Case Report

Imaging findings of a case report of intravenous lipoleiomyomatosis

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Background: Intravenous leiomyomatosis (IVL) is a special type of uterine leiomyoma and is rare. Intravenous lipoleiomyomatosis (LPL) is a rare subtype of IVL, distinguished by the presence of adipose tissue. Although histologically benign, this disease exhibits aggressive biological behavior such as local invasion and high recurrence rate. The disease initially presents with no obvious clinical features, and cardiac symptoms may only appear in the later stages. Diagnosis primarily relies on imaging studies, and due to its rarity and atypical clinical presentation, imaging diagnosis can be challenging, leading to misdiagnosis and missed diagnosis. Previously, there was no report on the imaging findings of this disease.

Case Description: This article reports a case of a 52-year-old patient who presented with lower abdominal discomfort due to IVL, and who underwent surgical resection and had a good recovery.

Conclusions: This is the first time we report the imaging features of a disease of intravenous LPL with an extension of the inferior vena cava (IVC), and its characteristic imaging features [ultrasound shows a mass with high echogenicity, computed tomography (CT) shows low-density signal similar to fat, magnetic resonance imaging (MRI) shows high signal on T1-weighted (T1W) image and low signal on T1W with fat-suppression (T1FS)] can lead to an accurate preoperative diagnosis and guide clinical treatment.

Keywords: Lipoleiomyomatosis (LPL); imaging manifestations; adipose tissue; intravenous; case report

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Introduction

Intravenous leiomyomatosis (IVL) is a specific type of uterine leiomyoma that is classified as a benign lesion with irregular smooth muscle growth patterns, according to the World Health Organization (WHO) Classification of Tumours of Female Reproductive Organs (2014) (1). However, it exhibits aggressive biological behavior, including a tendency for local invasion and recurrence (2). The tumor can extend along the uterine veins and iliac veins, involving the inferior vena cava (IVC), and in rare cases, it can spread to the right heart and pulmonary arteries. Intravenous lipoleiomyomatosis (LPL), on the other hand, is a rare subtype of IVL. Both conditions are rare, and their coexistence is even rarer (3). LPL refers to the presence of adipose tissue within IVL. It is typically asymptomatic in the early stages but can manifest with cardiac symptoms in advanced stages (4). The diagnosis of this disease relies primarily on imaging, as its rarity and atypical clinical presentation pose challenges for accurate diagnosis, often leading to misdiagnosis. As of now, only four case reports have been identified, all of which involve early misdiagnosis and missed diagnoses. There has been no documented literature on the imaging diagnosis of this condition. The characteristic imaging features described in this article can enhance the likelihood of a preoperative

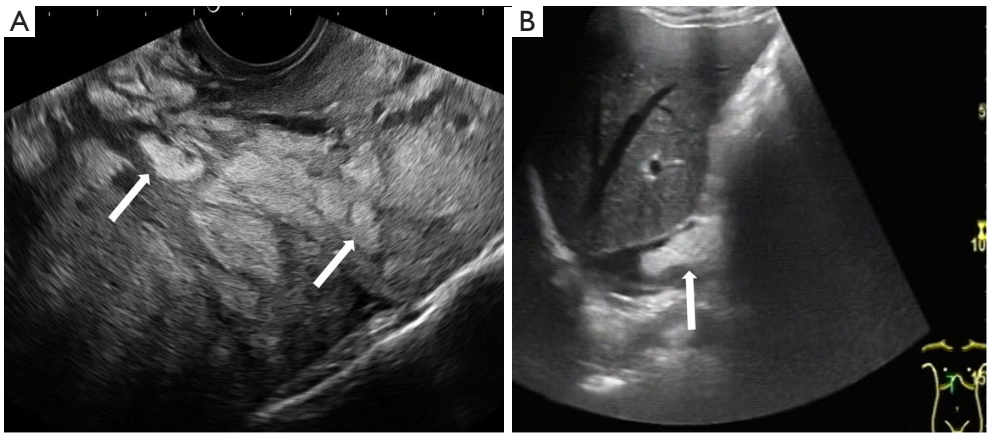

Figure 1 Ultrasonoscopy. (A) Multiple hyperechoic nodules in the uterine cavity on transvaginal ultrasound. The arrows point to the hyperechoic nodule. (B) Abdominal ultrasound showing a hyperechoic mass within the IVC. The arrow points to the hyperechoic mass. IVC, inferior vena cava.

diagnosis by physicians, enabling early detection and treatment. We present this case in accordance with the CARE reporting checklist (available at https://acr.amegroups.com/article/view/10.21037/acr-24-21/rc).

