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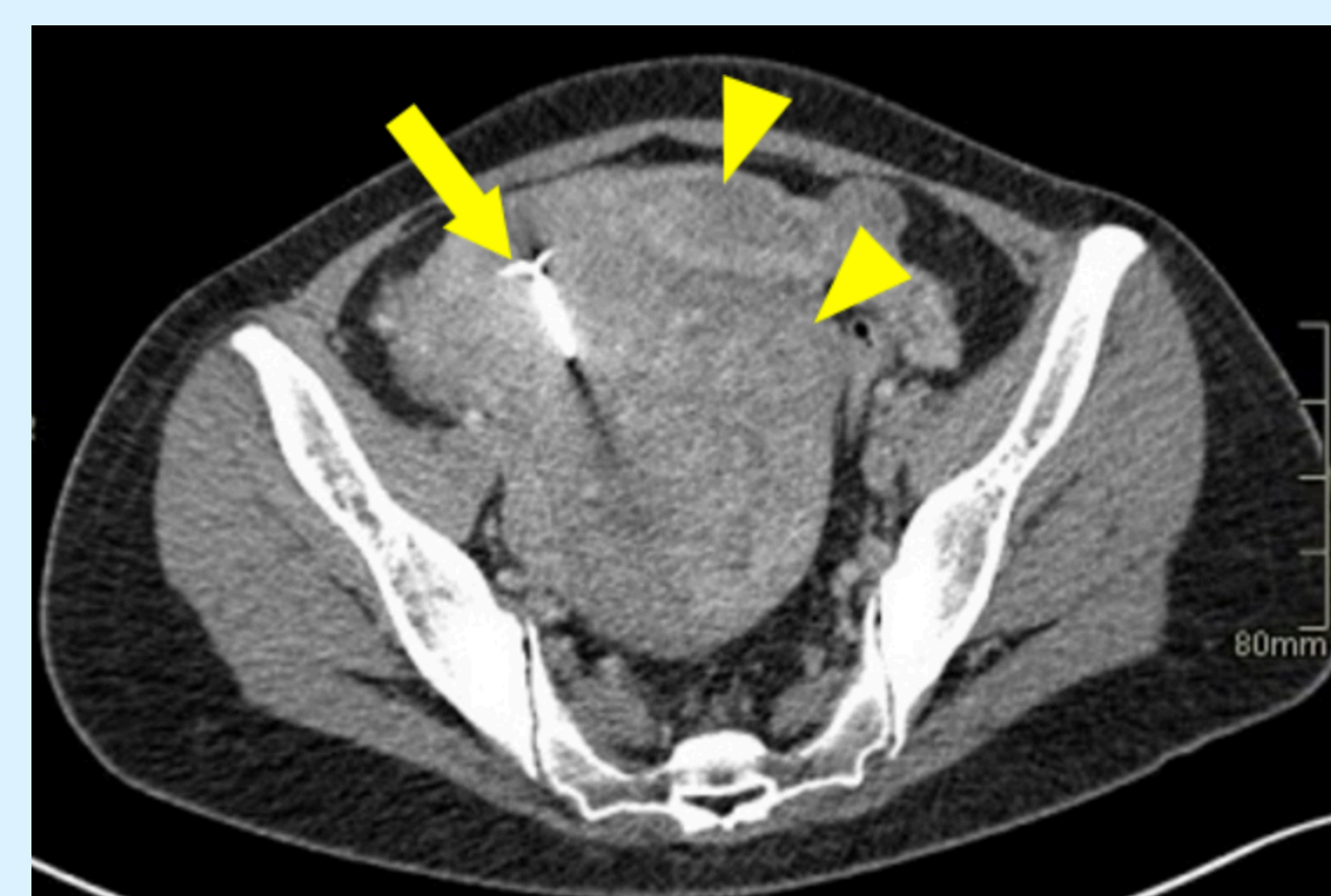
Introduction

Intravascular leiomyomatosis (IL) is an extremely rare, benign condition where smooth muscle cells arising from the uterus proliferate in the pelvic vasculature. It can, rarely, extend into the IVC, right heart and the pulmonary trunk. We report the first case of an asymptomatic patient with IL extending through the IVC, into the right ventricle and pulmonary trunk. The focus of this poster is on the diagnostic and management challenges presented by these cases.

