Journal of Geriatric Cardiology | JGC
LETTER TO THE EDITOR
J Geriatr Cardiol 2025; 22(12): 992–994

Intravenous leiomyomatosis presenting as pulmonary embolism: a cardiovascular perspective in two cases

Xia WU*, Shi-Jie YANG*, Ying LIANG✉

Emergency Critical Care Center, Beijing Anzhen Hospital, Capital Medical University, Beijing, China
*The authors contributed equally to this manuscript
✉ Correspondence to: cp_liangying@126.com
https://doi.org/10.26599/1671-5411.2025.12.008

Intravenous leiomyomatosis (IVL) is a rare, histologically benign uterine smooth muscle tumor with malignant biological behavior due to its propensity for intravascular extension.[1] While gynecological in origin, its most severe manifestations are cardiovascular, arising from tumor propagation through the venous system into the inferior vena cava (IVC), right heart, and pulmonary arteries, mimicking thromboembolic disease.[2,3] This can lead to pulmonary embolism (PE), right heart obstruction, and even sudden cardiac death.[4] Diagnosis is challenging, often delayed by misdiagnosis as conventional PE. We present two cases of IVL initially presenting with PE, highlighting the critical cardiovascular implications and diagnostic pitfalls.

Case 1 A 49-year-old postmenopausal woman with history of hypertension and prior total abdominal hysterectomy for uterine fibroids presented with a one-year history of progressive exertional dyspnea, culminating in a presyncopal episode. Initial computed tomographic pulmonary angiogram (CTPA) at a local hospital revealed filling defects in the main and branch pulmonary arteries, prompting a referral for suspected PE. Upon admission, vital signs were stable. Laboratory tests showed mildly elevated D-dimer (439 ng/mL). Electrocardiogram revealed an $S_1Q_3T_3$ pattern and T-wave inversions in leads V1-V4 (Figure 1). Crucially, transthoracic echocardiography demonstrated multiple cystic masses within the right atrium, mild right atrial enlargement, moderate tricuspid regurgitation, and mild pulmonary hypertension. CTPA identified multiple filling defects in the segmental and subsegmental branches of both pulmonary arteries, consistent with multiple pulmonary emboli. Additionally, low-density lesions within the right atrial and ventricular cavities were noted, suggestive of a space-occupying lesion requiring clinical correlation (Figure 2). This finding, inconsistent with bland thrombus, raised strong suspicion for IVL with intracardiac extension. The patient underwent surgical resection (bilateral oophorectomy and ovarian vein tumor removal). Histopathology confirmed IVL, with immunohistochemistry positive for desmin, estrogen receptor, progesterone receptor, and smooth muscle actin. The final diagnosis was stage IV IVL presenting as chronic PE. This case underscores that right heart masses and signs of pulmonary hypertension in a patient with prior uterine pathology should alert clinicians to the possibility of IVL, a critical cardiovascular mimic of thromboembolic disease.

Case 2 A 26-year-old primigravida at nine weeks of gestation presented with acute-onset pleuritic right-sided chest pain and dyspnea. CTPA confirmed acute segmental and subsegmental pulmonary emboli in the right lung (Figure 3). Despite immediate initiation of therapeutic anticoagulation with enoxaparin, serial imaging revealed persistent filling defects extending from the left ovarian vein into the left iliac venous system and IVC. These lesions showed no significant reduction in size, which was atypical for acute thrombosis. Bedside Doppler ultrasound and contrast-enhanced computed tomography venography detailed the extent of the intravascular material. Positron emission tomography/computed tomography showed physiologic fluorodeoxyglucose uptake (SUV_{max} = 1.7) in the IVC lesion, favoring a benign process but not excluding tumor (Figure 4). Given the anticoagulation-refractory nature of the "thrombus" and its continuity with the pelvic venous system, a vascular smooth muscle tumor was suspected. The patient underwent IVC filter placement and iliac vein thrombectomy, which intraoperatively suggested IVL. This case highlights a cardiovascular diagnostic challenge in