Highlight box

Key findings

- Although lipoleiomyomatosis (LPL) is rare, its imaging features are relatively significant due to the presence of adipose tissue [ultrasound shows a mass with high echogenicity, computed tomography shows low-density signal similar to fat, magnetic resonance imaging shows high signal on T1-weighted (T1W) image and low signal on T1W with fat-suppression], which are characteristic imaging findings that help us to distinguish LPL from other diseases.

What is known and what is new?

- LPL initially presents with no obvious clinical features, and cardiac symptoms may only appear in the later stages. Diagnosis primarily relies on imaging studies, and due to its rarity and atypical clinical presentation, imaging diagnosis can be challenging, leading to misdiagnosis and missed diagnosis.
- LPL has characteristic imaging findings that was first reported.

What is the implication, and what should change now?

- This is the first time we reported the imaging features of a disease of intravenous LPL with an extension of the inferior vena cava, and its characteristic imaging features can lead to an accurate preoperative diagnosis and guide clinical treatment.

Case presentation

A 52-year-old postmenopausal female patient presented to our hospital with a six-month history of lower abdominal discomfort and distension. During the physical examination, a 20 cm hard mass with poor mobility was palpated in the pelvic cavity, with no tenderness. The patient has no history of digestive or cardiovascular diseases, and no family history or genetic history. Hormonal examination revealed elevated prolactin levels at 30.67 μg/L (normal range: 2.74–19.6 μg/L). Subsequent transvaginal ultrasound examination revealed an irregularly shaped, well-defined, hyperechoic mass measuring 169 mm × 77 mm in the pelvic cavity, with unclear demarcation from the uterus. Multiple slightly hyperechoic nodules of varying sizes were observed inside (*Figure 1A*), suggesting a pelvic solid mass with undetermined nature. Abdominal ultrasound further indicated an irregularly shaped hyperechoic mass measuring approximately 46 mm × 17 mm within the posterior segment of the hepatic end of the IVC, suggestive of an intraluminal mass (*Figure 1B*). Given the presence of an intraluminal mass in the IVC, further investigations were performed, including contrast-enhanced computed tomography (CT) of the upper abdomen and pelvic magnetic resonance imaging (MRI). CT scan showed a long, strip-like, mixed low-density lesion within the IVC, containing fat density signals (*Figure 2A*); contrast-enhanced CT transverse section (*Figure 2B,2C*) and coronal section