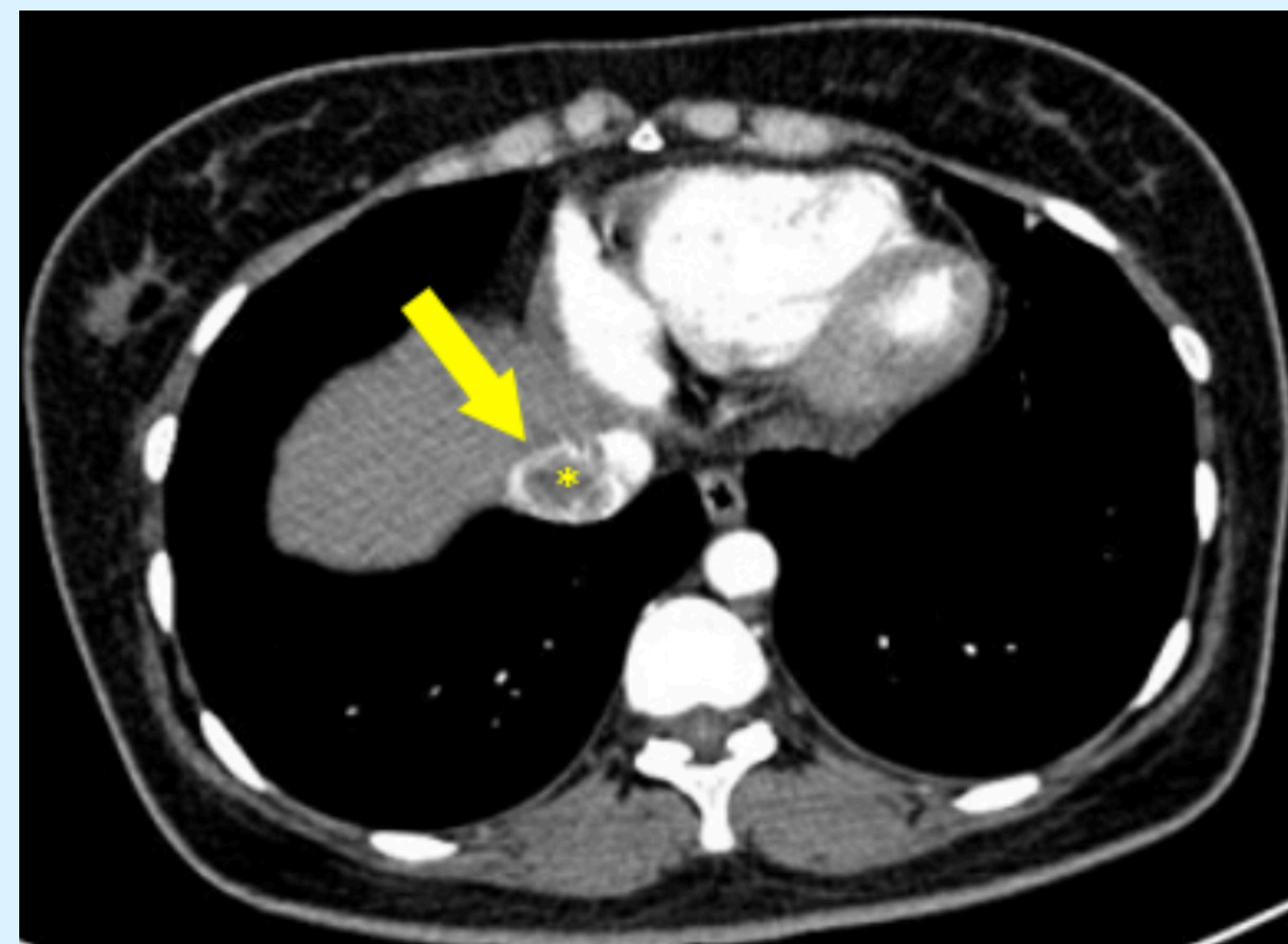
Case Report

A 39 year old patient attended the GP as she could not feel her IUCD threads. An ultrasound scan was performed which found a large pelvic mass. A CT scan demonstrated a 19cm mass involving the uterus and adnexae with suspected tumour thrombus extending from the iliac veins, into the IVC, right atrium and right ventricle. Image-guided biopsy was performed with the histology reported as a smooth muscle tumour, likely originating from the female genital tract with no evidence of malignant transformation. She was admitted for therapeutic anticoagulation and MDT review by gynae-oncology, urology, hepatobiliary and cardiothoracic surgery.

