Case Report

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Ultrasound appearance of intravenous leiomyomatosis

A case report

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Abstract

Rationale: Intravenous leiomyomatosis (IVL) is a rare benign smooth muscle tumor that can develop from the pelvic or uterine veins and spread into the central veins and heart. Here, we report a case of recurrent IVL in a 48-year-old woman. To the best of our knowledge, this is the first case report of IVL that describes the characteristic ultrasound features of the tumor, including the rainbow sign.

Patient concerns: A 48-year-old woman developed a solid-cystic lesion in the inferior vena cava (IVC) 3 years after undergoing a right heart tumor resection and 5 years after undergoing hysterectomy.

Diagnoses: Physical examination was unremarkable. However, ultrasonography showed a solid-cystic lesion in the IVC, and a diagnosis of IVL was made.

Interventions: The patient underwent complete surgical removal of the tumor by a multidisciplinary team. The tumor was resected successfully.

Outcomes: Pathological examination confirmed that the IVC tumor thrombus was consistent with IVL. During follow-up, there were no signs of local or distant recurrence.

Lessons: The preoperative diagnosis of IVL is difficult, and the tumor is usually misdiagnosed as a thrombus or right atrial myxoma. A thorough understanding of the characteristic imaging features of IVL is essential for an accurate preoperative diagnosis. The lesion in our patient showed multiple tracts, a honeycomb appearance, and vividly colorful blood flow that resembled a rainbow, which we termed the rainbow sign.

Abbreviations: CDFI = color Doppler flow imaging, IVC = inferior vena cava, IVL = intravenous leiomyomatosis.

Keywords: benign tumors, intravenous leiomyomatosis, ultrasound diagnosis

1. Introduction

Intravenous leiomyomatosis (IVL) is a rare intravascular smooth muscle tumor. IVL typically occurs in women with uterine hysteromyoma, and is believed to originate from the smooth muscle cells in the pelvic or uterine veins. Although IVL is histologically benign, it has the potential to grow along the veins and spread beyond the pelvic region and into the central veins and heart.[1] It can, therefore, cause severe complications, such as right ventricular outflow tract obstruction, pulmonary embolism, and sudden death.

The early diagnosis of IVL is challenging, and these tumors are usually misdiagnosed as a thrombus or a right atrial myxoma. The correct diagnosis is typically made during hysterectomy, through histopathological examination, or during subsequent imaging studies. A comprehensive understanding of the characteristic imaging features of IVL is essential for an accurate preoperative diagnosis.

To clarify the imaging features of IVL, we present herein a case of IVL in a 48-year-old woman, and describe in detail the ultrasound features of this tumor. We also review the literature on IVL. We hope that our findings will facilitate the preoperative diagnosis of IVL.

2. Case report

A 48-year-old woman (gravida 2 and para 2) complained of dizziness and asthenia for 2.5 years and occasional syncope for 2 months. She had undergone a hysterectomy for uterine hysteromyoma at the age of 43 years at a local hospital, but no postoperative pathological examination had been performed

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Informed written consent was obtained from the patient for the publication of this case report and the accompanying images.

The study was approved by the Institute Research Ethics Committee of Peking Union Medical College Hospital.

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at that time. Three years ago, she had undergone a heart tumor resection. The postoperative pathological examination at that time revealed the presence of IVL, but further treatment was not administered. Six months ago, during a routine follow-up, a hypoechoic lesion was found in the right heart and inferior vena cava (IVC) on ultrasonography.

She was then referred to our hospital for further treatment and was admitted to the cardiothoracic surgery department on 10 January 2018. A physical examination on admission was unremarkable. We performed a meticulous and comprehensive ultrasound examination. Two adjacent, irregular bar-like hypoechoic lesions were observed in the right uterine adnexal area, 1 measuring 5.5 cm × 1.7 cm, and the other measuring 4.3 cm × 1.0 cm. Color Doppler flow imaging (CDFI) showed that the lesions had abundant blood flow, and the Doppler spectrum was of high velocity and low resistance. Furthermore, a hypoechoic, intraluminal, consecutive, cord-like mass was detected. This mass extended from the right internal iliac vein and right common iliac vein into the IVC, almost completely filling the IVC lumen and entering the right atrium.