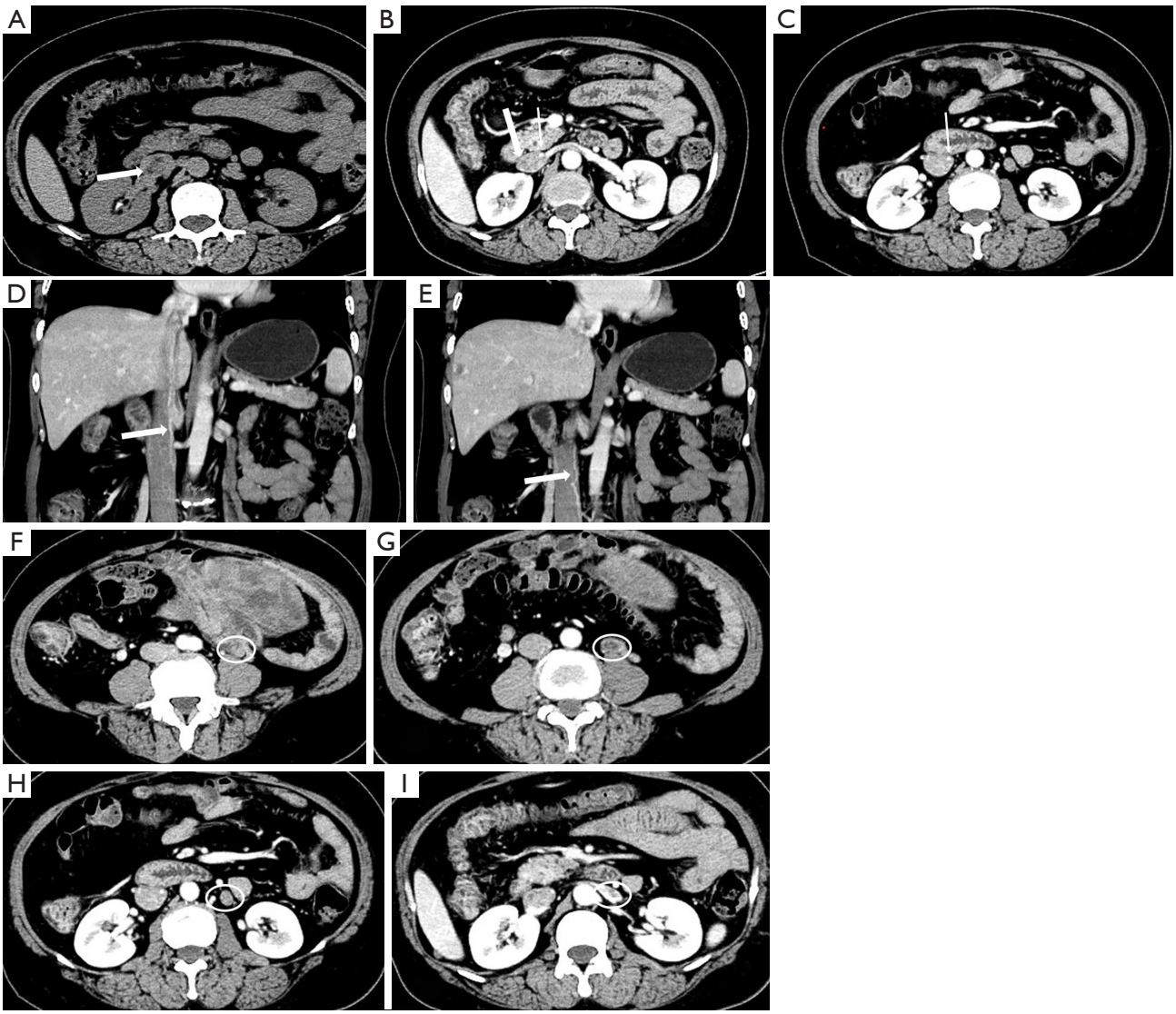

Figure 2 CT images. (A) The arrow points to a low-density occupying lesion within the IVC containing fat density. (B,C) Arterial phase of CT transverse section, thick arrow: IVC filling defect; thin arrow: small vessel visualization. (D,E) CT coronary artery phase, small vessel visualization. The arrow points to a small vessel visualization. (F-I) CT cross-sectional arterial phase of a pelvic venous filling defect extending upward from the vicinity of the pelvic mass to the left renal vein; the picture sequence is caudal to cranial of the patient. The filling defect of the pelvic vein indicated by the circle. CT, computed tomography; IVC, inferior vena cava.

(*Figure 2D*,*2E*) revealed a filling defect in the IVC with visualization of a long, thin, enhancing vessel. Additionally, a filling defect was observed in the pelvic vein extending upward to the left renal vein, subsequently joining the IVC (*Figure 2F-2I*). On T2-weighted image with fat-suppression (T2FS) MR Images, irregular mixed signal masses were found in the pelvic cavity (*Figure 3A*). Some areas containing adipose tissue showed high signal intensity on T1W images (*Figure 3B*), but this area was suppressed as low signal on the T1W image with fat-suppression (T1FS) (*Figure 3C*). Based on the aforementioned imaging findings, the diagnosis of IVL involving the pelvic veins and IVC was considered.

