CASE REPORT

Intravenous Leiomyomatosis of the Uterus: An Intriguing Case Revealed through Anatomopathological Examination

Léiomyomatose intraveineuse de l'utérus: Un cas intriguant révélé par un examen anatomopathologique

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Abstract

Introduction: Intravenous leiomyomatosis (IVL), a rare type of uterine leiomyoma (its incidence is about 0.25% to 0.40% of patients who present uterine fibroma), is characterized by the formation and growth of benign leiomyoma tissue within the vascular wall or lymphatic lumen.
Herein, we presented a case of early stage of IVL successfully treated by surgical removal and a review of actual medical recommendations.
Observation: A 49-year-old woman, gravida 2 para 2, presented to our department with hypogastric pain. On physical examination, a palpable mass in the hypogastrium was noted. Pelvic ultrasound showed a huge uterus with multiple heterogeneous leiomyomas. As the patient was symptomatic and as she had completed their family plan, the decision to perform a total abdominal hysterectomy with bilateral salpingo-oophorectomy was taken. On pathological examination, intravascular growth of benign smooth muscle cell was found within venous channels lined by endothelium. The diagnosis of IVL of the uterus without malignant transformation was confirmed. The patient was monitored for 14 months, and subsequent computed tomography did not reveal any evidence of tumor recurrence.
Conclusion: IVL is a benign, rare and potentially lethal pathology. Clinical manifestations are nonspecific. IVL needs surgical treatment for diagnosis and therapeutic purposes. They require close and prolonged follow-up because of the high risk of recurrence.

Key words: Benign Neoplasm, hysterectomy, intravenous leiomyomatosis, prognosis

Résumé

Introduction: La léiomyomatose intraveineuse (LIV) est un type rare de léiomyome utérin (son incidence est de 0,25% à 0,4% des patientes qui présentent un fibrome utérin) qui se caractérise par la formation et la croissance des cellules musculaires lisses dans la paroi vasculaire ou la lumière lymphatique.
Nous présentons un cas de stade précoce de LIV traitée avec succès par un traitement chirurgical radical et une revue de la littérature.
Observation: Une femme âgée de 49 ans, deuxième pare deuxième geste, s'est présentée dans notre service pour des douleurs hypogastriques. À l'examen physique, une masse palpable dans l'hypogastre a été notée. L'échographie pelvienne a montré un utérus augmenté de taille avec multiples léiomyomes hétérogènes. Vu que la patiente était symptomatique et ayant son capital d'enfants, la décision a été de réaliser une hystérectomie abdominale totale avec annexectomie bilatérale. À l'examen anatomopathologique, une croissance intravasculaire de cellules musculaires lisses bénignes a été notée. Le diagnostic de LIV de l'utérus sans transformation maligne a été confirmé. La patiente a été surveillée pendant 14 mois et la tomodensitométrie ultérieure n'a révélé aucun signe de récidive tumorale.
Conclusion: La LIV est une pathologie bénigne, rare et potentiellement mortelle. Ces manifestations cliniques sont non spécifiques. Elle nécessite un traitement chirurgical le plus souvent radical et un suivi étroit et prolongé en raison du risque élevé de récidive.

Mots clés: Hystérectomie, léiomyomatose intraveineuse, pronostic, tumeur bénigne

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Introduction

Leiomyomas beyond the uterus are defined by benign smooth muscle cell tumors outside of the uterus (1). Either the uterine leiomyoma invades the venous system or it is a growth of the smooth muscle cells of the vascular wall (2). Intravenous leiomyomatosis (IVL) or angioleiomyoma is a rare type of uterine leiomyoma and features formation and growth of benign leiomyoma tissue in the vascular wall or lymphatic lumen without necessarily invading them (1).
The tumor can grow along blood vessels, extending to the iliac vein, inferior vena cava, and even heart (2). IVL is a potentially lethal pathology if the invasion reaches vital organs and the diagnosis is not made in time. The incidence of this tumor is about 0.25% to 0.40% of patients who present uterine fibroma (3). IVL mainly affects pre-menopausal women between the age of 40 and 50(3).
Herein, we presented a case of IVL successfully treated by surgical removal and a review of actual medical recommendations.