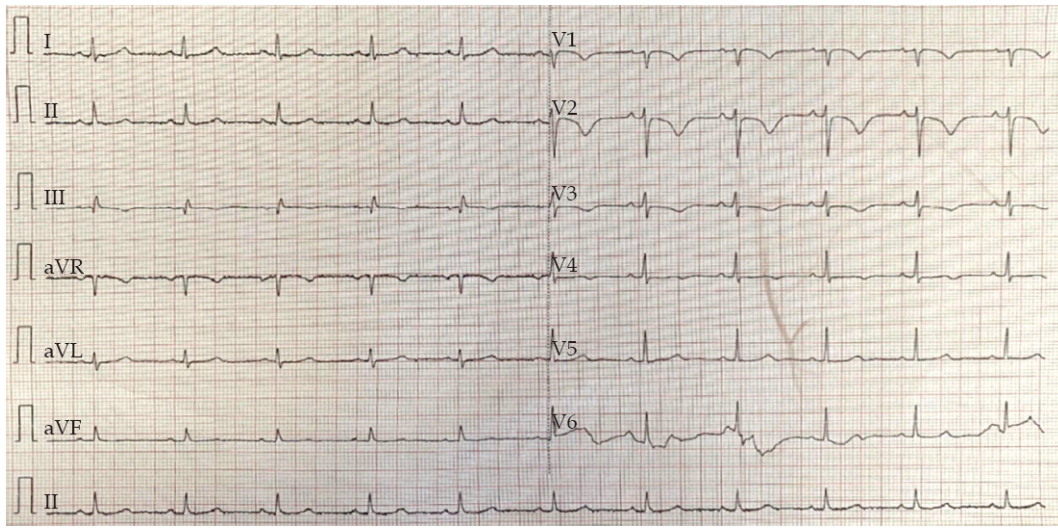

Figure 1 The electrocardiogram showed sinus rhythm with the $S_1Q_3T_3$ pattern, T-wave inversions in precordial leads V1-V4, and T-wave flattening in lead aVF.

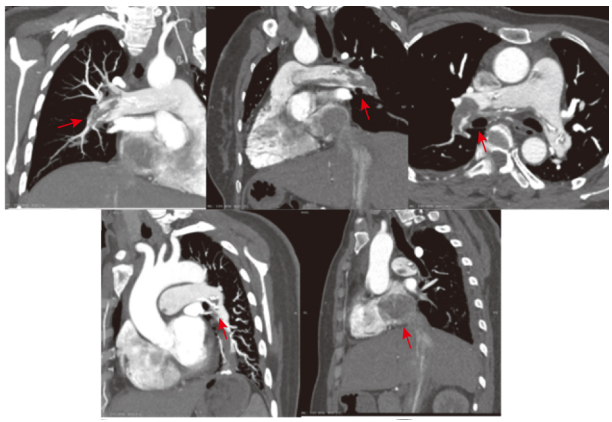

Figure 2 Computed tomographic pulmonary angiogram revealed multiple filling defects (red arrows) in the segmental and subsegmental branches of both pulmonary arteries.

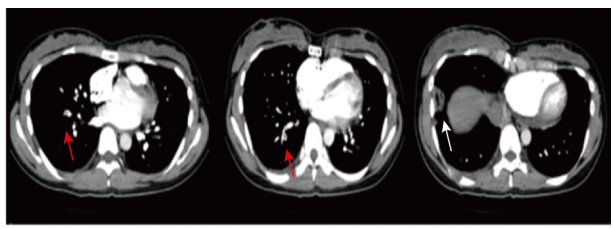

Figure 3 Computed tomographic pulmonary angiogram revealed multiple filling defects (red arrows) in both pulmonary arteries and consolidation in the right lower lung (white arrow).

pregnancy: IVL can masquerade as acute PE. The key clue was the failure of the intravascular filling defects to resolve with anticoagulation, prompting consideration of a tumorous etiology and necessitating a multidisciplinary vascular-gynecological approach to secure the diagnosis and plan definitive management.

