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Case Series

Hydropic leiomyoma, a considerable differential diagnosis: a case series

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ABSTRACT

Uterine leiomyomas can show a wide variety of degenerative changes. In the actual work, we report two cases of hydropic leiomyomas. Hydropic leiomyomas are characterized by a massive intra-tumoral accumulation of watery edema. This variant is often associated to atypical clinical and radiological presentations, raising the possibility of aggressive neoplasms, particularly uterine sarcomas. Although imaging can be helpful to diagnose uterine neoplasms, in particular ultrasounds and MRI, the radiological features of Hydropic leiomyomas, as for other leiomyomas variants, are often atypical and worrisome for uterine sarcomas. The pathological evaluation is often needed to have a final diagnosis.

Keywords: Leiomyoma, Hydropic leimyoma, Leiomyosarcoma

INTRODUCTION

The incidence of leiomyomas reaches up to 40% during the fifth decade, making it the most frequent mesenchymal neoplasms of the female genital tract.[1]

Leiomyomas can present histologically with a wide variety of features that can raise a suspicion for uterine sarcomas. These features are secondary to changes such us hyaline, hydropic and myxoid degenerations and calcifications.

Research in the English literature reveals very few cases of leiomyomas with extensive hydropic degeneration.[2-10]

In our work, we report 2 cases of hydropic leiomyomas mimicking aggressive uterine neoplasms, along with a discussion of the radiological, the pathological features and the differential diagnoses of this rare subtype of leiomyoma.

CASE SERIES

In this work, we report two cases of hydropic leiomyomas, in which clinical and radiological features were highly suggestive of malignancy.

The pathological evaluation was the only way to make a final diagnosis of benign leiomyoma with extensive hydropic degeneration.

Case 1

A 46-year-old, married and multiparous woman presented with an abdominopelvic mass. The patient reports a urinary frequency and chronic pelvic pain.

The patient reports no particular medical history, no family history of gynecological malignancies, in particular.

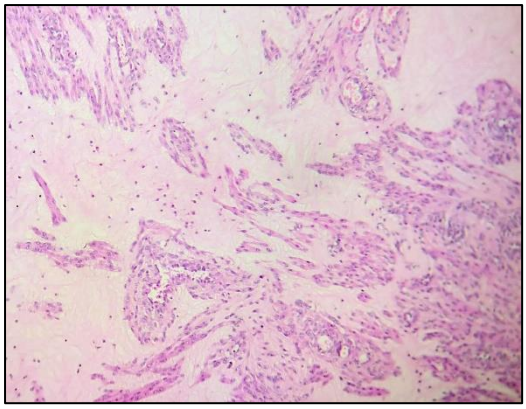

Figure 1: Microphotography from the mass of the first case, showing proliferating smooth muscle bland neoplastic cells, being dissected by an abundant edematous stroma (HE, 200X).

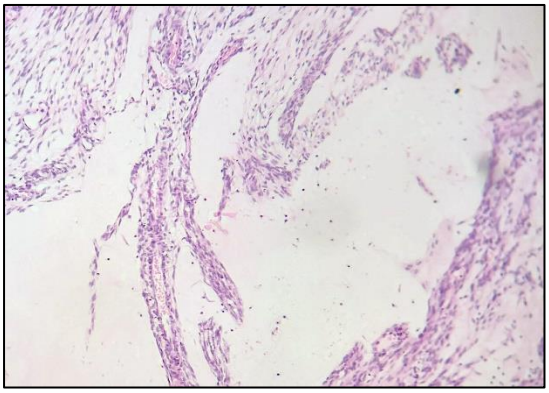

Figure 2: Microphotography showing an edematous stroma dissecting smooth muscle neoplastic cells into thin cords, the co-called felligree pattern (HE, 200X).

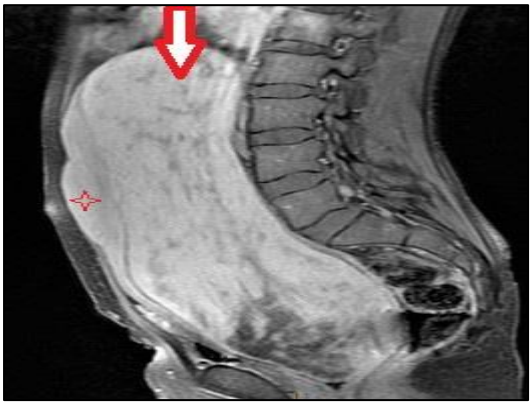

Figure 3: MRI identifying a voluminous well limited abdomino-pelvic mass (257×236×114 mm) (arrow), of the posterior wall of the uterus (star). The mass had heterogeneous enhancement after injection of contrast product in this sagittal section.