Following consultation, it was decided that the gynecologist would perform a pelvic mass resection surgery along with total hysterectomy and bilateral salpingo-

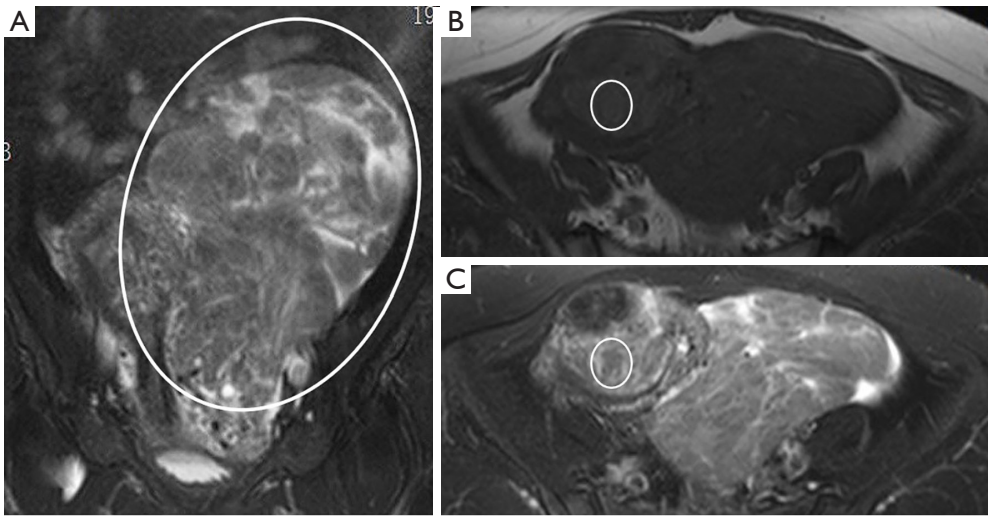

Figure 3 Magnetic resonance imaging. (A) The circle refers to an irregular mixed signal masse in the pelvic cavity on coronal T2FS. (B) The circle refers to adipose tissue in the mass with high signal intensity on T1W images. (C) The circle refers to adipose tissue in the mass with suppressed and hypointense on T1FS. T2FS, T2-weighted image with fat-suppression; T1W, T1-weighted; T1FS, T1-weighted image with fat-suppression.

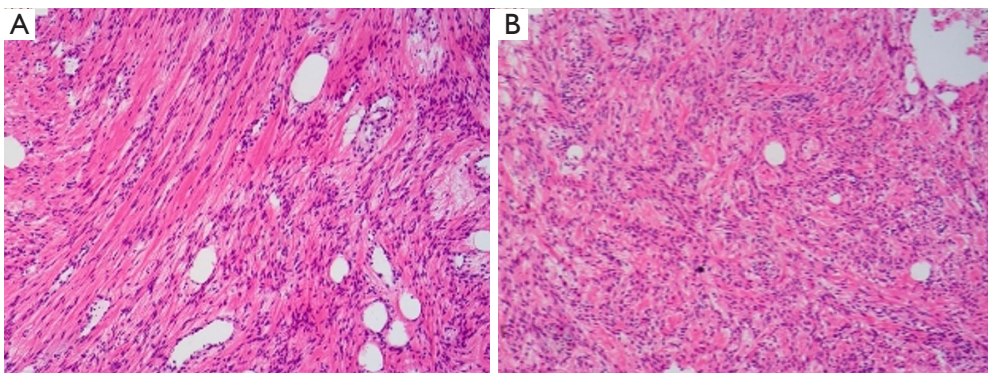

Figure 4 Images of pathological sections (hematoxylin-eosin, 10×). (A) The pathological section shows a large number of spindle cells of smooth muscle tissue, which are mixed with adipocytes. (B) The spindle cells on the left of image (A) are arranged in a regular braid, while the spindle cells on the right and image (B) are arranged disorderly and intertwined in a vortex.

oophorectomy. Intraoperative frozen section pathology (*Figure 4A*,*4B*) suggested a smooth muscle-derived mesenchymal tumor. Intraoperatively, a tumor measuring approximately 20 cm × 20 cm × 18 cm was palpated at the left broad ligament of the uterus. The tumor was solid, irregular in shape, extending posteriorly to the back of the uterus, with a complete capsule, smooth surface, and no adhesions. The left ovary and fallopian tube were attached to the surface of the tumor, while the uterus and right ovary and fallopian tube appeared normal. Postoperative pathology immunohistochemistry results showed that the tumor contained muscle, vascular, and fat components, suggesting the LPL. Subsequently, vascular surgery performed a resection of the IVC tumor and pelvic vein: after isolating the IVC (from the level of the hepatic vein to the level of the renal vein), both ends were bagged separately. A longitudinal incision was made from the level of the hepatic vein to the level of the renal vein, allowing the tumor to be extracted slowly by gentle traction. After the surgery, the patient received anti-estrogen therapy and experienced a good recovery. Subsequent follow-ups over a period of 5 years showed no recurrence.

