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Case Report

Intravenous Leiomyomatosis, Case Report and Literature Review

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Abstract

Intravenous leiomyomatosis (IVL) is a rare and unique variant of uterine fibroids, characterized by the benign proliferation of smooth muscle cells within the venous system. Despite its benign histological nature, IVL exhibits aggressive behavior by extending beyond the uterus, potentially reaching the inferior vena cava, heart, and pulmonary arteries. Due to its rarity and nonspecific early clinical presentations, IVL is often misdiagnosed or detected late, leading to challenges in treatment. We present a case of a 45-year-old woman initially suspected to have leiomyosarcoma, who underwent radical hysterectomy. Postoperative histopathology confirmed IVL. Complete surgical excision was performed, and a five-year follow-up was planned due to the risk of recurrence. This case put the light on the importance of high clinical suspicion and thorough surgical management in ensuring optimal outcomes for IVL.

Keywords: *Intravenous Leiomyomatosis; Uterine Fibroid; Hysterectomy*

Abbreviations

IVL: Intravenous Leiomyomatosis; AP: Anterior Posterior Diameter; CT: Computerized Tomography; MDT: Multidisciplinary Team

Introduction

Uterine fibroids, or smooth muscle leiomyomas, are common benign tumors of the uterus. Despite their benign nature, there are several rare variants where these tumors extend into distant organs, such as the great vessels or the lungs. These rare conditions present unique challenges in diagnosis and treatment due to their atypical growth patterns and locations outside the uterus [1].

Histologically, these variant leiomyomas resemble typical benign uterine leiomyomas both grossly and microscopically but exhibit different growth patterns that extend beyond the uterus. These variants include: intravenous leiomyomas, disseminated peritoneal leiomyomas, parasitic leiomyomas, and benign metastasizing leiomyomas [2].

Intravenous leiomyomatosis (IVL) is a rare and special type of smooth muscle tumor, that is classified as a benign disease according to its histological features but is malignant in terms of its behavior. Typically, IVL originates in the uterus, but it can extend along the venous system to the inferior vena cava, the right heart, and even the pulmonary artery. The clinical manifestations and ultrasound images of IVL in the early stages are not typical; thus, IVL is easily misdiagnosed or completely missed. As recurrence is frequent, it should be followed up more vigorously than ordinary uterine leiomyomas. IVL is relatively rare [3].

Incomplete tumor resection, extent of tumor lesions and size of the pelvic tumor were risk factors for post- operative recurrence and progression of intravenous leiomyomatosis [4].

Because of its rarity, uterine leiomyomatosis is occasionally misdiagnosed or diagnosed only latterly, leading to improper treatment [2].

We present a rare case of IVL, managed surgically as a leiomyosarcoma and histopathology confirmed IVL.

Case Report

45 years old lady who was referred to hospital with uterine fibroids and heavy menstrual bleeding, she is up to date with her cervical smears. she is para 2 previous vaginal deliveries.

Her pelvic scan showed bulky uterus, measures in excess of length of 132 mm and AP diameter of 86mm with multiple intramural and subserosal fibroids.

She had MRI (magnetic resonance image), which showed there is a large highly vascular serpiginous posterior myometrial mass measure 14.5 cm, extending all the way to the cervix, encasing it and indenting on the superior margin of the bladder. with tiny foci of intralesional necrosis or invasion of adjacent organs is seen. No invasion of the surrounding structures. No significant lymphadenopathy demonstrated or ascites. The mass shows high intensity T1 areas? haemorrhage. Impression=? Sarcoma (Figure 1 and 2).

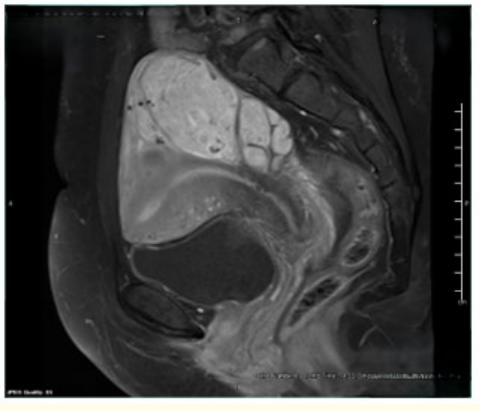

Figure 1

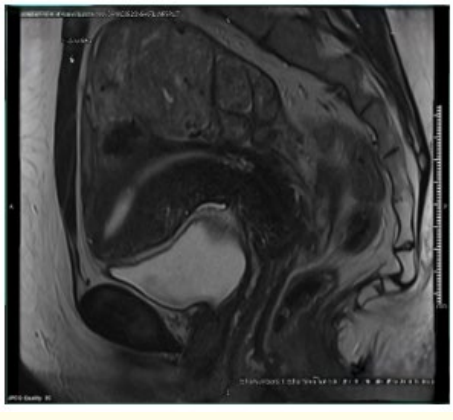

Figure 2

She had CT (computerized tomography) of chest abdomen and pelvis-with no metastatic evidence.

She case was discussed in the oncology MDT (multidisciplinary team) which advised for radical hysterectomy.

She had radical abdominal hysterectomy and bilateral salpingo-oophorectomy.

Intraoperative findings

Distorted anatomy on both uterine sides, retroperitoneal mass distorting the anatomy of uterus.

Medium side fibroid? Covering the anatomy of the uterus.

Histopathology report came as intravenous leiomyomatosis of the uterus. No invasive malignancy.

With a follow up plan for 5 years as intravenous leiomyomatosis has a small risk of local relapse.

Discussion

Intravenous leiomyomatosis is marked by the growth of a histologically benign smooth muscle cell tumor mass within veins, without invading the surrounding tissue [2]. With fewer than 200 cases reported overall, uterine leiomyomatosis is occasionally misdiagnosed or diagnosed late due to its rarity, which can result in improper treatment [5].

Our case was suspected to have leiomyosarcoma and underwent radical hysterectomy for that, fortunately histopathology was benign.

IVLs are generally asymptomatic and incidentally detected by ultrasound examination. Reliable biomarkers for the early detection of IVL have not yet been reported; therefore, early preoperative detection is difficult [5].

The number of IVL cases is likely underestimated, as the diagnosis is frequently missed, particularly in the early stages when the tumor is confined to the small vessels of the myometrium and cannot be detected through preoperative imaging. Accurate diagnosis depends on maintaining a high level of suspicion [1].

Surgery is the treatment of choice, and complete removal of the tumor is mandatory. Incomplete resection of the tumor may result in recurrences up to 15 years after the primary occurrence [6]. Bilateral oophorectomy is also advisable because the tumor is estrogen dependent [7] functioning ovarian tissue may remain. Therefore, long-term treatment with GnRH agonist may be useful in preventing recurrence of this disease [7].

Conclusion

Intravenous leiomyomatosis (IVL) is a rare, histologically benign smooth muscle tumor that can be challenging to diagnose due to its asymptomatic nature and lack of reliable biomarkers. Misdiagnosis or late diagnosis is common, particularly in the early stages when the tumor is confined to small myometrial vessels and may not be detectable on preoperative imaging. Despite its benign histology, IVL requires surgical intervention, as incomplete resection can lead to recurrence even years after the initial surgery.

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