Incidental intravascular leiyomyomatosis: a case report and review of the literature.

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Intravascular leiyomyomatosis (IVL) is a rare smooth muscle cell tumour that is histologically benign with metastatic behaviour. The tumour arises from the uterus and grows within the venous system, extending to the inferior vena cava, right-sided cardiac chambers and pulmonary vessels. IVL can be fatal, resulting in thromboembolic events, congestive heart failure and intra-pulmonary leiyomyomatosis. There is also a risk of recurrence if the tumour is not completely resected.[1]

Presentation usually occurs after the disease has advanced with symptoms of haemodynamic instability, dyspnoea, palpitations, chest or abdominal pain.[2] Typically, diagnosis is based on macroscopic description of worm-like tumour projections in the veins and/or microscopic evidence of intraluminal leiyomyomas.[3]

We report a case of a 44-year-old woman who underwent a total abdominal hysterectomy and bilateral salpingectomy for simple hyperplasia, diagnosed from uterine curettage performed for menorrhagia. Intraoperatively, increased vascularity and aberrant vessels were noted on the serosal surface. The macroscopic pathology examination was normal, however, the histopathology demonstrated smooth muscle tumours in 2 vessels, consistent with IVL. The patient underwent computed tomography imaging of the chest and abdomen to rule out metastases and is scheduled for yearly follow-up due to the risk of recurrence.

This case highlights a rare but important diagnosis that gynecologists should be suspicious of when abnormal vascularity is seen on the uterus, especially due to the high risk of morbidity and recurrence with IVL. We review the literature and discuss management options for optimal outcomes of this disease.

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References:

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