These cases underscore IVL as a formidable cardiovascular mimicker. Tumor extension via the uterine or ovarian veins into the IVC and right heart (intracardiac leiomyomatosis) can cause PE, right ventricular inflow obstruction, tricuspid regurgitation, and pulmonary hypertension.[3-5] The initial presentation is frequently cardiovascular, leading to misdiagnosis as acute or chronic PE, as seen in both our patients. Key diagnostic clues include a history of uterine fibroids or hysterectomy, persistent intravascular "thrombus" refractory to anticoagulation, and echocardiographic findings of a mobile, serpentine right heart mass

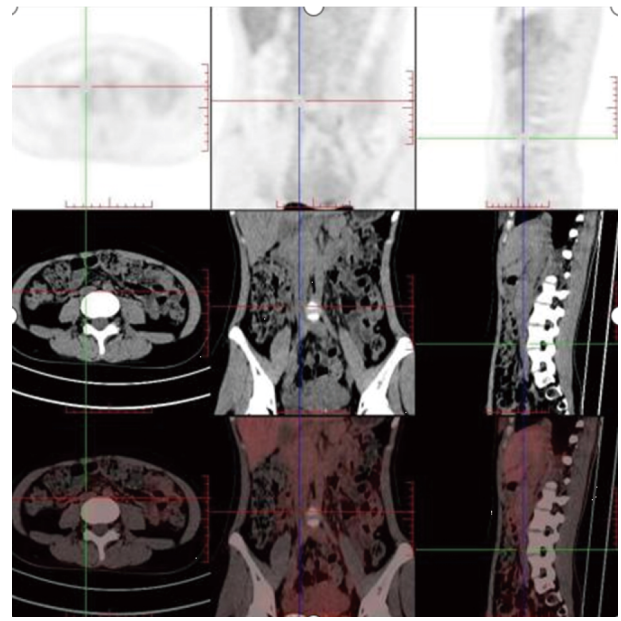

Figure 4 Positron emission tomography/computed tomography showed a low-density lesion was observed in the lower segment of the inferior vena cava extending to the left common iliac vein, showing general radiopharmaceutical uptake with an SUV_{max} of 1.7.

contiguous with the IVC.[6,7] Multimodal imaging (echocardiography, computed tomography angiography/magnetic resonance imaging venography) is crucial for mapping tumor extent and planning intervention.[8,9]

From a cardiovascular management perspective, complete surgical resection remains the cornerstone to prevent fatal embolization or obstruction.[10] For intracardiac leiomyomatosis, a coordinated single-stage (cardiothoracic and abdominal) or two-stage surgical approach is required, involving cardiopulmonary bypass for intracardiac tumor extraction.[11,12] Perioperative risks include massive hemorrhage and tumor embolization, necessitating a heart team approach. Hormonal therapy (gonadotropin-releasing hormone agonists/aromatase inhibitors) can be adjunctive but is not curative.[13] Long-term cardiovascular surveillance is essential due to recurrence rates of 16%–30%.[14-16] Recurrence can manifest as recurrent PE or right heart failure, emphasizing the need for periodic echocardiography and cross-sectional imaging.[16,17]

In conclusion, IVL is a rare condition with predominantly cardiovascular presentations. Cardiologists should consider IVL in the differential diagnosis of PE, particularly in women with a history of uterine fibroids and intravascular masses refractory to anticoagulation. Early recognition, multimodal imaging, and multidisciplinary management involving cardiology, cardiac surgery, and gynecology are vital to mitigate life-threatening complications and guide definitive therapy.

ACKNOWLEDGMENTS

This work was supported by the Capital Medical University Undergraduate Innovation Project 2025 (XSKY20-25251). All authors had no conflicts of interest to disclose.