Within the hypoechoic lesion in the IVC, multiple stripe-like hyperechoic lines were observed parallel to the long axis of the vein, with anechoic lesions between the lines. We referred to this appearance as multiple tracts (Fig. 1). A transverse section through this lesion had a honeycomb appearance (Fig. 2), and CDFI showed that the anechoic lesions were filled with blood flow. These lesions vividly mimicked a rainbow in the sky after a heavy rain (Fig. 3), and we termed this feature the rainbow sign.

We then conducted a contrast-enhanced ultrasound examination. A 2-mL bolus of sulfur hexafluoride (SF_6) was rapidly injected through an elbow vein. After the injection, the hypoechoic mass in the IVC soon filled with microbubbles in the early stage of the arterial phase. In the venous phase and delayed phase, the microbubbles slowly subsided. The lesion in the vein was enhanced, and the enhancement was consistent with the long axis of the vein (Fig. 4).

Two weeks later, on 29 January 2018, an operation to resect the tumor was carefully designed and conducted by a multidisciplinary team. The tumor was eventually removed successfully. During the operation, the IVC mass was found to be completely separate from the vein and had no adhesions with the wall of the right atrium or the IVC. After the operation, the patient was immediately transferred to the intensive care unit, where she underwent an unremarkable postoperative course and was transferred to the ward after 36 hours.

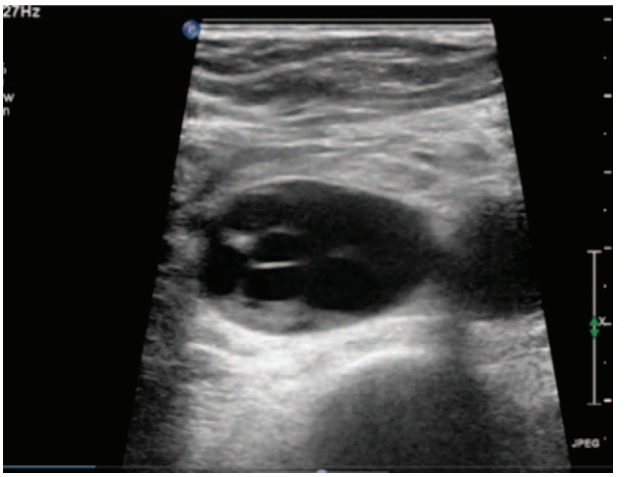

Figure 2. A transverse section through the inferior vena cava has a honeycomb appearance.

The surgical specimen from the IVC mass was firm, rubbery, and pale on gross examination. Upon opening the tumor for exploration, we observed several long cavities in the mass, with diameters of 0.5 to 1.2 cm. Pathological examination confirmed that the IVC tumor thrombus was consistent with IVL. The postoperative course was uneventful, and the patient was advised to return home 7 days after the surgery. Because the tumor had estrogen receptors,[2] the patient was treated with tamoxifen. During a 6-month follow-up, she remained asymptomatic with no evidence of tumor recurrence on ultrasonography.

3. Discussion

IVL was first reported by Birch-Hirschfeld in 1896 as a tumor with the potential to infiltrate veins.[3] IVL is a rare condition that

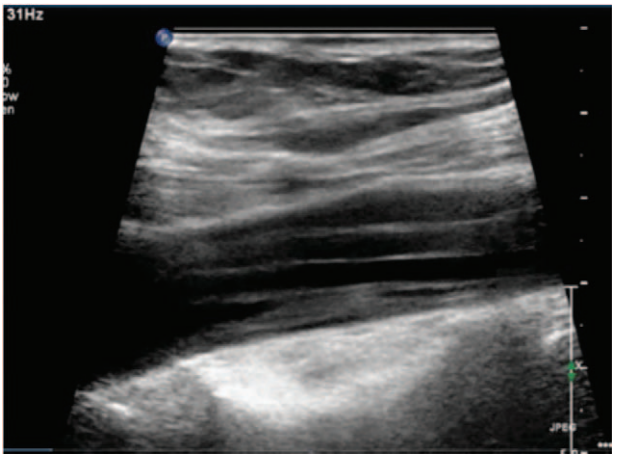

Figure 1. Multiple stripe-like hyperechoic lines parallel to the long axis of the inferior vena cava.