Physical examination showed stable vital signs, and abdominal examination a painless, irregular pelvic mass. No lymph nodes were clinically found. Laboratory investigations were within normal limits. Tumor markers were also normal.

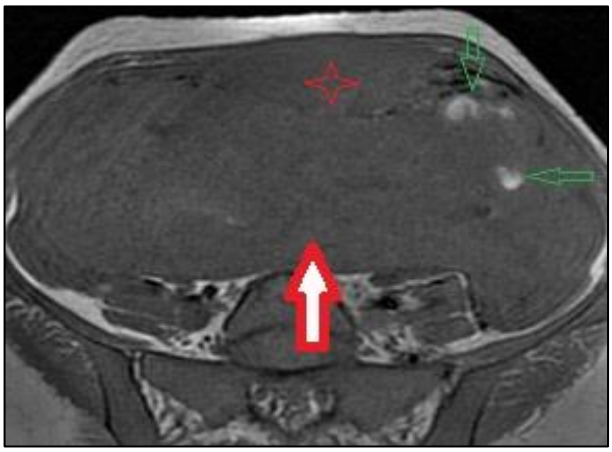

Figure 4: Axial T1 weighted sequence MRI identifying the mass (arrow), merging form the posterior wall of the uterus (star), with visible foci of hemorrhage (green arrows).

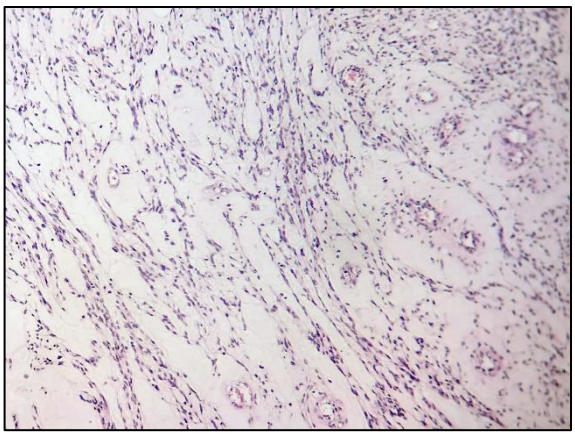

Figure 5: Microphotography from the second case, showing the same filigree pattern, secondary to the abundant edematous stroma (HE, 100X).

An abdominal and pelvic CT scan was performed showing a 16×10×8 cm large mass, with a solid and cystic component. The solid component showed enhancement after contrast administration. The mass was inducing compression on adjacent abdominal and pelvic organs. Examination of the thorax showed no lesions suggestive of metastases.

Magnetic resonance imaging of the pelvis was performed and showed a 15×10×8 cm abdomino-pelvic mass with both a solid and a cystic component, the later showing thick septations. The mass showed an intermediate or low T2 signal intensity with enhancement after Gadolinium administration. A mass effect was observed on adjacent structures, particularly on rectum, uterus and bladder. Both ovaries, and cervix were unremarkable.

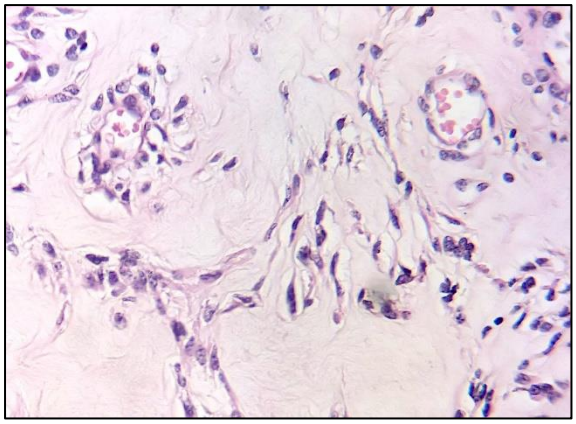

Figure 6: High power view showing bland nuclei and absence of mitoses (HE, 400X).

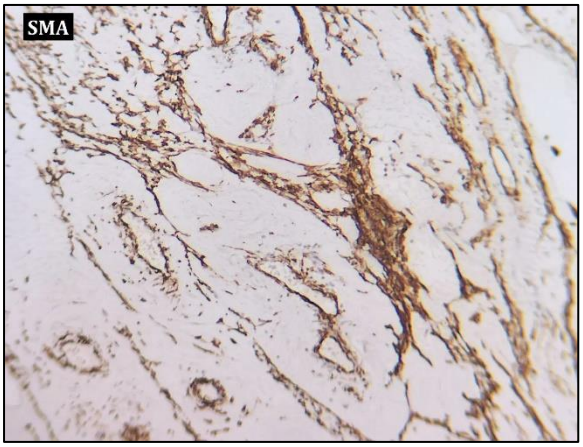

Figure 7: Neoplastic cells showed expression of SMA.

