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CASE REPORT

A case of intravascular leiomyomatosis, a pathology of infrequent diagnosis

Un caso de leiomiomatosis intravascular, una patología de diagnóstico infrecuente

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Abstract

A case of intravascular leiomyomatosis is presented in a 45-year-old woman, a benign pathology of very infrequent diagnosis. The clinical circumstance of its diagnosis and its histological characteristics are described.

Key words: Intravascular, Leiomyomatosis.

Resumen

Se presenta un caso de leiomiomatosis intravascular en una mujer de 45 años, una patología benigna de diagnóstico muy infrecuente. Se describe la circunstancia clínica de su diagnóstico y sus características histológicas.

Palabras clave: Leiomiomatosis intravascular.

Intravascular leiomyomatosis is a benign alteration caused by the proliferation of smooth muscle cells of the myometrial vessels or uterine fibroids. Its diagnosis should be considered when faced with uterus-dependent pelvic masses, especially if it occurs in women with previous or current uterine fibroids. It's important to keep in mind that it can spread to the cava vein and also to the right heart chambers.

A 45-year-old woman (LGB), with menarche at 12 years of age, menstrual type 4/28, with no history of interest, who consulted for the first time on 12/21, providing a a magnetic resonance imaging (**Figures 1, 2**). that depicted: "Increased uterus with leiomyomas and occupation of most of the pelvis by a large solid-cystic or solid left pelvic mass with necrosis and to a lesser extent a predominantly cystic or necrotic right para-uterine mass, which although they raise doubts due to their size and heterogeneity, they are suggestive of large pedunculated leiomyomas with degeneration or necrosis. Both ovaries normal".

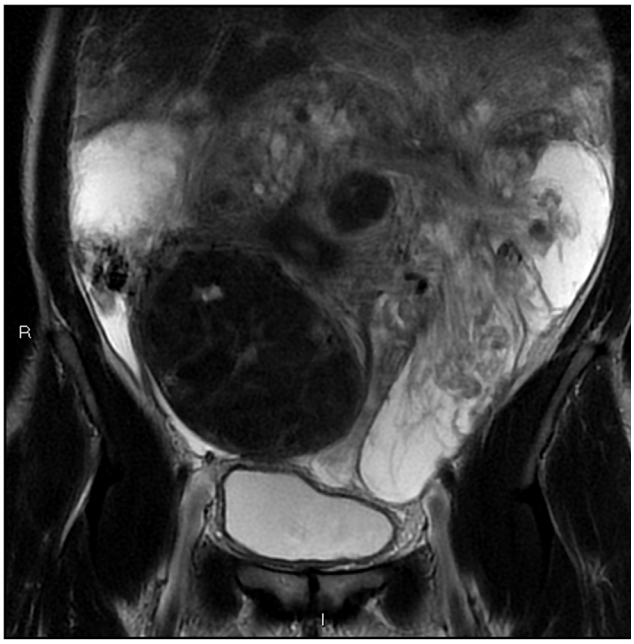
Surgical intervention was scheduled, which was performed on 01/22. It was found a large uterus with a polimyomatous appearance occupying the entire pelvis and a tumour of about 6cm with a cystic appearance. Also, in the left para-adnexal area and retroperitoneally a 40 cm tumour with a predominantly cystic appearance and a highly vascularized cystic not adherent to deep layers or intestine occupying the bottom of the pouch of Douglas. It was decided to perform a total hysterectomy with double adnexectomy and retroperitoneal lumpectomy.

Histological report

Total hysterectomy piece with right adnexectomy and left salpingectomy. It weighs 1002 g as a whole and has a polylobulated appearance with multiple myomatous lesions, the largest measuring 8 cm. At the level of the left horn, a poorly defined 6 x 5 cm area with an edematous and vesicular appearance was identified in relation to several small myomatous lesions. Collapsed endometrial cavity with smooth-surfaced mucosa 0.1 cm thick. Right ovary and tube without macroscopic alterations.

Resection piece of 20 x 20 x 7 cm multinodular with a vesicular appearance that presents an ovary adhered to one of its faces of 5 x 2 cm without alterations. The rest is made up of edematous material with small cystic areas and some hemorrhagic areas. Scattered myomatous-looking nodules are seen.

Figure 1, 2: Magnetic resonance imaging.



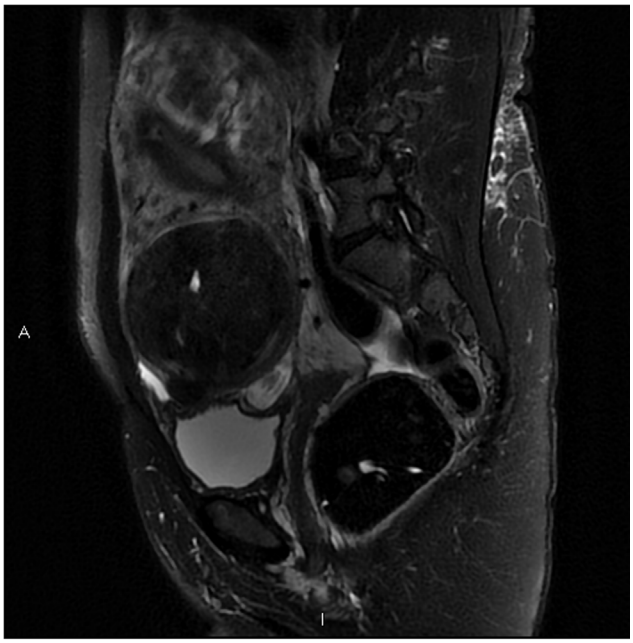


Figure 3: Hematoxylin-eosin. x16.