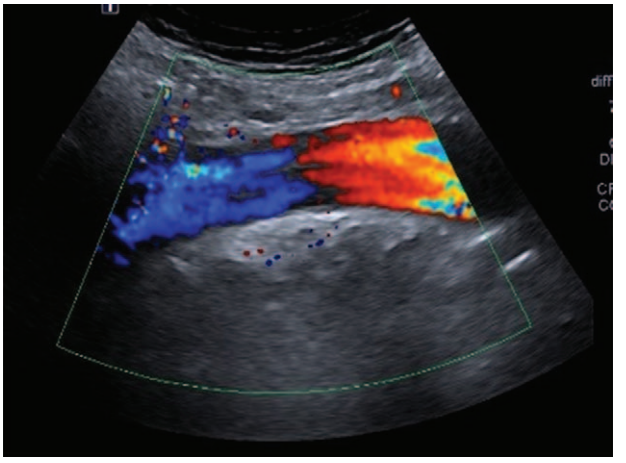

Figure 3. The anechoic lesion, filled with colored blood flow, is termed the rainbow sign.

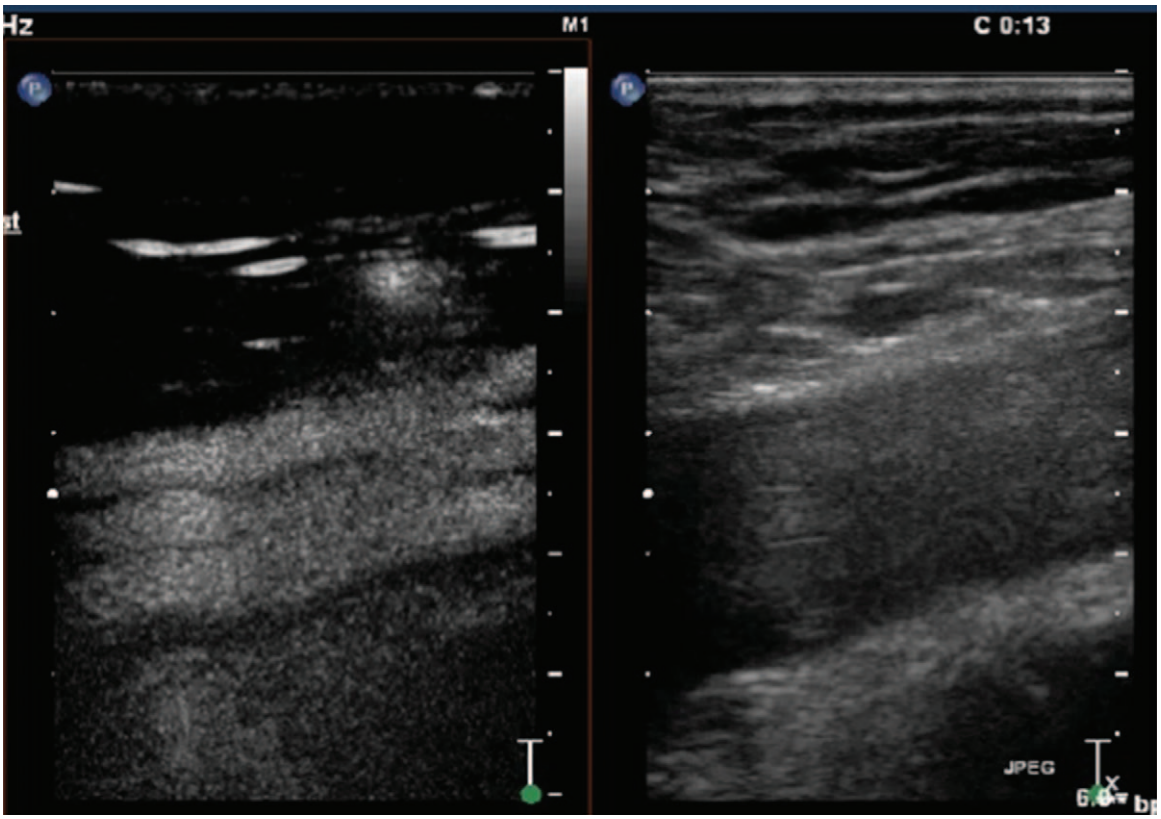

Figure 4. On contrast-enhanced ultrasonography, the lesion shows enhancement that is consistent with the long axis of a vein.

