

Intravenous Leiomyomatosis: Case Report and Review of the Literature

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Introduction

Intravenous leiomyomatosis (IVL) is a rare pathology, characterised by formation and growth of leiomyoma within vascular wall. Although histologically benign, IVL demonstrates some malignant behaviours, with spread and localised invasion of the vasculature typically through the ovarian veins to the inferior vena cava (IVC)¹.

Case report

We report a case of intravenous leiomyomatosis with inferior vena cava and intracardiac extension in a 56 year old post-menopausal woman. Primary presentation was with progressive shortness of breath on exertion and initial imaging demonstrated a large right aortic mass.

Subsequent findings include extension of a uterine fibroid into internal iliac and common iliac veins, extending into IVC, reaching the right atrium and pulmonary artery.

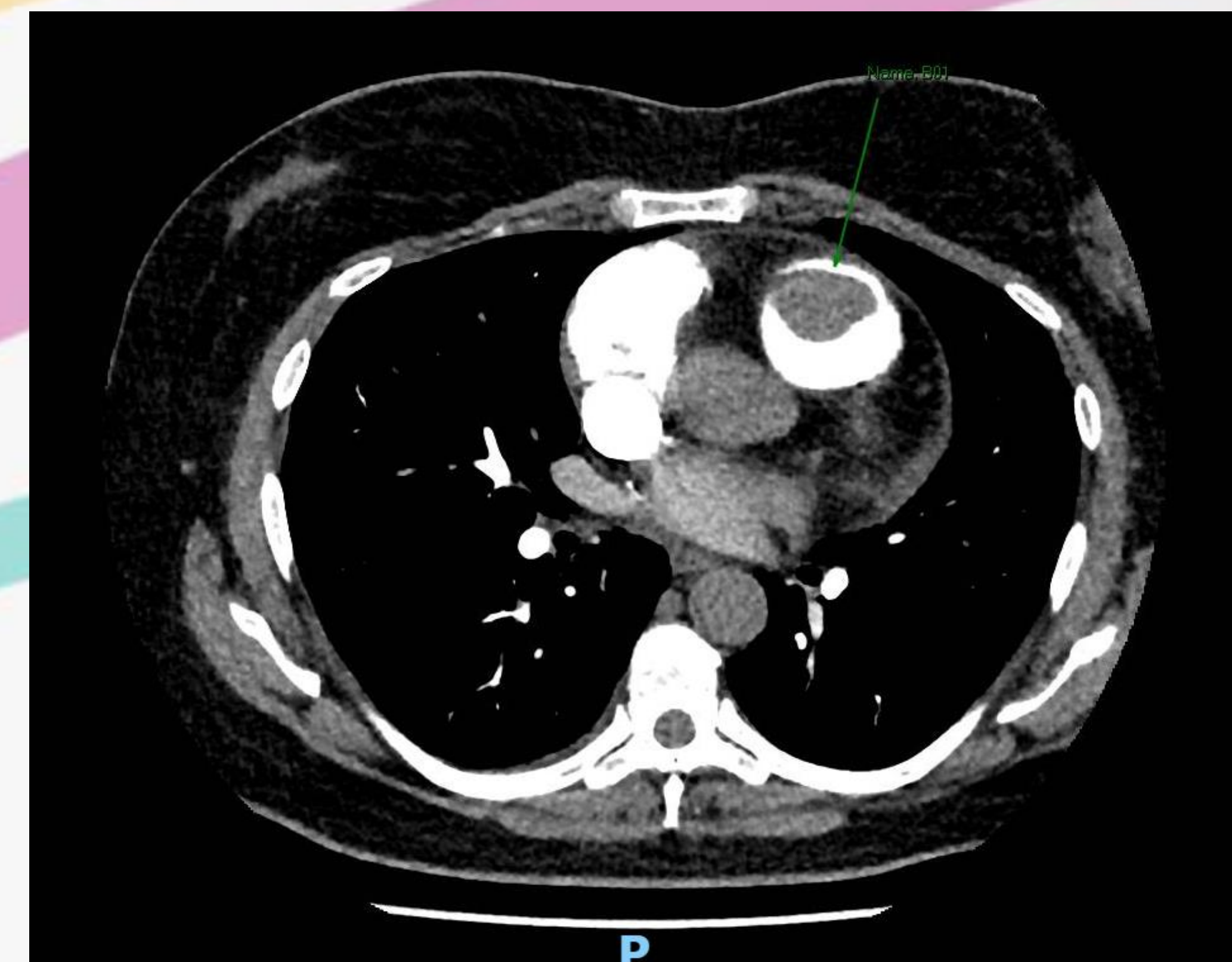
Thoracic debulking was performed and initial histology confirmed diagnosis of intravenous leiomyomatosis.

She has been commenced on Letrozole and second surgery is planned with gynae, vascular and GI surgical teams to strip the IVC and perform hysterectomy, BSO, adnexal clearance.

IVL Staging³

- Stage I: tumours invading the uterine vein and limited to the pelvis
- Stage II: tumours extending to the abdominal cavity but without involvement of the renal vein
- Stage III: tumours invading the renal vein and inferior vena cava and extending further up to the right atrium but without reaching the pulmonary arteries
- Stage IV: tumours reaching the pulmonary arteries or metastasizing to the lungs

Fig.1 (below): Stage 4 IVL, tumour reaching pulmonary artery
Fig.2 (right): ILV extending full length of the IVC



- IVL is a rare disease with benign histology but can behave aggressively and lead to death through cardiac obstruction¹.
- Current evidence supports mainstay treatment as a one or two stage complete surgical resection of the tumour. Treatment strategies should be individualised and consideration for hormone therapy may be supported when surgery is not possible.
- All cases of IVL should receive long term follow up

Literature Review

Fewer than 400 cases of IVL have been reported in the English literature¹. Mean age of onset has been estimated between 45 and 47 years and up to 65% have a history of uterine surgical procedures, such as caesarean section, myomectomy and hysterectomy².

The differential diagnosis considered in this case is of a uterine PEcoma. Histology has demonstrated widespread expression of HMB45 and cathepsin K. However, distribution of vascular plugging is more in keeping with IVL. Further histological samples will aid diagnosis.

Recurrences of intravenous leiomyomatosis may lead to suspicion of a missed diagnosis of a leiomyosarcoma.

Given the relative rarity of the condition, there is limited evidence to support a single best management approach.

Consensus from the literature supports one or two stage surgical procedure, with complete resection reducing risk of recurrence^{1,2}. Intravenous leiomyomatosis cells exhibit estrogen and progesterone receptor positivity. Use of gonadotropin releasing hormone agonist was previously recommended as a postoperative approach to reducing the risk of recurrence in cases with preservation of both ovaries or impossibility of complete resection, but recent studies have questioned its effectiveness^{3,5}. Use of tamoxifen and aromatase inhibitors has also been reported^{4,5}.

References

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