

Rare case of high grade endometrial stromal sarcoma with extensive metastasis.

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Case Summary

- 46 years old lady presented with heavy menstrual periods and MRI suggestive of uterine fibroid with possible sarcomatous changes. Presentation was delayed due to pandemic.
- Primary surgery planned was total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH+BSO).
- Intra-operatively, tumour mass was felt invading the paravaginal and right gonadal veins and intra-operative ultrasound confirmed extension into the inferior vena cava(IVC) but could not define upper extent of the tumour thrombus.
- TAH+ BSO was done.Post operative contrast imaging confirmed tumor extension into IVC. Possibility of tumour thrombus in the pulmonary artery was raised but it could not be concluded if pulmonary emboli(PE) were secondary to tumour or blood clots. Trans thoracic echocardiogram showed tumor thrombus in right atrium.
- On balance, anti-coagulation(Apixiban) was started.
- Final histopathology confirmed the tumor as high grade endometrial stromal sarcoma(HG- ESS) with molecular analysis showing YWHAE/NUTM2A+B gene fusion.
- The case was discussed with the Sarcoma Multidisciplinary Team(MDT) and she was given letrozole and 3 cycles of doxorubicin.
- Post chemotherapy imaging showed reduction in PE, slight reduction in IVC, right gonadal vein and right internal iliac tumour thrombus.
- Second Surgery with vascular team 6 months after primary surgery: Excision of right gonadal vein with IVC intravenous thrombus and right iliac vein with branches.
- Post-operatively patient received 3 cycles of doxorubicin.
- Imaging 3 months after second surgery showed no evidence of recurrent or metastatic disease.
- She was then continued on hormonal treatment- letrozole 2.5mg OD.
- On repeat imaging 6 months after the second surgery, there is no evidence of recurrent or metastatic disease.