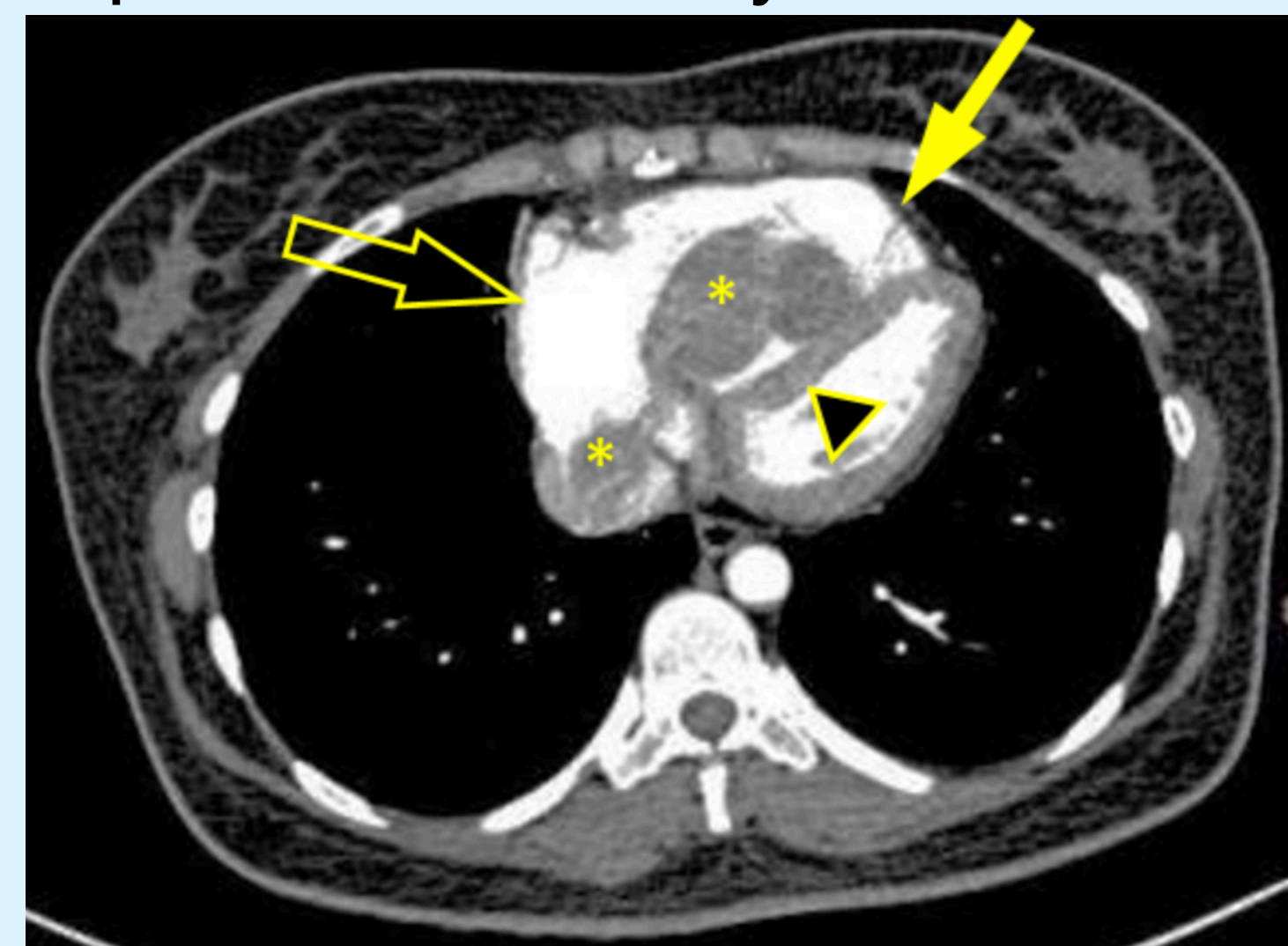
On pre-operative CT the lesion had progressed into the pulmonary trunk (see images below). The patient remained asymptomatic throughout. At operation cardiopulmonary bypass was commenced. The right atrium was opened and the IL identified. The tumour was fully excised from the chest and IVC. A total abdominal hysterectomy and bilateral salpingo-oophorectomy was undertaken to remove the pelvic disease. A small residual section densely adherent in the left common iliac vein was left due to clinical deterioration of the patient. Post-operative recovery was uneventful. At 6 months she was well with a tiny, stable nodule in the left pelvic side wall on imaging.