Observation

A 49-year-old woman, gravida 2 para 2, with no family history of uterine myomas mentioned, having a history of arterial hypertension presented to our department with hypogastric pain and abnormal uterine bleeding in the prior five months resulting in anemia, which required iron supplementation. On physical examination, we noted a blood pressure at 130/ 70 mmHg and pulse at 80 bpm. A palpable mass in the hypogastrium was noted. The speculum examination was normal. On vaginal examination coupled with abdominal palpation, the uterus was found to be about 14 weeks of gestation. Pelvic ultrasound showed an enlarged uterus measuring 16x14x12cm with multiple fibroids. The biggest fibroid measured 5cm. Computed tomography (CT) scan and magnetic-resonance imaging were not done initially due to the unaffordability of the patient. The initial diagnosis was a leiomyoma or sarcoma given the rapid increase in uterine volume. As the patient was symptomatic and as she had completed their family plan, the decision to perform a total abdominal hysterectomy with bilateral salpingo-oophorectomy was taken after patient's consent. Abdomen was opened by midline vertical incision. During surgery, multiple subserosal, intramural, and submucosal fibroids ranging from 2×3 to 10×10 cm were seen. On pathological examination, the uterus measured 19 cm in largest diameter and weighed 1.3 kg. Cut section showed white nodular myometrial masses (Figure 1). Microscopically, intravascular growth of benign smooth muscle cell was found within venous channels lined by endothelium (Figure 2).

The diagnosis of IVL of the uterus without malignant transformation was confirmed. The postoperative course was uneventful, and the patient recovered well without any complications. The patient was monitored for 14 months, and subsequent CT did not reveal any evidence of tumor recurrence. The follow up will be performed annually until the age of menopause.

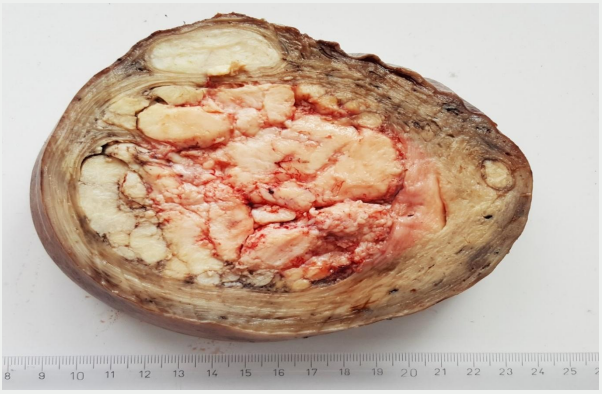

Figure 1. Cut section of the uterus: multiple white nodular myometrial masses within the myometrium (yellow arrow)

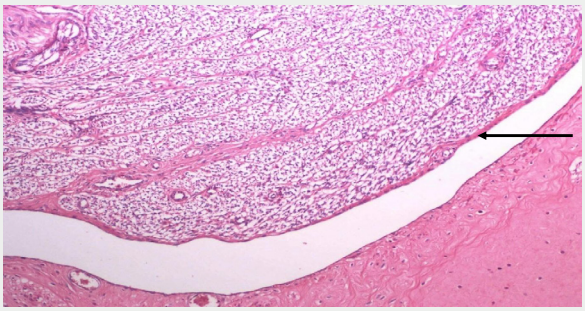

Figure 2. Microscopic appearance: Intravenous leiomyomatosis is found within venous channels lined by endothelium (HEx100) (black arrow)