REFERENCES

[1] Demirkiran F, Sal V, Kaya U, *et al*. Intravenous leiomyoma with extension to the heart: a case report and review of the literature. *Case Rep Obstet Gynecol* 2013; 2013: 602407.
[2] Hanke H, Lenz C, Finking G. The discovery of the pathophysiological aspects of atherosclerosis: a review. *Acta Chir Belg* 2001; 101: 162–169.
[3] Norris HJ, Parmley T. Mesenchymal tumors of the uterus. V. Intravenous leiomyomatosis. A clinical and pathologic study of 14 cases. *Cancer* 1975; 36: 2164–2178.
[4] Clement PB. Intravenous leiomyomatosis of the uterus. *Pathol Annu* 1988; 23: 153–183.
[5] Fornaris RJ, Rivera M, Jiménez L, *et al*. Multimodality evaluation of intravenous leiomyomatosis: a rare, benign but potentially life-threatening tumor. *Am J Case Rep* 2015; 16: 794–800.
[6] Lam PM, Lo KW, Yu MY, *et al*. Intravenous leiomyomatosis: two cases with different routes of tumor extension. *J Vasc Surg* 2004; 39: 465–469.
[7] Li B, Chen X, Chu YD, *et al*. Intracardiac leiomyomatosis: a comprehensive analysis of 194 cases. *Interact Cardiovasc Thorac Surg* 2013; 17: 132–138.
[8] Nakai G, Maeda K, Yamamoto K, *et al*. Uterine intravenous leiomyomatosis with cardiac extension: radiologic assessment with surgical and pathologic correlation. *Case Rep Obstet Gynecol* 2015; 2015: 576743.
[9] Quade BJ, Dal Cin P, Neskey DM, *et al*. Intravenous leiomyomatosis: molecular and cytogenetic analysis of a case. *Mod Pathol* 2002; 15: 351–356.
[10] Du J, Zhao X, Zhang X, *et al*. A clinicopathologic study of intravenous leiomyomatosis: a series of 23 cases. *Eur J Gynaecol Oncol* 2014; 35: 399–404.
[11] Riess FC, Dobritzsch B, Kormann J, *et al*. Lepirudin for cardiopulmonary bypass surgery in a patient with terminal renal insufficiency and acute heparin-induced thrombocytopenia. *Thorac Cardiovasc Surg* 2006; 54: 140–142.
[12] Du J, Zhang X, Li W, *et al*. Intravascular leiomyomatosis: a review of clinical and surgical management. *Eur J Obstet Gynecol Reprod Biol* 2020; 245: 54–59.
[13] Li B, Chen X, Li F, *et al*. Intravenous leiomyomatosis of the uterus: a clinicopathological analysis of 76 cases. *Chin Med J (Engl)* 2010; 123: 1991–1996.
[14] Clay TD, Riccio MC, Khabiri H, *et al*. One-stage versus two-stage resection of intravascular leiomyomatosis with intracardiac extension: a systematic review and meta-analysis. *J Card Surg* 2022; 37: 5353–5361.
[15] Wang J, Yang J, Huang H, *et al*. Management of intracardiac leiomyomatosis: a 10-year single-institution experience. *J Thorac Cardiovasc Surg* 2014; 147: e1–e3.
[16] Liu F, Zhang H, Li Y, *et al*. Recurrence and risk factors of intravenous leiomyomatosis: a retrospective single-center study. *Orphanet J Rare Dis* 2022; 17: 45.
[17] Clay TD, Khabiri H, Sanaiha Y, *et al*. Follow-up and surveillance for intravascular leiomyomatosis: a systematic review. *Gynecol Oncol Rep* 2021; 38: 100875.

Please cite this article as: WU X, YANG SJ, LIANG Y. Intravenous leiomyomatosis presenting as pulmonary embolism: a cardiovascular perspective in two cases. *J Geriatr Cardiol* 2025; 22(12): 992–994. DOI: 10.26599/1671-5411.2025.12.008