Unusual Case of Intravenous Leiomyomatosis with Intracardiac Extension and Pulmonary Metastases

Ng Jing Long Raymond*, Guo Shuyi, Haja Mohideen Salahudeen Mohamed

Singapore General Hospital, Singapore

***Correspondence:** Ng Jing Long Raymond, Singapore General Hospital. Outram Road. Singapore 169608

ngjinglong@hotmail.com

Radiology Case. 2025 March; 19(3):1-7 :: DOI: 10.3941/jrcr.5530

Authors' Contributions
Ng Jing Long Raymond: Methodology, Writing – original draft, Data curation, Writing—review and editing, Visualization
Guo Shuyi: Methodology, Conceptualization, Resources, Supervision, Writing—review and editing, Visualization.
Haja Mohideen Salahudeen Mohamed: Methodology, Conceptualization, Resources, Supervision, Writing—review and editing, Visualization.

Acknowledgements
None.

Disclosures
None.

Consent
Yes.

Human And Animal Rights
Not applicable.

ABSTRACT

Intravenous leiomyomatosis and benign metastasizing leiomyomatosis are subsets of extrauterine leiomyomas, a rare condition that also includes disseminated peritoneal leiomyomatosis, parasitic leiomyoma, and retroperitoneal leiomyomatosis. We present an unusual case of intravenous leiomyomatosis with intracardiac extension and pulmonary metastases in a 52-year-old female.

CASE REPORT

CASE REPORT

A 52-year-old, previously healthy female presented in 2010 with abdominal distension lasting several months. Computed tomography (CT) of the abdomen and pelvis showed a massive heterogeneously enhancing mass arising from the uterine fundus (Figures 1A,1B) extending superiorly up to the upper abdomen. Small filling defects in the left renal veins (Figures 2A,2B) were observed retrospectively.

She subsequently underwent a hysterectomy and left salpingo-oophorectomy. Histology revealed leiomyoma with tumor extension into vessels within the broad ligament. Subsequent yearly follow up pelvic ultrasound studies did not show any evidence of local recurrence in the pelvis before she was lost to follow up in 2017.

She re-presented to the emergency department in 2023 with right iliac fossa pain. CT demonstrated a similar appearing heterogeneously enhancing soft tissue mass in the right hemipelvis (Figure 3) extending up to the mid abdomen. The mass was in close proximity to the left ovarian vein which exhibited several small filling defects (Figure 4A). Filling defects were also present in the left renal vein (Figure 4C) and in the right atrium with coarse calcifications (Figure. 5A). A cluster of small solid pulmonary nodules (Figure 5B,5C) was noted in the right lower lobe.

The patient subsequently underwent a CT-guided biopsy of the dominant right lower lobe pulmonary nodule, with histology yielding benign metastasizing leiomyoma. She was commenced

on parenteral leuprorelin and oral letrozole therapy. At the time of this case report, the patient was not keen on any surgical intervention.

DISCUSSION

Intravenous leiomyomatosis (IVL) and benign metastasizing leiomyomatosis (BML) are subsets of extrauterine leiomyomas, a rare condition that also includes disseminated peritoneal leiomyomatosis, parasitic leiomyoma and retroperitoneal leiomyomatosis [1]. These conditions predominantly affect women of reproductive age, with a median age of 48 years [2].

IVL is an aggressive benign smooth muscle tumor believed to arise from the direct extension of a leiomyoma into the pelvic veins [3]. Mural invasion is uncommon. IVL can extend variable distances superiorly, involving the inferior vena cava, right atrium, and even the pulmonary arteries.

BML, on the other hand, is a benign smooth muscle tumor, typically involving the lungs, although other sites, such as the abdomen, skin, mediastinum and nervous system have also been reported [4]. In BML, the tumor is hypothesized to spread to the lungs via pulmonary embolism as some case reports describe imaging findings consistent with BML but without features of IVL.

