

Uterine Intravascular Leiomyomatosis; A Rare Variety of Myoma, with an Unpredictable Evolution: A Case Report

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Abstract

Intravascular leiomyomatosis (IVL) is a benign pathology of the uterus. It is characterized by a leiomyocytes proliferation in the lumen of the myometrial and pelvic veins. Prognosis is related to its hormonal dependence and its potential vascular extension. Surgical excision must be complete to avoid recurrence's risk. In the light of a 31-year-old nulliparous patient case report, with a pelvic mass developed at the uterus, which histological examination confirmed the IVL diagnosis, we will discuss the clinical, evolution, prognosis and management features of this rare variety of myoma.

Keywords: Uterine intravascular leiomyomatosis, Myoma, Benign tumor of the uterus, Vascular extension.

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INTRODUCTION

Intravascular leiomyomatosis (IVL) is a rare benign tumor of the uterus. It usually occurs in the course of a fibromatous pathology, and can sometimes be revealed by complications related to its proliferation and migration. This tumor, although benign, has a high potential for intravascular development and requires specific management and in-depth knowledge to avoid recurrence.

We report an observation of IVL limited to the gynecological sphere. Based on this case and a review of the literature, we will discuss the clinical, evolution and management features of this exceptional variety of myoma.

CLINICAL CASE

Ms. SC, 31 years old, nulligest, regular menstrual cycle, without any particular pathological history, was treated for a recent pelvic mass associated with dysmenorrhea, pelvic pain dysuria and constipation without menometrorrhagia.

Clinical examination revealed a well-limited, firm, painless pelvic mass reaching the halfway of umbilicus. The cervix was macroscopically healthy.

Ultrasound showed a uterus of a normal size and homogeneous structure, a thin hyperechoic vacuity

line in place, surmounted by a heterogeneous echogenic pelvic mass, tissue echostructure, measuring 83/67 mm and located laterally and retro uterine left (Figure 1). The ovaries were not seen, which made the diagnosis difficult. The tumor marker CA 125 was negative, and liver ultrasound was normal.

Surgical exploration revealed a uterus of normal appearance giving rise to a large left latero-uterine mass, well limited, with a firm consistency and fibromatous appearance, measuring 12 cm in diameter, invading the whole left fallopian tube as well as the left broad ligament, and partly the left ovary, without ascites or associated peritoneal carcinosis. We performed a total resection of the mass, removing the left adnexa with it (Figure 2).

Anatomopathological analysis concluded to a disseminated intravascular leiomyomatosis without any sign of malignancy. On macroscopic section, intravascular invasion was evident. On microscopy, this tumor invasion was composed of leiomyocytes organized in clear limited, hyalinized nodules with intravascular development.

As the vascular axes had not been explored either during the preoperative imaging and the surgery, a CT scan with an injection of a radiological contrast medium at the venous time was performed. It did not detect any intra-luminal venous extension, either at the

iliac, ovarian, or inferior vena cava. There was no residual mass.

No additional surgical or hormonal treatment was therefore proposed, but a clinical and ultrasound monitoring. The postoperative period was normal, and prophylactic anticoagulation was prescribed. Doppler ultrasound monitoring of the ilio-caval venous axes was implemented because of the risk of long-term vascular extension. The patient did not present any complications after five months.

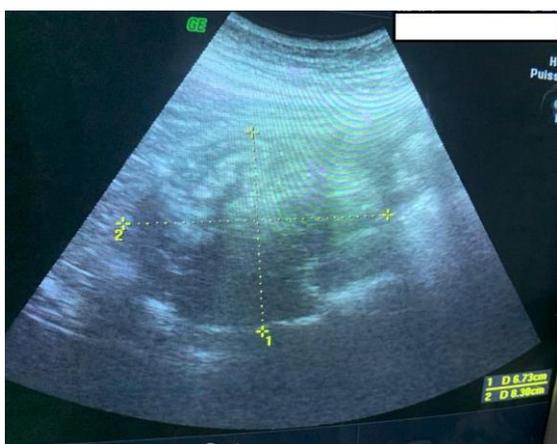


Figure 1: Image of an ultrasound section showing the echostructure and dimensions of the uterine mass



Figure 2: Surgical resection piece removing the tumor and the left adnexa

DISCUSSION

IVL is a rare pathology, corresponding to a benign leiomyocytes proliferation located in the vascular lumens of the uterus, which may occur locally or extend into the veins of the pelvis to the right heart chambers, leading to the death of the patient [1]. The average age of diagnosis is 47 years, with extremes of 28 to 80 years [2].

Two pathogenic hypotheses are frequently advanced. These are either invasion of the venous vascular lumen by a leiomyoma, or invasion of the vascular lumen by leiomyocytes of the venous wall [3]. For some, a vascular endothelial origin is also considered [4].

The clinical symptoms depend essentially on the tumor location. If it is located in the uterus, it follows the symptomatology of a fibroid uterus: enlarged uterus, menometrorrhagia, pollakiuria, heaviness and pelvic pain [5,6,7,8]. In case of intravenous evolution, the symptomatology depends on the ectopic extension topography: in the pelvic veins, there will be a lower limbs edema [9, 10]. If the tumor reaches the heart, the patient will present rhythm disorders [11, 12], a heart failure [13, 14], syncope [15], or will die from that complication [1].

The differential diagnoses are: diffuse leiomyomatosis, benign metastasis of a uterine myoma [11], uterine leiomyosarcoma, or endometrial stromal sarcoma [16].

Preoperative diagnosis of IVL is difficult and often fortuitous, as in our case. There are no specific imaging features. It is most often a pathological finding [17]. That's why any myoma's removal must be subject of a careful histological examination [18]. Magnetic resonance imaging (MRI) may be more helpful than CT to determine the vascular nature of this tumor and its extension [15, 19]. Additional informations can be provided by Doppler ultrasound and phlebocavography in matter of abdominal metastases [1]. MRI is recommended for the diagnosis of cardiac extensions [16].

The treatment consists of a total hysterectomy with bilateral adnexectomy, with removal of ectopic extensions. The excision must be total in order to avoid recurrence from residual tumor elements.

Moreover, it is a hormone-sensitive tumor, secreting oestradiol itself, which explains the disturbed hormonal balance in postmenopausal patients [6]. Several teams have used gonadotropin-releasing hormone (GnRH) analogues as a complement to surgical treatment, either preoperatively [8], or when resection was not complete [15].

In the absence of scientific data on the predictive factors of vascular evolution, it seems licit, considering the reported cases, to carry out a vascular extension assessment at the time of diagnosis [17].

Follow-up should be long and periodic with MRI and/or ultrasound; some authors recommend control every six months [19]. If abdominal or pelvic extension is noted, transesophageal cardiac ultrasound is indicated [17].

CONCLUSION

Due to the lack of a uniform diagnosis consensus and the rarity of this pathology, IVL can be multifaceted. In general, the diagnosis is histological. MRI imaging is necessary to assess vascular extension. Complete surgical removal is necessary to avoid any risk of recurrence. Its hormonal dependence requires definitive castration and prohibits any hormone replacement therapy. Treatment with anti-estrogens may be associated. Periodic and prolonged follow-up with imaging is recommended to detect any recurrence.

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