A case series of Intravenous leiomyomatosis and literature review

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IVL is a histologically benign smooth muscle tumor which intravascularly extends into uterine and systemic veins, in the absence of, or beyond the confines of a leiomyoma (1). Approximately 80% of the tumor growth into the venous system of the myometrium and parametrium, cardiac involvement is observed in 10% to 40% of cases and haematogenous spread to the lung has been described (2). IVL is rare and there are fewer than 150 cases described in the literature (2).

Patients are commonly of reproductive age and rarely post-menopausal (1, 3). Clinical symptoms varies; patients with pelvic mass have symptoms included abnormal uterine bleeding, pelvic pain and abdominal distention (3), whereas patient with intracardiac involvement can present with symptoms of right heart failure (1,3).

The rarity of IVL poses a diagnostic challenge pre-operatively. CT scan and MRI are valuable but are not routine imaging modalities. IVL resembles uterine fibroids macroscopically and microscopically. The diagnosis is commonly made on histopathological examination (1).

Hysterectomy is the treatment of choice with the resection of extrauterine tumor when technically feasible (3).

We will present a case series of eight patients diagnosed with IVL on histopathology in a seven month period at The Royal Women's Hospital and a literature review of the current management of this rare disease.

All patients presented with menorrhagia. Mean age at presentation was 43.5 yo (33- 50 yo). One patient had ultrasound and MRI features suggestive of atypical cellular fibroid. Six patients underwent hysterectomy and two patients had laparoscopic myomectomy. There was no significant intra-operative findings although suspicion of IVL was raised in one of the cases. Histopathology revealed venous invasion and extension beyond the tumor mass from which IVL originates.

No patient had extra uterine spread.

Patients were educated about signs and symptoms of spread of IVL. CT scan of chest, abdomen and pelvis will be performed in 2 and 5 years to detect intravascular recurrence in the future.

Conclusion
IVL is rare and can be a diagnostic challenge. There should be a high clinical suspicion for recurrence and patients should have long term follow up.

PROJECTS

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