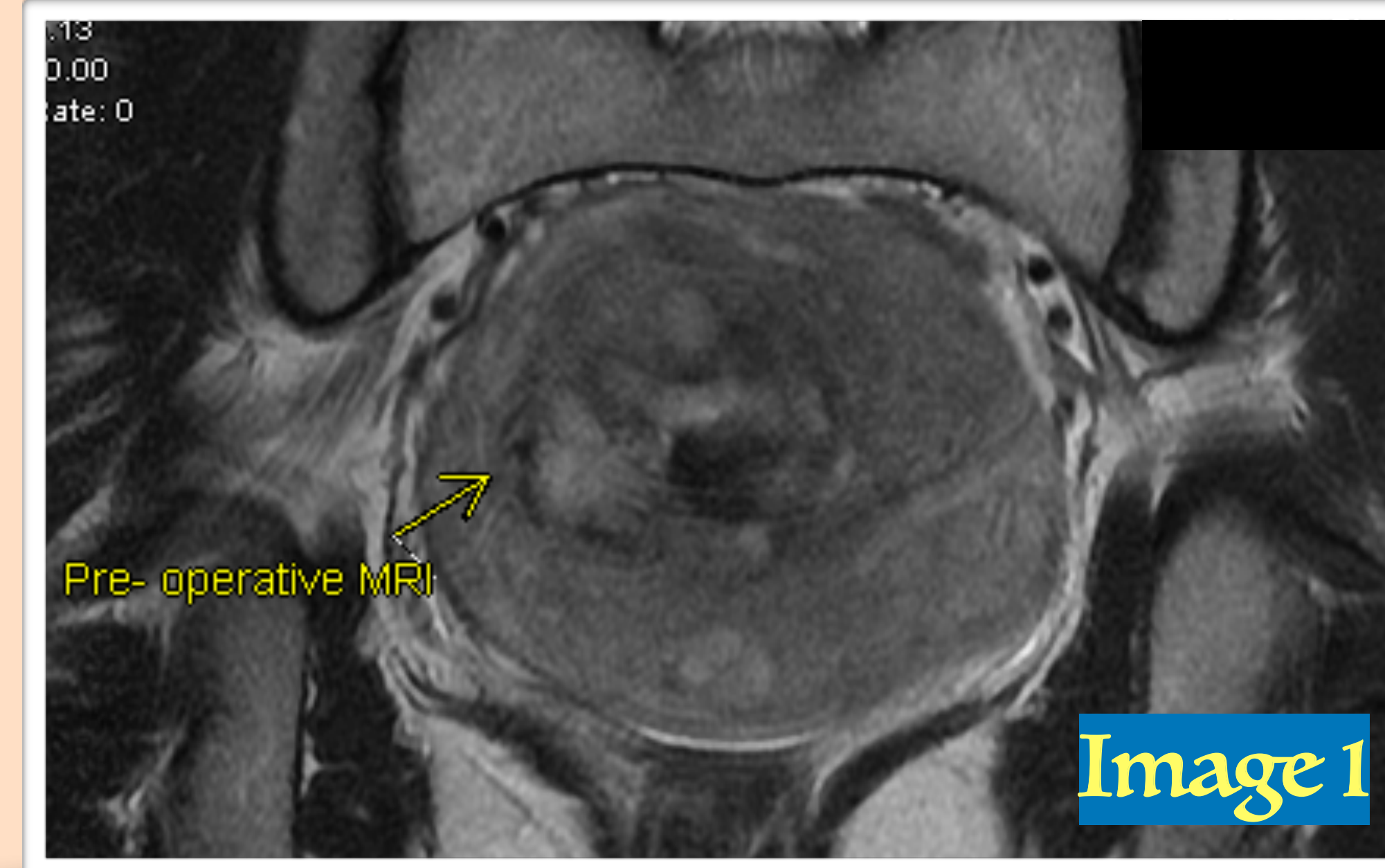
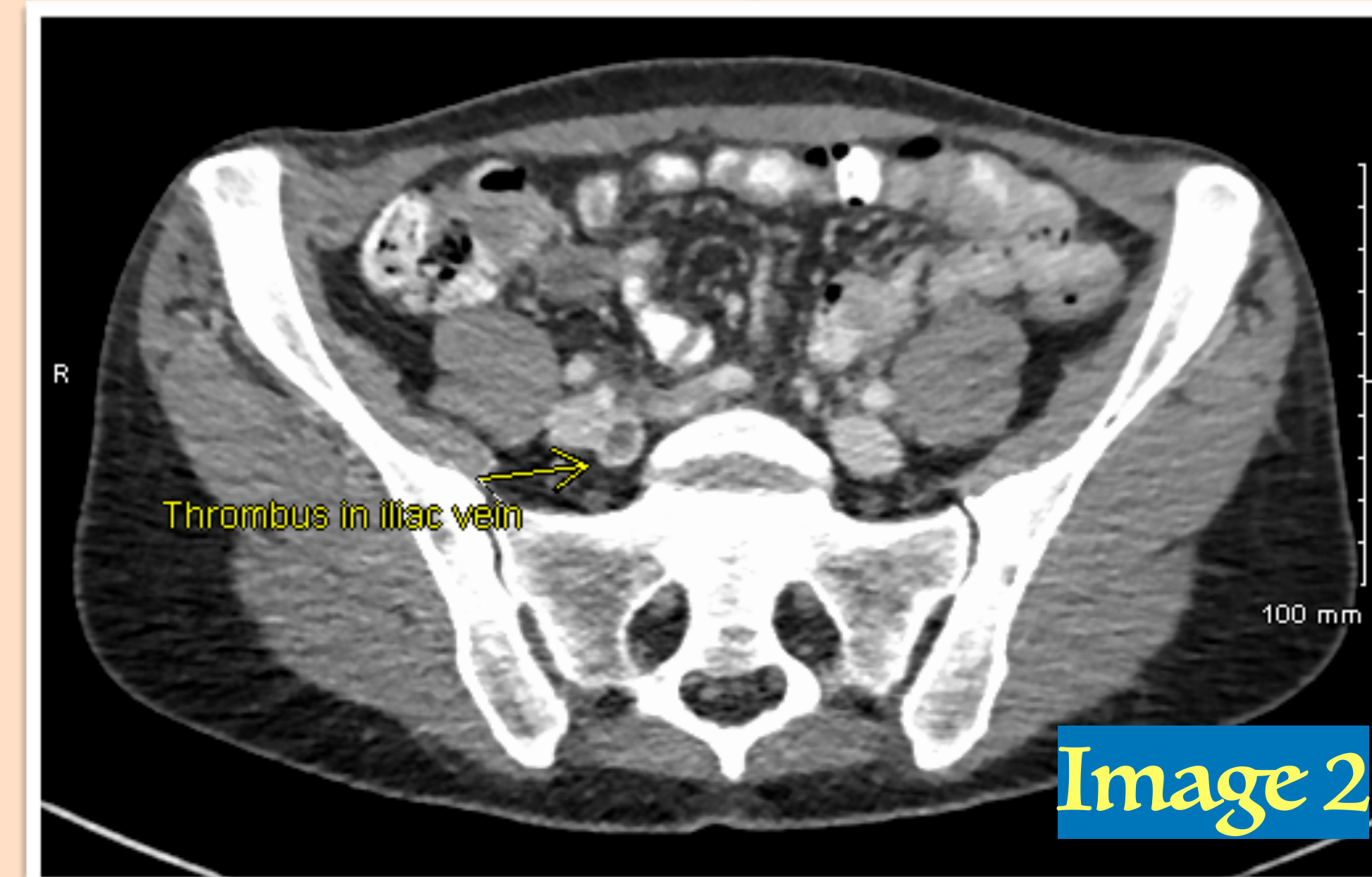