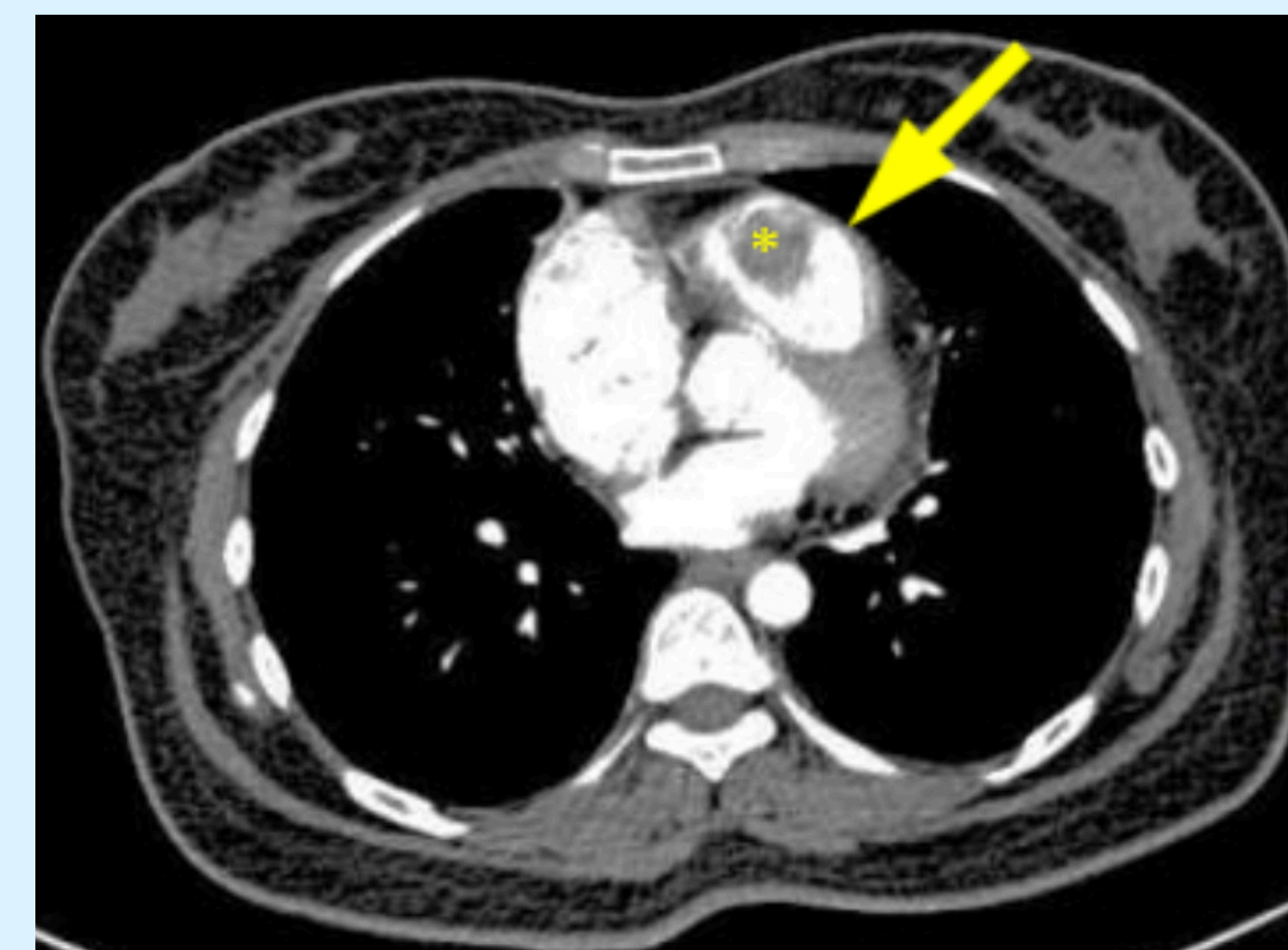
Fibroid uterus. IUD (solid arrow). Fibroids (arrow heads)