The diagnosis of both conditions requires a high degree of clinical suspicion. Clinical presentation can vary widely, ranging from asymptomatic cases to abdominal pain, heart failure or shortness of breath due to pulmonary embolism [2,3]. Instances of sudden death have also been reported [2,3].

Recurrent or metastatic disease often presents a diagnostic challenge and may be underdiagnosed [2]. Prior history of hysterectomy for leiomyomas can be a useful clue. As illustrated in our case, a recurrent mass with similar morphology involving the remnant ovarian vein was an important diagnostic indicator. Furthermore, the presence of tumor thrombi in the right atrium and contiguity of the pulmonary nodules to the pulmonary arterial branches helped to clinch the diagnosis of pulmonary metastatic leiomyoma.

Given the rarity of these conditions, there are no established standard treatment guidelines. However, surgical resection is generally considered the treatment of choice [5]. Long term management with gonadotropin-releasing hormone agonists and long-term regular follow-up are also recommended to prevent recurrence and monitor for new lesions [5].

TEACHING POINT

Given their rarity, IVL and BML necessitate a high level of clinical suspicion for accurate diagnosis. In patients with a history of hysterectomy for leiomyomas, the presence of intravenous filling defects or solid pulmonary nodules should raise the consideration of IVL and BML in the differential diagnosis.

QUESTIONS AND ANSWERS

1. Which is the most common site of metastasis for benign metastasizing leiomyomas?
   A. Central nervous system
   B. Lung
   C. Liver
   D. Bladder

Answer: (B). The lungs are the most common site of metastasis for benign metastasizing leiomyomas.

2. Benign metastasizing leiomyomas of the lung can appear as which of the following on CT?
   A. Solitary solid pulmonary nodule
   B. Multiple solid pulmonary nodules
   C. Cavitating pulmonary nodules
   D. All of the above

Answer: (D). All of the above imaging findings can be seen in benign metastasizing leiomyomas of the lung.

3. In which age group are intravenous leiomyomatosis and benign metastasizing leiomyomas most commonly seen?
   A. Perimenopausal
   B. Under age 30
   C. Post menopausal
   D. Similar incidence across all age groups

Answer: (A). Extrauterine leiomyomatosis is most commonly seen in perimenopausal females.

4. Which of the following correctly describes the imaging finding of intravenous leiomyomatosis?
   A. Ultrasound – Non vascularised thrombus in IVC
   B. CT – Right atrial filling defect with calcification
   C. MRI – High T2W signal lesion in the IVC with enhancement and restricted diffusion
   D. None of the above

Answer: (B). Intravenous leiomyomatosis appears as vascularised thrombus on ultrasound. Option (C) describes features of leiomyosarcoma.

5. Which of the following best describes the signal characteristics of extrauterine leiomyomas on MRI?
   A. High T1W, low T2W and homogenous contrast enhancement
   B. High T1W, high T2W and heterogenous contrast enhancement
   C. Intermediate T1W, low T2W and homogenous contrast enhancement
   D. Intermediate T1W, intermediate T2W and homogenous contrast enhancement

Answer: (C). Low T2W signal intensity is characteristic of extrauterine leiomyomas, mimicking that of smooth muscle.

Homogenous contrast enhancement and low to intermediate T1W signal are usually seen.

REFERENCES

1. Fasih N, Prasad Shanbhogue AK, Macdonald DB, et al. Leiomyomas beyond the uterus: Unusual locations, rare manifestations. *Radiographics.* 2008; 28(7): 1931-1948. PMID: 19001649.

2. Clay TD, Dimitriou J, McNally OM, Russell PA, Newcomb AE, Wilson AM. Intravenous leiomyomatosis with intracardiac extension – A review of diagnosis and management with an illustrative case. *Surg Oncol*. 2013; 22(3): e44-e52. PMID: 23642379.

3. Wei JL, Ji X, Zhang P, Chen WJ, Zhao YN, Liu M. Complete intravenous leiomyomatosis: a case report and literature review. *Ann Palliat Med*. 2021; 10(11): 12039-12045. PMID: 34872328.