Discussion

In this case, IVL limited to the uterus has a favorable prognosis after radical treatment with disappearance of pelvic pain. In fact, postoperative clinical and radiological monitoring did not reveal any local or extragynecological recurrence.
The precise etiology of IVL is still unclear, although there are two hypotheses for the origin of intravenous leiomyomatosis(4). The first is that the uterine leiomyoma invades the uterine vein and then grows in the venous system (4). The second suggests that the smooth muscle cells of the vein abnormally proliferate in the venous system (4). However, the second hypothesis was recently refuted and studies found no endothelial tissue markers (CD10, CD31 and CD34) in IVL tumors (4). Uterine leiomyomas can grow along the veins or lymphatic vessels, but there are two main ways to grow up; one is through the uterine vein into the internal iliac veins; common iliac veins, inferior vena cava, and rarely goes to the right atrium, ventricle, and pulmonary artery. Another route is through the right ovarian vein (4).
The early diagnosis of IVL is usually difficult. At the early stage of IVL, most of the patients are asymptomatic despite intravenous extension, and the tumor extension remains inside the small vessels of the myometrium and

cannot be detected by CT scan or magnetic resonance imaging (5). Clinical manifestations are nonspecific depending on the location and size of the tumor (5). For example, in the case of a pelvic mass or uterine leiomyoma, patients may either be asymptomatic or complain of abnormal uterine bleeding, pelvic pain, and abdominal distension (6). Regarding the inferior vena cava and heart, patients may have symptoms such as chest pain, dyspnea, lower extremity edema, and even sudden death (6). The diagnosis of certainty is based on anatomopathological analysis (2). On gross examination worm like growths of smooth muscle can be seen on the blood vessels or lymphatics(2). Microscopically, the tumor is positive for desmin and actin(2). There is also the presence of hormonal receptors to progesterones and estrogens (2). The mitotic analysis reveals a low activity of the cells(2).

IVL needs surgical treatment for diagnosis and therapeutic purposes (2). Total hysterectomy is necessary to prevent recurrence (4). Resection should include the uterus, fallopian tubes, ovaries and intravascular tumor (4). Myomectomy is only suitable for young women with fertility needs (4). If an extrauterine invasion is present; a multidisciplinary team is needed to ensure a successful surgery (1). A one or two-stage operation can be considered depending on the general condition of the patient and tumor invasion (7).

The recurrence rate of IVL varies from 14% to 31% (8). The difference in recurrence rate is due to the difference in surgical methods (4). Women who have had myomectomy or hysterectomy alone are more likely to recur compared to women who have undergone hysterectomy with bilateral salpingo-oophorectomy (8). However, an incomplete resection increases the risk of recurrence of IVL. The earliest recurrence was noted after 6 months (8). The risk factors for recurrence were the incomplete tumor resection, involvement of the iliac vein or genital vein, involvement of the inferior vena cava, and size of the pelvic tumor more than 15 cm (8).

IVL strongly expresses estrogen receptors (2). In fact, women steroids have been incriminated in the pathogenesis of this disease (2). For this reason, several authors have evaluated the efficacy of hormonal therapy in the treatment of the IVL as an adjuvant for the patients with incomplete resection or who do not have surgery (2). The commonly used agents are gonadotropin releasing hormone agonists, tamoxifen, and medroxyprogesterone to prevent recurrence (5). New molecules have been used such as Crizotinib in tumor's recurrence (9). However, data on the effectiveness of this treatment are limited due to the rarity of this tumor. Since postoperative recurrence of IVL may occur after many years of surgery, long-term follow-up is mandatory, and annual CT examination is recommended (10).

This case report presents two limitations. First, we did not have radiological explorations to better characterize the leiomyoma before the operation. Second, the postoperative period was too short to assess the long-term risk of recurrence.

Conclusion

IVL is a benign leiomyoma developed within the vascular or lymphatic lumen. It's a rare pathology but potentially lethal if there is a cardiac extension. Clinical manifestations are nonspecific depending on the location and size of the tumor. Surgical resection is the main treatment for IVL. The choice of treatment depends on the parity and extension of the tumor. In the event of extrauterine invasion, multidisciplinary approach is necessary. They require close and prolonged follow-up because of the high risk of recurrence.

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