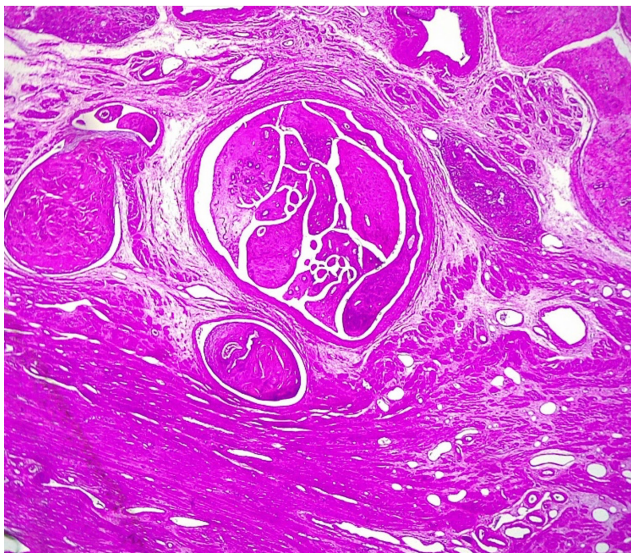
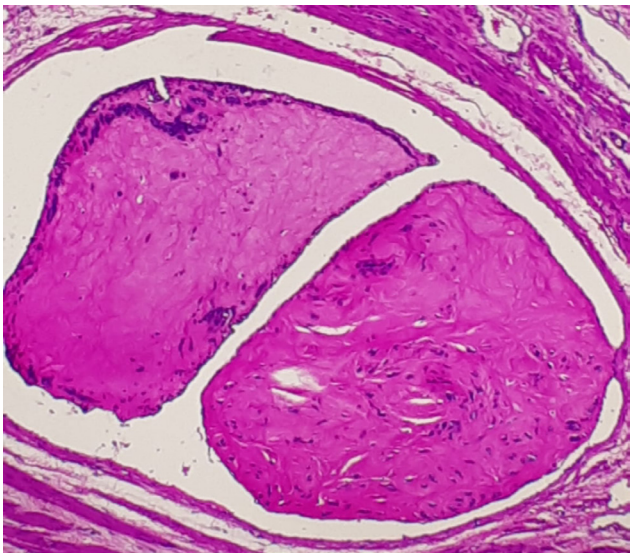


Figure 4: Hematoxylin-eosin. x200.



Both at the uterine and extra-uterine levels, a myomatous proliferation associated with ectasia of the vascular walls, with intraluminal growth, is observed (**Figures 3,4**). Extensive areas with marked edema. The nuclei are oval and norm-chromatic, with no tumor necrosis, atypical mitoses, or signs of aggressiveness proliferative endometrium. Tubes and ovaries without alterations.

Based on these observations, INTRAVENOUS LEIOMYOMATOSIS is diagnosed.

In March 2022 (03/14/2022) the Aortic CT Angiography revealed venous occupation by a single mass that extended from the right gonadal vein to the artery of the lower lobe of the right lung, consistent with his pathology.

Cardiac surgery was performed on 03/14/2022, observing normal-appearing lungs and heart. A white mass is observed inside the inferior vena cava. A hard, lobulated, pearly white tumor was completely removed (**Figure 5**). With extension of the superior and inferior lobar branches of the right pulmonary artery, a branch of the Left Pulmonary Artery crosses the right cavities outflow tract RV, RA and inferior vena cava (about 20 cm in IVC) with rest of old clots probably related to previous surgery.

The macroscopic description of pathological anatomy describes an elongated fragment of 40cm in length, beaded and made up of whitish tissue of a firm and well-defined consistency. The microscopic description defines fragments corresponding to a well-differentiated

Figure 5: lobulated, pearly white tumor.

tumor made up of bundles of spindle cells of smooth muscle lineage. Presence of deposits of abundant hyaline substance in the stroma. No necrosis or hemorrhage is evident. Absence of signs of malignancy. Diagnosis: intravenous leiomyomatosis.

In May 2022, a whole body CT angiography was performed to rule out endovascular satellite fibroids, with a negative result.

Comment

If "endovascular leiomyomatosis" and 2022 as the search period are entered in PubMed as the keywords, three publications are displayed[1], 163 if the word is "leiomyomatosis"[2]. This great difference reflects the diagnostic rarity of the case discussed here. This fact is clearly expressed in the publication that the Gynecology group of the MD Anderson Hospital in Madrid published in 2016[3] describing it as an "exceptional entity" and pointing out the difficulty of diagnosis and the need for its diagnosis to be suspected when, as in the case that we present here, the evaluation of uterine pathology is addressed in women with pelvic masses of polimyomatous appearance and especially if it is also suspected that there may be associated abdominal or thoracic vascular pathology.

It is a benign pathology but it needs to be addressed in a multidisciplinary way in its surgical approach, as was done in the case discussed here. The recommendation also includes long-term follow-up with adequate imaging techniques to control the possible recurrence of the pathology in the areas that may be affected, of special relevance for diagnosis, therapy and prognosis of possible pulmonary or cardiac involvement[4,5].

Conflict of interest

All authors declare no conflict of interest for this publication.

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