4. Adair LB 2nd. CT findings of pathology proven benign metastasizing leiomyoma. *Radiol Case Rep.* 2020; 15(11): 2120-2124. PMID: 32944110.

5. Chin H, Ong XH, Yam PK, Chern BS. Extrauterine fibroids: a diagnostic challenge and a long-term battle. *BMJ Case Rep.* 2014; 2014: bcr2014204928. PMID: 25395465.

6. Bednarova I, Frellesen C, Roman A, Vogl TJ. Case 257: Leiomyosarcoma of the Inferior Vena Cava. *Radiology.* 2018; 288(3): 901–908.

FIGURES

Figure 1: Pre-operative Contrast enhanced CT abdomen and pelvis. Axial (A) and sagittal (B) images show a very large heterogeneous abdominopelvic mass arising from the uterus, containing several small nodular enhancing foci.

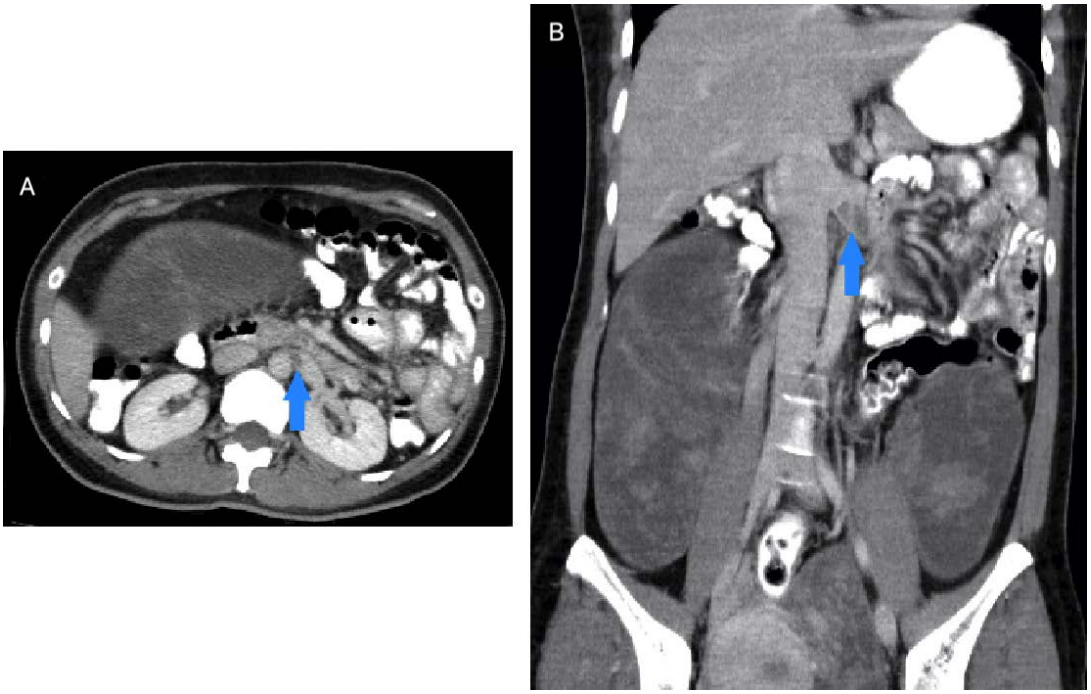

Figure 2: Pre-operative Contrast enhanced CT abdomen and pelvis. Axial (A) and coronal (B) images demonstrate partial filling defects in the left renal vein (blue arrows).