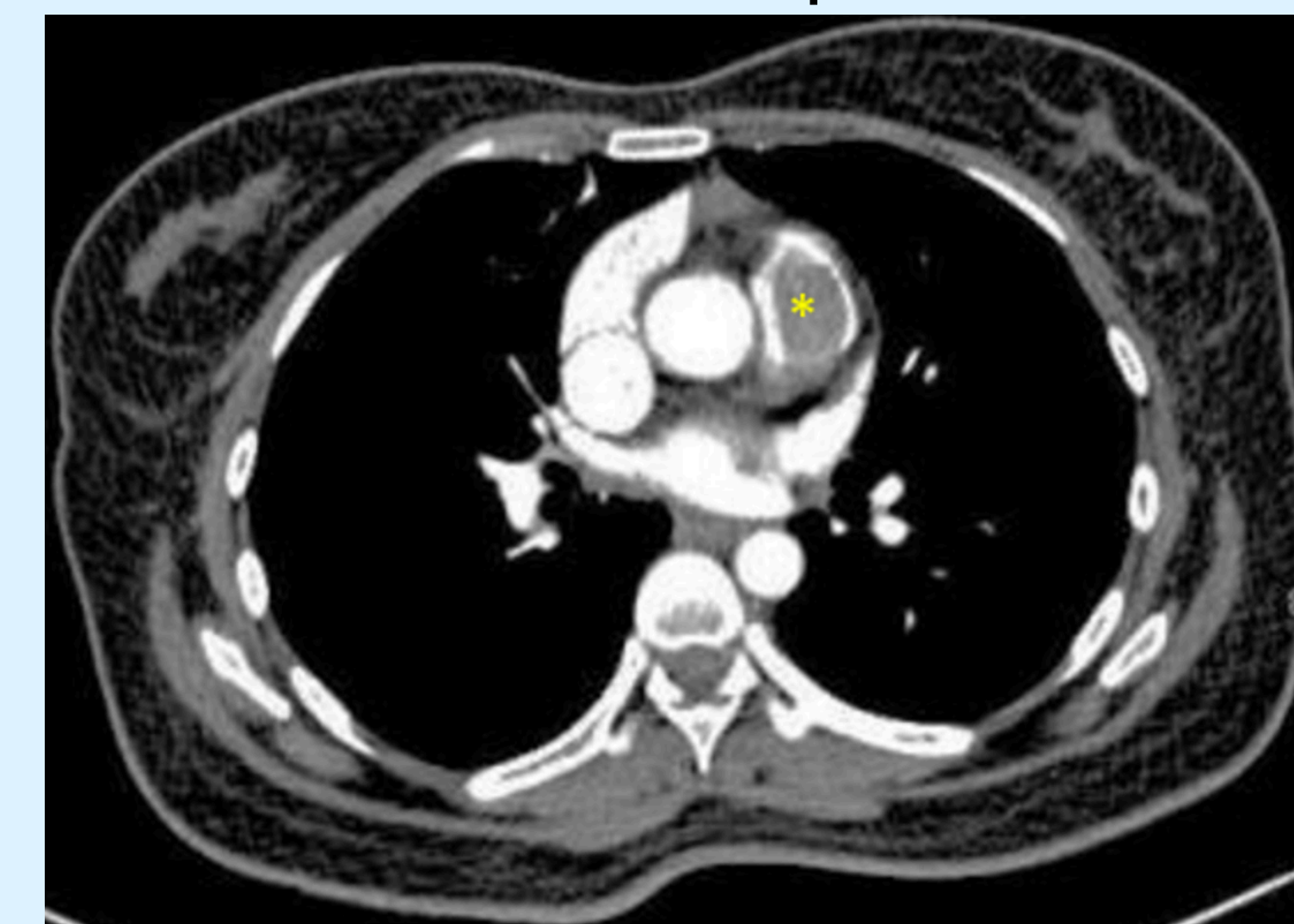
Tumour (*) within retrohepatic IVC (arrow)