All procedures performed in this study were in accordance with the ethical standards of The First Affiliated Hospital of Ningbo University Lung-approved 2024 Research No. 033RS and with the Helsinki Declaration (as revised in 2013). Publication of this case report and accompanying images was waived from patient consent according to the ethics committee of the First affiliated Hospital of Ningbo University.

Discussion

IVL is a special type of uterine leiomyoma, characterized by the growth of uterine smooth muscle tumors into the veins or the protrusion of smooth muscle tissue from the vessel wall into the lumen (5). The tumor often invades adjacent uterine venous or parametrial veins. It predominantly occurs in postmenopausal women, and most patients have a history of uterine leiomyoma or previous uterine surgery. Elevated estrogen levels, venous congestion, and local injury (such as a history of uterine surgery) may be important factors contributing to the development of this disease (6). IVL is histologically characterized by the benign growth of smooth muscle cells within the lumen of veins or lymphatic vessels, but it exhibits biological behavior similar to malignant tumors, with a tendency for recurrence and dissemination. Some studies have indicated that IVL shares certain molecular and cytogenetic characteristics with uterine leiomyomas, but its gene expression profile is more similar to that of smooth muscle sarcomas (2).

LPL are a rare subtype of intravenous IVL, primarily composed of fat, smooth muscle, and blood vessels. The clinical symptoms caused by this disease are similar to IVL. In the early stages, although there is extensive venous dilation, the expansion of the tumor remains within the small blood vessels of the muscle layer, and cannot be detected by CT or MRI. It is easily misdiagnosed as a common uterine leiomyoma in the early stages, with clinical symptoms mainly manifesting as menstrual changes, lower abdominal pain, and palpable pelvic masses. When the disease invades the cardiovascular system in the late stages, patients may experience symptoms such as chest pain, dyspnea, lower limb edema, syncope, and sudden death (7).

The clinical symptoms of this disease are atypical, but its characteristic imaging findings can help with diagnosis. Due to the presence of fat components, ultrasound shows higher echogenicity (while venous leiomyomas generally show lower echogenicity), similar to renal angiomyolipomas. In this case, transvaginal ultrasound of the uterus revealed nodules of varying sizes with slightly higher echogenicity, which were actually leiomyomas containing adipose tissue (as leiomyomas typically show low echogenicity). Abdominal ultrasound showed a high echogenic mass within the IVC, which was also due to the presence of adipose tissue within the tumor, resulting in high echogenicity (as thrombi usually show low or moderate echogenicity). CT and MRI findings of fat and vascular tissue are also important diagnostic tools. The contrast-enhanced CT scan revealed a large filling defect within the IVC, containing low-density fatty components. Post-contrast imaging did not show significant enhancement of the fatty component, but a slender small vessel within it demonstrated marked enhancement. MRI showed that the soft tissue mass had high signal intensity on T1W images and was completely suppressed on T1FS images, further indicating that the mass was not just a leiomyoma but a lipoma-like liposarcoma containing fat tissue. Additionally, a filling defect in the pelvic vein extending upward to the left renal vein and draining into the IVC was identified. Intraoperatively, the uterus was found to be normal in size and morphology, with the mass located at the broad ligament of the uterus. From this, we infer that the primary site of the lesion is located in the broad ligament of the uterus within the pelvic cavity, then it spreads through the pelvic vein and extends into the IVC. Based on these imaging findings, which revealed the presence of fat and vascular tissue within the mass, along with the involvement of the pelvic and IVC veins, the diagnosis of LPL is considered appropriate. It is crucial to differentiate how filling defects in the IVC are not misdiagnosed as poor opacification of contrast agent during enhancement. If there is contrast agent filling in the distal part of the lesion but not in the proximal part, then we can confirm the presence of an occupying lesion in the IVC causing the filling defect rather than incomplete opacification of the contrast agent. Additionally, when the left renal vein joins the IVC, turbulence is created that may appear as a filling defect. How can we distinguish between turbulence and a defect caused by an occupying lesion? Turbulence-induced defects are generally irregular in shape and only present at the entrance of the turbulence, while defects caused by an occupying lesion have clear boundaries and are present throughout the section of the occupying lesion, requiring careful observation for differentiation.