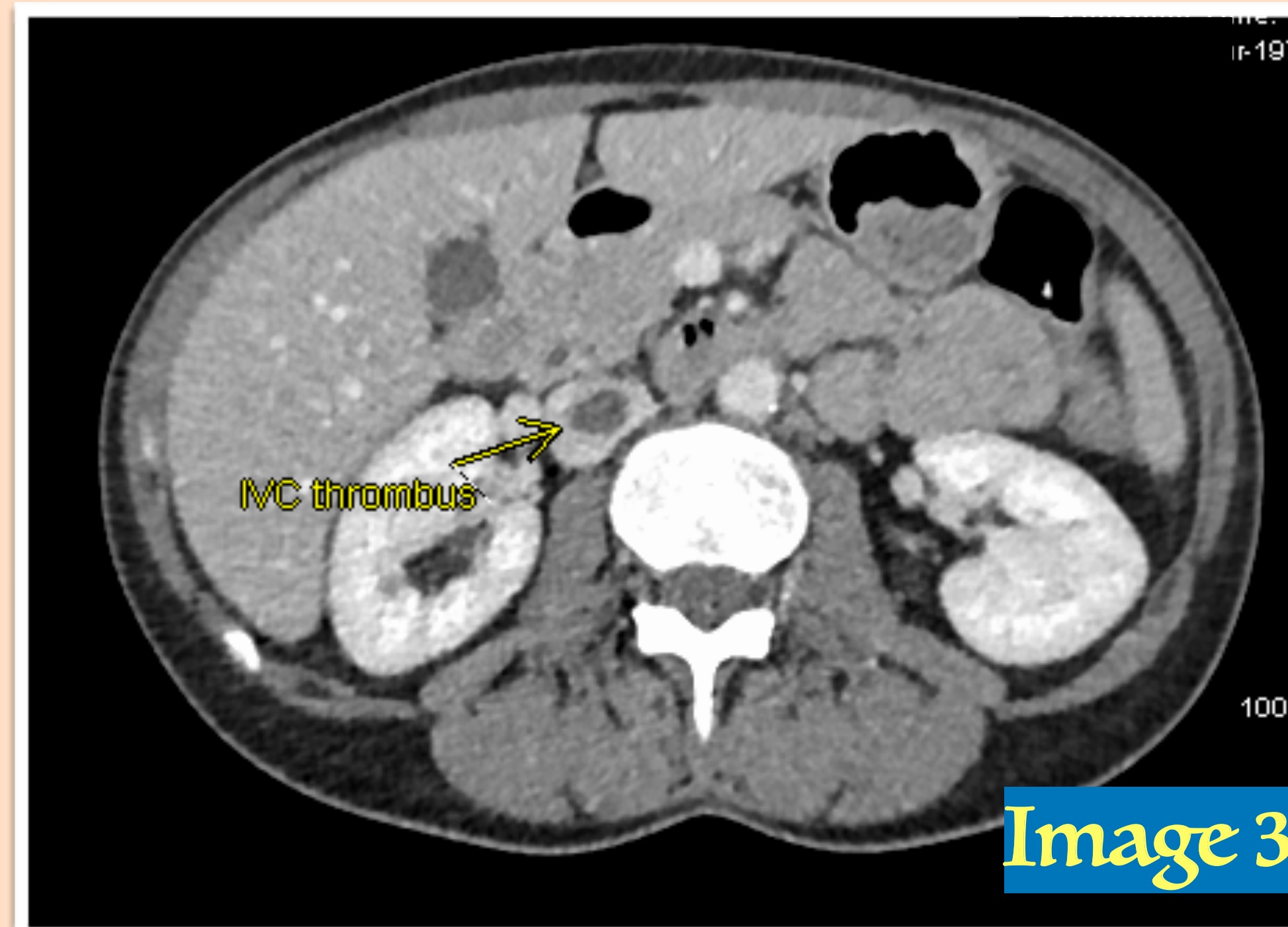
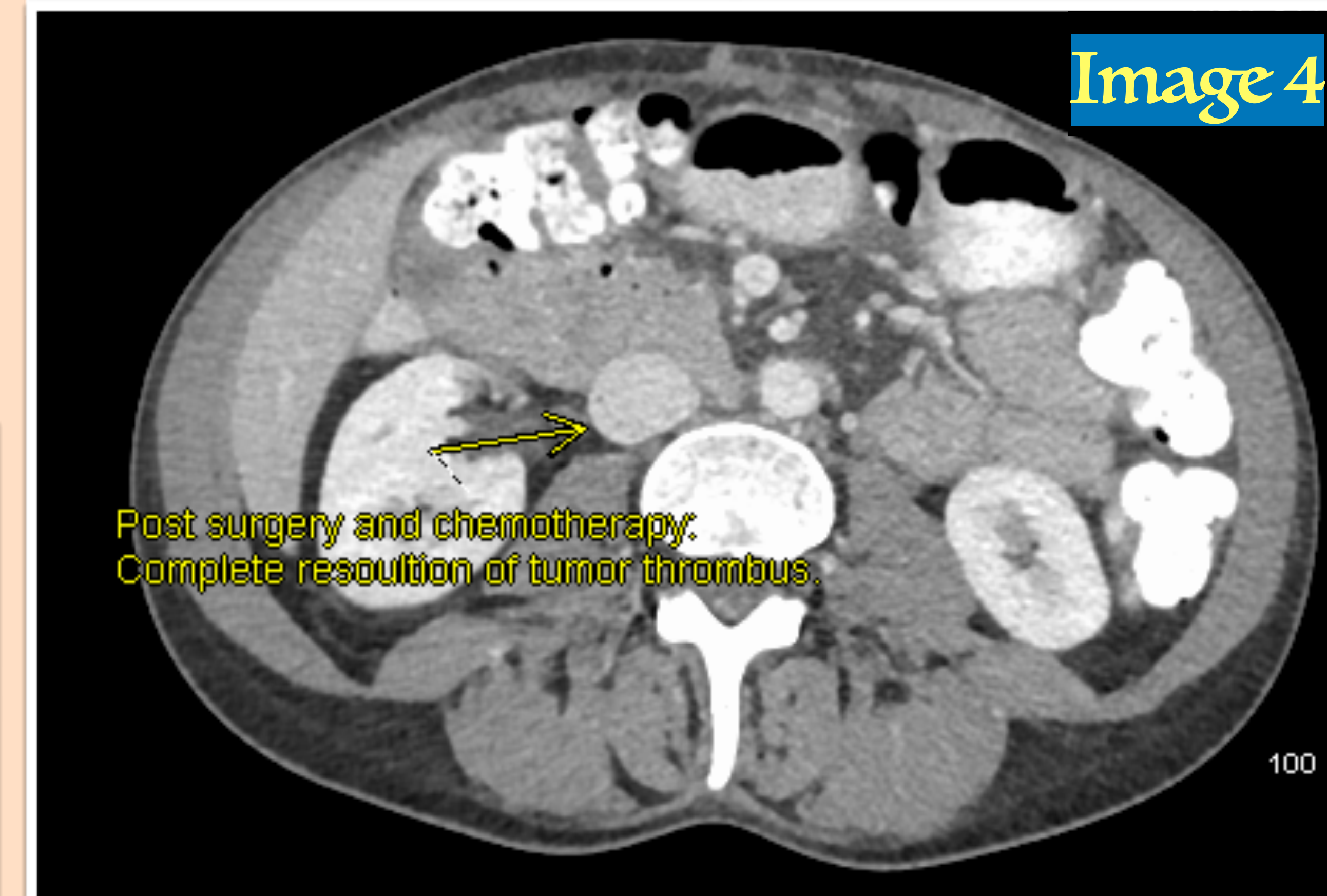