predominantly affects premenopausal women or women of childbearing age, usually around the age of 40 years. Many patients have a history of hysterectomy, with or without bilateral salpingo-oophorectomy or myomectomy. The clinical symptoms of IVL depend on the extent of the lesions. In the early stages, many patients are asymptomatic, and the tumor may be discovered incidentally. Most patients eventually develop symptoms, such as abdominal or pelvic pain or discomfort.[4–6] If the systemic veins and right atrium are involved, symptoms of impaired venous circulation will develop, including swelling of the lower extremities, chest tightness, chest pain, congestive heart failure, and even sudden death. Rarely, patients present with thromboembolic events involving the lower extremities, pulmonary circulation, or hepatic veins. In a few patients, IVL can metastasize to the lungs.[7,8]

Thus far, no consensus has been reached regarding the cause of IVL, but there are 2 principal theories that are widely accepted.[1,9] The first states that leiomyomatous cells originating from the uterine myometrium invade the intima of the myometrial sinuses. The second holds that the tumor consists of proliferating smooth muscle cells and arises directly from the venous wall of the uterine or pelvic veins.

The preoperative diagnosis of IVL mainly depends on imaging studies. Magnetic resonance imaging and computed tomographic angiography are considered the most sensitive methods to diagnose this pathology and to plan the optimal surgical strategy,[10] as these techniques can show the overall structure of the lesion. In addition, ultrasonography is an indispensable diagnostic tool in IVL, as it enables the real-time imaging of the extent of the tumor and the sensitive analysis of blood-flow signals within the tumor. Many patients have a hypoechoic lesion in the uterine or systemic veins, which is usually misdiagnosed as a myoma or venous thrombosis. A cordlike hypoechoic lesion is usually found in the pelvic veins and can even extend into the IVC. In the final stages, a mass can be seen extending into the right atrium.[11–13]

The features described above are consistent with those observed in our patient; a cordlike lesion was indeed seen in the IVC. However, a detailed description of the ultrasound features of IVL has never been reported before. The early diagnosis of IVL is difficult because patients may be asymptomatic, despite much intravenous extension. When symptoms do appear, they are often secondary to direct cardiac involvement.

The gold standard treatment for IVL is complete surgical resection.[4] The treatment of IVL has 2 goals:[14] to remove the tumor burden and to prevent tumor recurrence. Treatment consists of total hysterectomy and bilateral salpingo-oophorectomy, with the removal of as much of the extrauterine tumor as possible to ensure a complete resection.[15] The tumor can be completely free within the vessel lumen, as in the case of our patient, or less commonly, can have slight adhesions to the vessel or atrial walls. If complete resection is not achieved, there may be a recurrence and further surgical treatment may be required. IVL resection may be performed as either a 1-stage or 2-stage operation.[16,17]

Thus far, no agreement has been reached as to the optimal surgical approach for resection of this type of tumor. The procedure chosen varies widely among surgeons, but mainly

depends on the range of the tumor and the patient's ability to tolerate surgery.[18] Single-stage surgery was performed in our patient because this technique enabled us to control both the cardiac and abdominal fields by means of bypass and hypothermic circulatory arrest. This facilitated IVL removal and minimized blood loss and risk to the patient. Some studies recommend using an IVC filter to prevent postoperative or late pulmonary embolism.[19]

On immunohistological examination, the tumor is positive for smooth muscle actin, CD34, vimentin, and desmin and negative for cytokeratin, CD10, and epithelial membrane antigen.[20] IVL may also express estrogen receptors,[3] and so, antiestrogens such as tamoxifen may be administered in patients with unresectable tumors, incomplete resection, or tumor recurrence.[21] However, outcomes vary among studies.[15,22,23]

4. Conclusions

A diagnosis of IVL should be considered when a mass is discovered extending from the IVC to the right atrium on ultrasonography in a middle-aged woman, especially a woman with a history of hysterectomy or uterine hysteromyoma.[1] Ultrasonography, combined with contrast-enhanced ultrasonography, is a useful tool for the diagnosis of IVL, as it can provide information on the blood flow within the lesion, which is valuable during the differential diagnosis. Given the tumor's tendency to relapse, long-term postoperative follow-up is critically important. No recurrence or metastasis was found in our patient after 6 months of follow-up, which is still ongoing.

To our knowledge, this is the first case report of IVL that describes the ultrasound appearance of the tumor in detail. We found that IVL exhibits multiple tracts and the rainbow sign on ultrasonography. We hope that this detailed description of the ultrasound features of IVL will facilitate the preoperative diagnosis of IVL.

Author contributions

Funding acquisition: Jianchu Li.
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