

Intravenous leiomyomatosis of the uterus: link with new fertilisation methods?

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A 41-year-old woman with a medical history of hormonal treatment for primary infertility eight years earlier and three subsequent operations for “recurrent uterine leiomyomas”, was admitted to our hospital with suspected iliac vein thrombosis extending to the inferior vena cava in CT. Hysterectomy had been performed two weeks earlier for an enlarged heterogeneous uterus on MRI that raised the possibility of leiomyosarcoma. Indeed, although some MRI features can suggest malignancy, there are no specific imaging criteria differentiating leiomyosarcoma from other uterine tumours.

The histopathological result of the hysterectomy specimen revealed uterine intravenous leiomyomatosis (IVL). The clinical picture pointed to the diagnosis of intravascular extension of the uterine IVL into the iliac vein and inferior vena cava (IVC). Excision of the intravascular mass was performed. The surgical specimen exactly fitted with the CT images (fig. 1a–b). There was no thrombotic component to the mass. Differential diagnosis of this IVC mass includes primary leiomyosarcoma or extension of adjacent tumours such as adrenocortical tumours, renal cell or hepatocellular carcinoma. In this case, the diagnosis of IVL was confirmed histopathologically, with characteristic smooth muscle spindle-shaped cells within a heterogeneous hyaline structure invading the vascular lumen without any atypical cellular signs. Immunohistochemical analysis was positive for smooth muscle actin. Interestingly, 80% of the cell nuclei were positive for oestrogen and progesterone receptors.

IVL is a rare, benign tumour of the uterus first described in 1896 by Birch-Hirschfeld. It typically extends into the vascular lumen with various extension patterns. [1] Tumour progression can involve the iliac veins, the inferior vena cava and even the



Figure 1a

Figure 1

Contrast-enhanced CT of the abdomen with curved coronal reconstruction (fig. 1a) and surgical specimen (fig. 1b) showing a complex mass in the lumen of the left internal iliac vein (small arrows) extending to the inferior vena cava (arrowheads).

right atrium. [2] Unusual extension patterns of uterine leiomyomatosis include peritoneal masses, sometimes mimicking carcinomatosis, or “metastasising” leiomyoma with benign pulmonary nodules. [3–5] Symptoms and signs depend on the sites of tumour extension. Definite diagnosis is confirmed by histopathology. [6] Treatment consists of complete resection of the mass to prevent recurrence.

There seems to be an increased number of IVL cases reported over the last few years, which could possibly be due to an increased incidence of this rare tumour. One hypothesis could be that the now widespread use of fertilisation methods may trigger tumour growth in hormone-sensitive tumours in which nuclear oestrogen and progesterone receptors can be demonstrated. The role of reproductive techniques as a risk factor has already been suggested in cases of leiomyomatosis peritonealis disseminata. [7] Interestingly, antioestrogens such as tamoxifen or raloxifene [8] or GnRH agonists [9, 10] have been used for treatment of IVL. The aromatase inhibitor letrozole has also been used in one patient. [11] However, these therapeutic non surgical options must still be considered hypothetical.

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Figure 1b

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