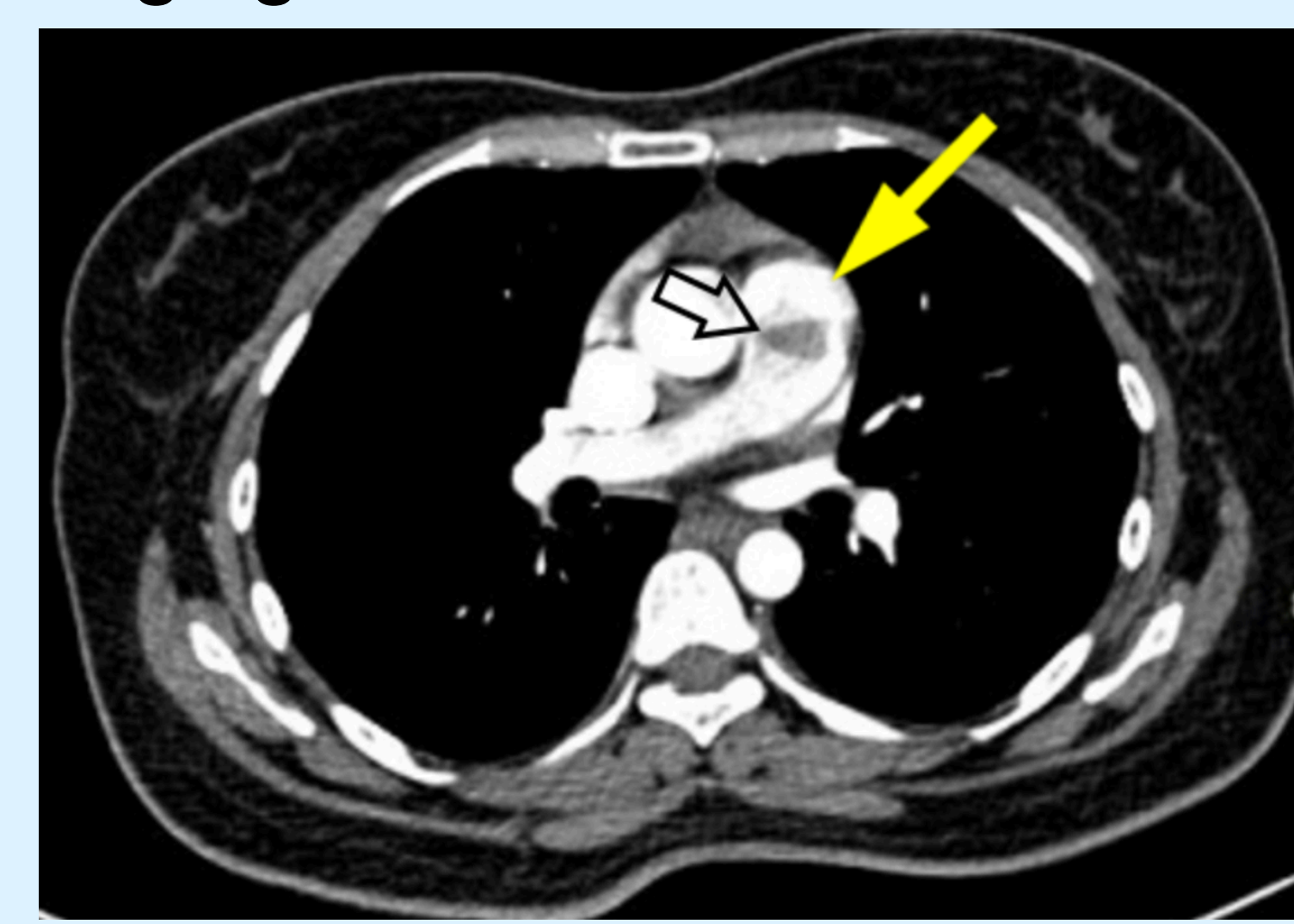
Tumour (*) is within the IVC, right atrium (hollow arrow) and right ventricle (solid arrow). Arrowhead - inter ventricular septum



