the lesion in the right pelvic cavity. The retroperitoneum was opened, hypogastric artery and vein was ligated and the IVL structures distal to the hypogastric artery were resected.

The final pathology report confirmed the presence of an IVL (Figure 1b and 1c). Immnuhistochemically, tumor cells

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were positive for smooth muscle actin (Figure 1d). Postoperative imaging examinations reported no pathologic lesions concerning the cardiovascular system of the patient including the heart and the lungs. GnRH agonist treatment was initiated for 6 months. Patient had an uneventful follow-up one year after surgery.

Discussion

Intravenous leiomyomatosis is an uncommon smooth muscle cell tumor of the uterus in which the tumor grows within venous channels without tissue invasion [4]. It is still unclear whether the neoplasm first originates from the uterus with subsequent vascular invasion or arises directly from the venous wall [5]. Uterine intravenous leiomyomatosis most commonly enters through the lumen of iliac vein and grows into the IVC, sometimes reaching the right atrium, ventricle and the pulmonary artery. Occasionally, the ovarian vein may provide an alternative route to the subphrenic segment of the inferior vena cava [4,6].

Figure 1a.

Figure 1a Macroscopic appearance of intravenous leiomyomatosis

It is reported the patients with intravenous leiomyomatosis have a median age of 45 years [1]. The symptoms of intravenous leiomyomatosis are determined by the location and the size of the tumors as followings: the pelvic mass, lower abdominal pain, abnormal vaginal bleeding due to the pelvic mass, and the uterine myoma; edema of the lower extremities due to occlusion of the IVC; shortness of breath, congestive heart failure and chest pain, or even sudden cardiac death associated with mechanical obstruction of the heart [5]. Since the diagnosis is easily missed particularly in the early stages, the cases of IVL were underestimated. The correct diagnosis of IVL in the perioperative period is also difficult, and IVL is usually detected intraoperatively during the exploration or postoperatively at the pathologic examination [7]. Presence of wormlike or nodular plugs in the pelvic veins and in descending order of frequency: in the broad ligament, uterus, ovaries, and vagina should raise the suspicion of an IVL. Encountering IVL with IVC involvement is not as uncommon as expected during hysterectomies. Since the patients with intravenous leiomyomatosis are often diagnosed by postoperative imaging studies after making a pathologic diagnosis, extrauterine involvement occurs in about 30% of the patients [8]. A preoperative diagnosis of IVL could not be made in our case.

Figure 1b.

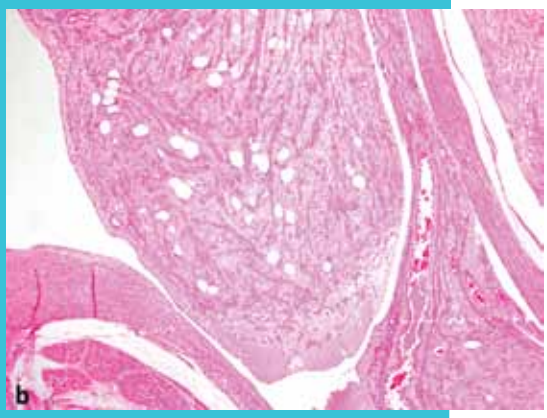

Figure 1c.

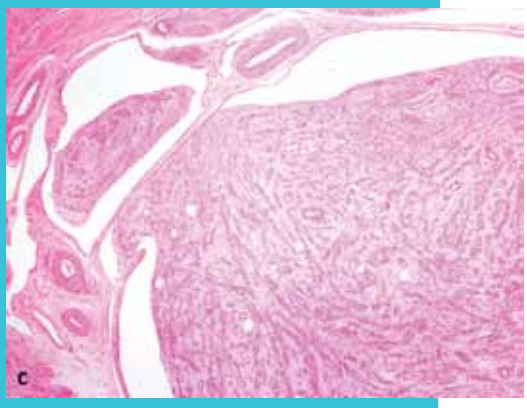

Figure 1b and 1c Microscopically, intravascular tumor component is composed of interlacing bundles of bland smooth muscle cells and few adipocytes in a hyalinized stroma (H&Ex40)