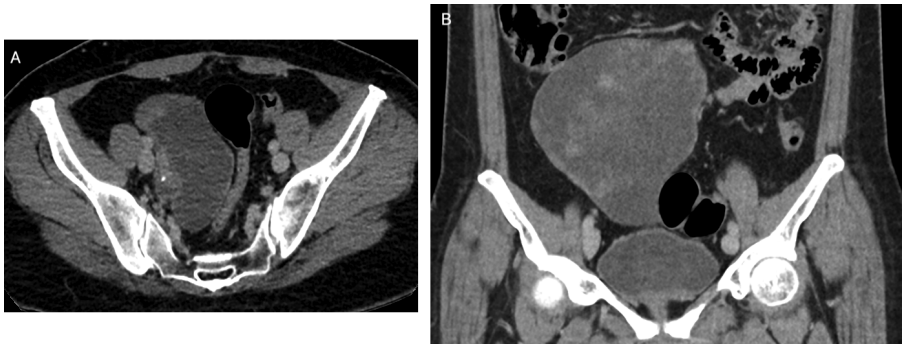

Figure 3: Patient re-presented years later with right iliac fossa pain. Contrast enhanced CT abdomen and pelvis, axial (A) and coronal (B) images demonstrating recurrent heterogenous soft tissue mass in the right hemipelvis, inseparable from the right ovary.

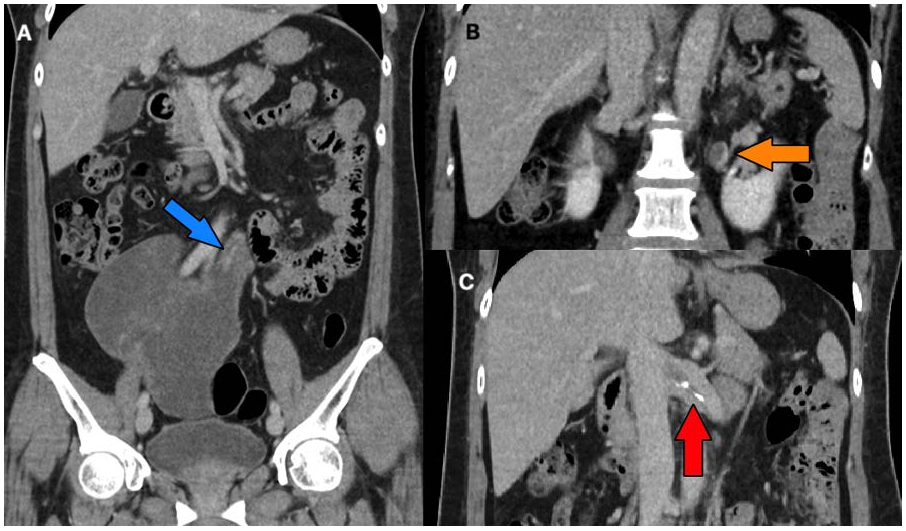

Figure 4: Contrast enhanced CT abdomen and pelvis. Coronal image (A) shows a heterogeneous right hemipelvic mass with a contiguous filling defect in the adjacent left ovarian vein (blue arrow). Coronal images (B) and (C) show partial filling defects in the left ovarian vein (orange arrow) and left renal vein (red arrow), the latter associated with coarse calcifications.