Tumour (*) within the right ventricle (arrow)



Tumour (*) within infundibulum of pulmonary artery



Tumour (hollow arrow) within the main pulmonary trunk (solid arrow)

Discussion

While uterine leiomyomas are extremely common, development of IL is very rare with few reported cases. Clinicians rely on case reports to advance our knowledge of such rare conditions. We have identified only 4 reported cases of extension of IL beyond the heart[1,2,3]. To our knowledge this is only case in which a patient, with such extensive disease, was completely asymptomatic with no signs of cardiac or respiratory compromise. Prior to her presentation she worked up to 16 hours per day in a takeaway!

Surgical excision is the mainstay of treatment with oophorectomy preferred. Medical oestrogen suppression has been used to reduce risk of recurrence or in cases when the diagnosis is made pre-operative to reduce tumour bulk prior to surgery. Prognosis appears to be good if the tumour is fully resected and oestrogen suppressed. However recurrence has been reported in premenopausal women.[1]

Conclusion

Intravascular leiomyomatosis should be considered when extensive thrombosis is reported in patients with a history of leiomyoma, especially if there is progression of the vascular filling defect despite anticoagulation. In cases with involvement of extrapelvic disease, multidisciplinary surgical, anaesthetic and critical care working is essential to ensure a good outcome for the patient.

- 1) Virzi G, et al. Intravenous leiomyomatosis extending from the inferior naval vein to the pulmonary artery. The Journal Of Thoracic and Cardiovascular Surgery. 2007; 133(3) pp831-832
- 2) Su Q, et al Intravenous leiomyomatosis of the uterus: a retrospective singe-centre study in 14 cases. Biomed Res Int. 2020
- 3) Murphy A, et al. Intravenous leiomyomatosis manifesting as saddle embolism. BMJ Case Reports. 2019; 12(3)