Typically, the characteristics of an affected uterus by IVL are followings: enlarged and heavy, the serosa is commonly bosselated. Tumorous nodules with irregular borders are frequently observed as multiple worm-like plugs of tumor. Nodules protrude from the cut-ends of the myometrial or broad ligament veins [8]. The following malignant tumors should be especially considered in the differential diagnosis of IVL: endometrial stromal sarcoma and leiomyosarcoma. Benign histology and immunohistochemical profiles are useful in the differential diagnosis. Histology reveals a proliferation of smooth muscle bundles invading vascular lumens and a 'vessel within a vessel' appearance characteristic of intravenous leiomyomatosis. There are no atypical cellular signs.

Surgery is the mainstay of treatment for IVL. When a complete removal of the tumor is not possible, ligation of the vein distal to the tumor has been suggested to prevent tumor growth along the IVC [8]. Most of IVL are commonly hormone receptor positive and estrogen dependent. Since the diagnosis is frequently made in a hysterectomy specimen, it is recommended to perform bilateral oophorectomy subsequent to surgery, if it has not already been performed. Resection should include the uterine tumors, parametrium tumors, ovaries and fallopian tubes, and intravenous and intracardiac tumors [4]. In our case, total hysterectomy was performed together with complete excision of the tumor disseminating into the right pelvic cavity, bilateral oophorectomy and ligation of the hypogastric arteries and veins distal to the tumor.

Since the tumor has the potential to recur and metastasize, careful long-term follow-up and use of postoperative medical treatment is advised in the literature. There are only a few case reports available in literature describing the use of a GnRH agonist for postoperative treatment. And the long-term therapy has been advised. Anti-estrogenic drugs (danazol, tamoxifen and gestrinone), gonadotropin-releasing hormone (GnRH) analogues and progesterone have also been used postoperatively [9]. The use of following antiestrogens has been considered for treatment of IVL: tamoxifen or raloxifene. However, their efficacy remains still controversial [10]. Follow-up with pelvic magnetic resonance imaging (MRI) or ultrasound and echocardiography is recommended for every 6 months for the first 5 years and recurrence has been reported in 30% of patients [11].

Figure 1d.

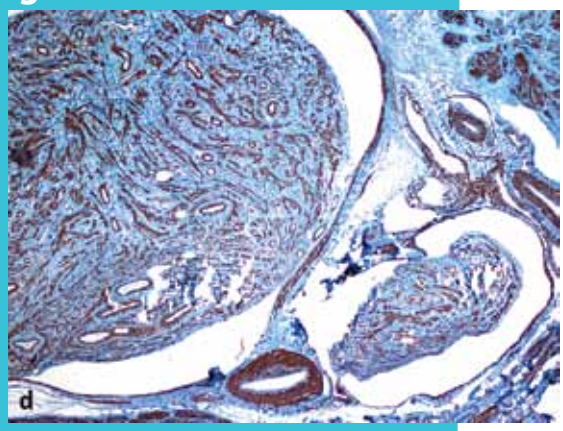

Figure 1d Immnuhistochemically, tumor cells positive for smooth muscle actin (anti SMAx40).

Although IVL is a rare and histologically benign tumor, it may sometimes be malignant in its mode of behavior. When the wormlike or nodular plugs are encountered in the pelvic veins of the broad ligament, uterus, ovaries, and vagina in a decreasing order of frequency, then IVL may be suspected intraoperatively. Intravenous leiomyomatosis is often diagnosed after pathological evaluation of the uterus. Hysterectomy together with complete resection of the tumor and bilateral oophorectomy is the best treatment of choice. Due to its potential to recur, careful, long-term postoperative follow-up is important for the success of the treatment.

Conflict of interest statement

The authors declare no conflict of interest.

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