Image 1: Pre-operative MRI showing fibroids suspicious of sarcomatous changes.

Image 2: Tumor thrombus seen in Iliac Vein

Image 3: Tumor thrombus seen in inferior vena cava(IVC)

Image 4: Post surgery, chemotherapy and hormonal therapy **complete resolution of the tumor thrombus.**



Discussion

- Endometrial stromal sarcomas(ESS) are the second most common pure mesenchymal tumours of the uterus, and they account for 10% of all uterine sarcomas.
- There have been numerous case reports and case series about the extension of low grade ESS(LG-ESS) tumour thrombus into the IVC, extending to the heart and even to the pulmonary vasculature. However there are no cases reported of HG-ESS showing such a pattern of spread. This potentially makes this the first case of HG-ESS to be reported with such extensive pattern of spread.
- Diagnostic dilemmas with regards to immuno-histopathology are not uncommon while differentiating LG-ESS from HG- ESS as was in this case as well. Molecular studies thus becomes very important for correct diagnosis.
- This case reiterates the importance of thorough vascular imaging including cardiac imaging in pre-operative staging and planning.
- Multimodal approach with surgery, chemotherapy and hormonal therapy in ER/PR positive cases is often needed.
- An MDT approach is important in managing these cases especially to plan chemotherapy and surgery.
- Newer therapies like apatinib(antiangiogenic agents) and pazopanib in treating HG-ESS are under evaluation.

References

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