Case 16256

Intravenous Leiomyomatosis
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Section: Genital (female) imaging
Area of Interest: Genital / Reproductive system female
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: MR
Imaging Technique: PACS
Special Focus: Pathology Case Type: Clinical Cases
Authors: Joao Cunha Salvador / Joao Pedro Caldeira
Patient: 61 years, female

Clinical History:
A 61 years-old woman complained of progressive pelvic pain for 4-months, with no abnormal vaginal bleeding. Physical examination only revealed a distended abdomen, with no pain on compression. On the gynecological observation the vagina and cervix were unremarkable.

Imaging Findings:
Pelvic post-contrast computed tomography (CT) revealed a large, well defined but markedly heterogeneous tumor in the uterine fundus, partially exophytic, with small coarse and peripheric calcifications, which was suggestive of subserosal leiomyoma. However, there was a tubular lesion with the same characteristics in the right parametrium, extending superiorly into the retro-peritoneal iliac vessels.

On magnetic resonance (MR) imaging both tumors were iso-intense to muscle on T1-weighted imaging (T1W) and heterogeneously hyper-intense on T2-weighted imaging (T2W). The fundus lesion was irregularly delineated with a hypo-intense layer of myometrium, and the tubular right parametrial lesion showed well-defined borders with lobulated contours. This lesion extended from the para-cervical region into the primitive iliac vessel's region, inducing pelvic venous congestion with some peri-uterine transudation. Both lesions demonstrated restricted diffusion, and almost homogeneous intense enhancement after gadolinium administration. Neither pelvic adenopathies nor distant lesions were documented.

Discussion:
Intravenous leiomyomatosis (IVL) is a rare manifestation of a benign uterine leiomyoma [1]. IVL is an aggressive form of this benign condition, representing growth of smooth muscle within the lumen of vessels, outside the limits of a uterine leiomyoma [2]. It can grow from the walls of the vein itself, but most commonly arises from an uterine leiomyoma, growing into the uterine vessels [3]. From here, tumor can extend upwards into the right atrial cavity [3], giving different clinical symptoms depending on the level of the tumor, ranging from asymptomatic, to pelvic pain and vaginal bleeding (typical leiomyoma symptoms), to leg edema and swelling from iliac vessels/inferior vena cava occlusion, and to cardiac failure, dyspnea and even syncope or sudden death from cardiac extension [2-4].

The radiologic diagnosis is suggested when a parametrial lesion with growth along pelvic and retroperitoneal vessels is present, with similar characteristics on MR and CT of a uterine leiomyoma, being confirmed when a connection between the two is seen [3-4]. The presence of coarse calcifications may help to establish the diagnosis. However, these may be mistaken with phleboliths, typically located in the pelvis. Many cases are found after
previous hysterectomy for leiomyomas, which may be a clue for this entity, but also hinder the radiological connection sign [3]. MR with gadolinium administration is of utmost importance, mainly to differentiate IVL from a bland venous thrombus, but also because IVL is usually very vascular [2]. The typical MR signal characteristics are iso-intensity to muscle on T1W, and iso- to hypo-intensity on T2W [1, 3, 4]. Hydropic degeneration is the most common degeneration of IVL [2], which translates into hyper-intensity on T2W.

If IVL reaches the inferior vena cava, other tumour thrombus type of lesions must be considered: Wilms tumour, hepatocellular carcinoma and adrenocortical carcinoma [3, 4]. Treatment of choice is complete surgical excision, which includes total hysterectomy, bilateral oophorectomy (hormonal dependent tumor) and excision of the vessels involved, sometimes even with cardiotomy [1, 3, 4]. Considering 30% of recurrence, long-term imaging follow-up at 3-6 months interval is recommended [4].

IVL is a rare form of a leiomyoma, that can manifest clinically from asymptomatic to sudden death, with similar imaging characteristics. Surgical excision is the norm, and follow-up is advocated attending to its high recurrence rate.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Intravenous Leiomyomatosis with Hyaline Degeneration., Uterine Leiomyosarcoma with parametrial invasion/venous tumour thrombus, Primary Venous Leiomyosarcoma

**Final Diagnosis:** Intravenous Leiomyomatosis with Hyaline Degeneration.

**References:**

**Figure 1**

*Description:* Axial post-contrast CT images revealing a large heterogenously enhancing lesion in the uterine fundus (*), partially covered by myometrium (white arrow) *Origin:* Department of Radiology, IPOLFG, Lisboa, Portugal
Description: And also a tubular lesion in the right parametrium (black arrows) extending from the para-cervical space (cervix - C) into the iliac vessels region (iliac vessels - IV). Origin: Department of Radiology, IPOLFG, Lisboa, Portugal
Description: Peripheral coarse calcifications are present on both lesions, as well as the same heterogenous density. Origin: Department of Radiology, IPOLFG, Lisboa, Portugal
Description: Peripheral coarse calcifications are present on both lesions, as well as the same heterogenous density. Origin: Department of Radiology, IPOLFG, Lisboa, Portugal
Description: Axial T2-TSE images (a, b, c, d) revealing both lesions as heterogeneously hyper-intense.

Origin: Department of Radiology, IPOLFG, Lisboa, Portugal

Description: This signal characteristic, along with the parametrial extension, were the findings that could support an uterine leiomyosarcoma into the differential diagnosis. Origin: Department of Radiology, IPOLFG, Lisboa, Portugal
**Description:** The lobulated contours of the parametrial lesion are better delineated in this image.

**Origin:** Department of Radiology, IPOLFG, Lisboa, Portugal

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**Description:** Note the right pelvic transudation from venous obstruction (arrow).

**Origin:** Department of Radiology, IPOLFG, Lisboa, Portugal
Figure 3

**Description:** Coronal and sagittal T2-TSE images showing the track of the right parametrial lesion. Note that it grows superiorly along the uterine veins into the internal iliac venous system. **Origin:** Department of Radiology, IPOLFG, Lisboa, Portugal.
**Description:** There is no invasion of the adjacent structures. Note the well-defined lobulated contours (white arrows). **Origin:** Department of Radiology, IPOLFG, Lisboa, Portugal
Description: The fundic lesion is circumscribed by a thin layer of myometrium, thus not being a subserosal lesion as CT images proposed (black arrow). Origin: Department of Radiology, IPOLFG, Lisboa, Portugal
Figure 4

a

Description: Axial T1-TSE image revealing the iso-intensity with muscle signal of both lesions (white arrows). Origin: Department of Radiology, IPOLFG, Lisboa, Portugal

b

Description: T1-fat suppression contrast enhanced image showing intense, mostly uniform, contrast enhancement after gadolinium administration (*). Origin: Department of Radiology, IPOLFG, Lisboa, Portugal
**Figure 5 a**

**Description:** Axial DWI images at b800 and ADC-map showing restricted diffusion on both lesions: hyper-intensity on b800 and respective hypo-intensity on ADC-map. **Origin:** Department of Radiology, IPOLFG, Lisboa, Portugal
**Description:** Axial DWI images at b800 and ADC-map showing restricted diffusion on both lesions: hyper-intensity on b800 and respective hypo-intensity on ADC-map. **Origin:** Department of Radiology, IPOLFG, Lisboa, Portugal
Figure 6

Description: Spindle cells arranged in fascicles, with areas of collagen deposition (*) features typical for hyaline degeneration of this intravenous leiomyomatosis. **Origin:** Department of Radiology, IPOLFG, Lisboa, Portugal.
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