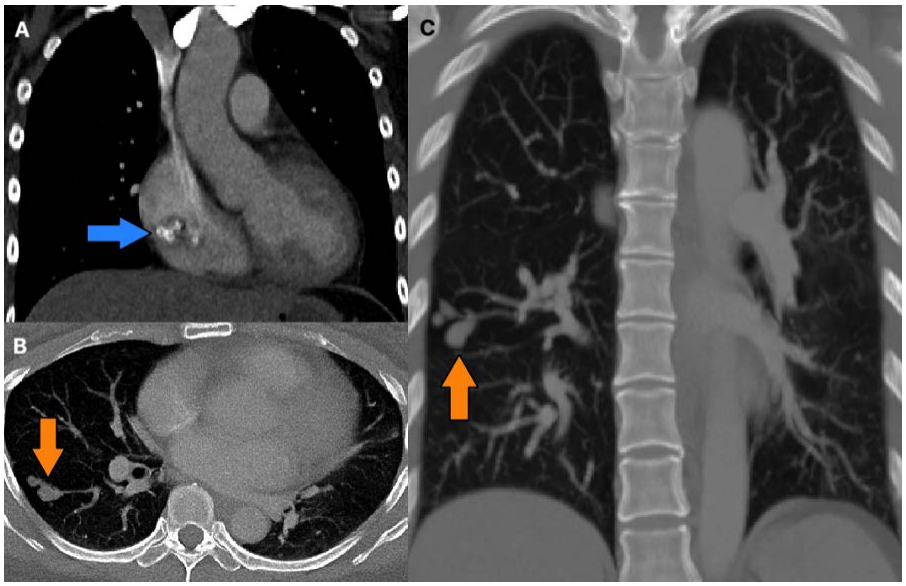

Figure 5: Findings: Contrast enhanced CT chest. Coronal (A) soft tissue reconstructed image shows filling defect in the right atrium with associated calcifications (blue arrow). Axial (B) and coronal (C) MIP lung images show small cluster of solid pulmonary nodules in the right lower lobe in close proximity to the pulmonary arterial branches (orange arrows).

SUMMARY TABLE

	Aetiology	**Incidence**	**Gender ratio**	**Age predilection**	**Risk factors**	**Treatment**	**Prognosis**	**Findings on imaging**
Intravenous leiomyomatosis	Intraluminal growth of leiomyoma into vessels	Very rare. About 150 cases in literature	Female exclusively	Perimenopausal	Not known. History of leiomyoma	Hormonal therapy targeting oestrogen and progesterone receptors. Surgical resection.	Fairly good. Tumour recurrence noted in up to 30% of cases.	US – vascularised thrombi within vessels. CT - Filling defects within vessels. MRI – Low to intermediate T1W and low T2W signal. Usually homogenous enhancement.
Benign metastasizing leiomyomatosis	Believed to be haematogenous metastases	Very rare. About 150 cases in literature	Female exclusively	Perimenopausal	Not known. History of leiomyoma	Hormonal therapy targeting oestrogen and progesterone receptors. Surgical resection.	Fairly good	CT - Solitary or multiple solid well defined smoothly marginated pulmonary nodules. Calcifications and cavitation may be present, albeit rare.

DIFFERENTIAL TABLE

	Ultrasound	CT	MRI	Pattern of enhancement
Intravenous leiomyomatosis	Vascularised thrombi within vessels.	Filling defect within vessels	Filling defect within vessels. Low to intermediate T1W and low T2W signal. Usually no restricted diffusion.	Homogenous
Leiomyosarcoma	Difficult to distinguish from intravenous leiomyomatosis.	Heterogeneously enhancing filling defect	Iso- to hyperintense on T2W images, enhances on T1W postcontrast images. Restricted diffusion [6]	Variable
Bland thrombus	Non vascularised thrombi within vessels.	Filling defect within vessels	High T1W/T2W signal during acute phase. Low T2W signal in chronic phase. No restricted diffusion.	No enhancement

	X-Ray	CT	Pattern of enhancement
Benign metastasizing leiomyomatosis (Pulmonary)	Solitary or multiple pulmonary nodules.	Solitary or multiple solid pulmonary nodules with smooth margins. Calcifications and cavitation may be present, albeit rare.	Usually homogenous enhancement
Pulmonary metastases	Solitary or multiple pulmonary nodules.	Solitary or multiple pulmonary nodules. Cavitation may be observed. May have enlarged lymph nodes, bony lesions or other sites of disease. History of known primary malignancy in the lung or elsewhere.	Usually enhancing.
Pulmonary sarcoidosis	Reticulonodular opacities. Enlarged paratracheal stripe, mediastinal widening, and bilateral hilar enlargement	Pulmonary nodules which are perilymphatic in distribution. Nodular thickening of the interlobular septa. In chronic cases, there may be evidence of pulmonary fibrosis. Enlarged mediastinal nodes with or without calcifications.	Usually enhancing.

KEYWORDS

Intravenous leiomyomatosis; Benign metastasizing leiomyomatosis; Extrauterine leiomyomatosis

ABBREVIATIONS

BML=BENIGNMETASTASIZINGLEIOMYOMATOSIS
CT = COMPUTED TOMOGRAPHY
IVL = INTRAVENOUS LEIOMYOMATOSIS