The differential diagnosis of LPL mainly includes venous lipoma, intravenous liposarcoma, venous thrombosis, and other diseases. Venous lipoma has clear margins, appears as high signal intensity on T1W and T2W images, and the

entire fat mass is completely suppressed on T1FS images (8), similar to the presentation in this case. However, venous lipoma does not involve neurovascular structures and does not have septations or thin septa. Intravenous liposarcoma on MR imaging features a partially ill-defined margin, neurovascular involvement, enhancing thick/nodular septum, and a partially bright signal intensity on T1W images (9). The radiological findings in this case are very similar to those of liposarcoma, making differentiation challenging. This also supports the view that the genetic expression profile of LPL is closer to that of leiomyosarcoma. LPL is relatively easy to differentiate from venous thrombosis, as thrombi do not enhance after contrast agent administration while LPL does enhance.

In the pathological section, we observed a significant number of spindle-shaped cells arranged in a partially regular woven pattern, with some areas exhibiting disordered arrangement intertwining in a vortex shape, suggesting that the mass originates from smooth muscle. Interspersed among them are numerous adipocytes, indicating the presence of adipose tissue within the mass. Immunohistochemical results demonstrate ERG (+), CD31 (+), D2-40 (+) expression, indicating the tumor's association with angiogenesis; S-100 (+) expression indicates the presence of lipomatous components; while positive expression of Desmin and Caldesmon suggests the existence of muscle cells. Positive estrogen receptor (ER) and progesterone receptor (PR) staining implies that the tumor is receptive to estrogen and progesterone receptors. The aforementioned findings suggest that the tumor comprises three components: muscle, blood vessels, and fat, leading to its pathological diagnosis as LPL.

The most commonly used treatment method currently is surgical removal of the tumor and postoperative anti-estrogen therapy. Complete removal of the tumor and removal of the ovaries are key to avoiding IVL recurrence (10). Anti-estrogen therapy is reasonable for those who have been found to have estrogen and progesterone receptors in the tumor's veins and cytoplasm (11). Tamoxifen is commonly used for treatment. In this case, pathology revealed positive staining for both estrogen and progesterone by immunohistochemistry, so anti-estrogen therapy was administered after surgery. Tumors recur in up to 30% of cases, and young age, large initial tumors, and involvement of the major veins may be high-risk factors for recurrence. Therefore, long-term follow-up of 3–6 months after surgery is necessary (12). The most common site of subsequent metastasis is the lungs (13). Follow-up should include reproductive hormone testing, ultrasound examination of the cardiovascular system and pelvis, and CT examination of the lungs (5).

Due to the rarity of the disease, the diagnosis of LPL and IVL is very difficult and often misdiagnosed. The imaging findings in this case are typical, but this is only a case report and more case studies are needed to confirm the diagnosis.

Patient's perspective

I was satisfied with the outcome of my treatment and I hope that my case will encourage physicians to study this disease and reduce the chance of missed diagnosis and misdiagnosis.

Conclusions

In summary, although LPL is rare, its imaging features are relatively significant due to the presence of adipose tissue (ultrasound shows a mass with high echogenicity, CT shows low-density signal similar to fat, MR shows high signal on T1W image and low signal on T1FS), which are characteristic imaging findings that help us to distinguish LPL from other diseases.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://acr.amegroups.com/article/view/10.21037/acr-24-21/rc

Peer Review File: Available at https://acr.amegroups.com/article/view/10.21037/acr-24-21/prf

Conflicts of Interest: Both authors have completed the ICMJE uniform disclosure form (available at https://acr.amegroups.com/article/view/10.21037/acr-24-21/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of The First Affiliated Hospital of Ningbo University Lung-approved 2024 Research No. 033RS and with the Helsinki Declaration (as revised in 2013). Publication of this case report and accompanying images was waived from patient consent according to the ethics committee of the first affiliated hospital of Ningbo university.

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