The worrisome radiological features were sufficient to indicate surgery for the patient. A total hysterectomy was performed with no secondary complications. The gross examination of the resected specimen showed a 11×10×7 cm parietal uterine mass. The cut surface of the mass showed a cystic component made of numerous cysts filled with a clear watery fluid. The solid component was nodular with a fascicular appearance and a firm to solid consistency. Foci of hemorrhage were found and occupied 5% of the mass, and many gelatinous foci were identified in contact with the cystic spaces.

Sampling from the solid regions of the mass showed a proliferation suggestive of a benign leiomyoma, with fascicles of smooth muscle cells with no mitosis, necrosis or atypia.

Sampling from the gelatinous and cystic regions showed the same benign smooth muscle cytology as observed on the solid regions. However, an abundant edematous background separated these cells in thin cords. We didn't observe necrosis, mitoses or atypia. A diagnosis of hydropic leiomyoma was made (Figure 1, 2).

After surgery, the patient was discharged on the sixth day after surgery.

Follow-up for one year was uneventful, with a complete resolution of the reported symptoms.

Case 2

This is the case of a 46-year-old, married and multiparous woman, with no family history of gynecological malignancies.

The patient presented with a huge abdomino-pelvic mass, with a history of a progressive abdominal distension, chronic pelvic pain and urinary frequency.

The patient had stable vital signs, and abdominal examination revealed a huge, painless irregular mass occupying the whole pelvic and abdominal cavities. No lymph nodes were clinically found.

Laboratory investigations were within normal limits. Tumor markers were also normal.

An abdominal and pelvic CT scan was performed showing a huge 26×24×12 cm large mass, with a solid and cystic component. The solid component and septas of the cystic component showed enhancement after contrast administration. There was a mass effect on the vena cava with secondary bilateral thrombosis of the iliac veins. Examination of the thorax showed no lesions suggestive of metastases.

On MRI, a voluminous well limited abdomino-pelvic mass (257×236×114 mm), of the posterior wall of the uterus, heterogeneously high on T2-weighted images and diffusion-weighted images without ADC (Apparent Diffusion coefficient) signal dropout, containing some hemorrhagic areas on T1-weighted images. The mass had heterogeneous enhancement after injection of contrast product with some areas of necrosis. (Figure 3, 4)

Since radiological features were worrisome for a sarcomatous nature of the mass and since the presence of the bilateral iliac veins thrombosis, a total hysterectomy was performed

The gross examination of the resected specimen showed a 26×24×13 cm parietal uterine mass of the posterior wall of the uterus. The cut surface of the mass showed a cystic component made of numerous cysts filled with a clear watery fluid. The solid component was nodular with a fascicular appearance and a firm to solid consistency. Foci of hemorrhage were found and occupied 10% of the mass.

Sampling from the solid regions of the mass showed a proliferation suggestive of a benign leiomyoma, with fascicles of smooth muscle cells with no mitosis, necrosis or atypia.

Sampling from the gelatinous and cystic regions showed the same benign smooth muscle cytology as observed on the solid regions. However, an abundant edematous background separated these cells in thin cords. We didn't observe necrosis, mitoses or atypia (Figures 5, 6).

Immunohistochemistry study was performed with anti-SMA, anti-H caldesmon and anti-desmin, confirming the smooth muscle nature of the neoplastic cells (Figure 7).

A diagnosis of hydropic leiomyoma was made. There were no secondary complications to surgery and follow-up for one year was uneventful, with a complete resolution of the reported symptoms.

DISCUSSION

Uterine leiomyomas are the most frequent uterine neoplasms. They present most often as asymptomatic masses in women in their fourth and fifth decades of life. When these neoplasms are symptomatic, they often present with uterine bleeding, pelvic pain or rarely with infertility.[11]

10% of uterine leiomyomas present signs of degeneration, 12 hyaline degeneration being the most frequent and the most described type (60% of cases).

Other types of degeneration include myxoid, hemorrhagic, hydropic, and cystic degenerations.[2,9,12,13]

Hydropic degeneration is the result of watery edema accumulation within the tumor. It is focally reported in up to 50% of leiomyomas.[14]

However, the occurrence of hydropic degeneration in an extensive manner is extremely rare with two described forms: diffuse hydropic cystic degeneration2-18 and perinodular hydropic degeneration.[12,13,15]

The massive accumulation of edema in a leiomyoma can result in giant tumors with difficulties and doubts regarding its uterine origin and it benign or malignant nature especially on radiological features.

The first series of this particular variant was reported by Clement et al, with 10 cases showing symptoms similar to those in patients with typical leiomyomas.[5,16]

In the literature, the age of occurrence ranges between 16 and 58 years old.[5,16]

HLs were also rarely described in pregnant women.[3,8]

Some cases of HLs were associated to an elevation of CA-125 which induces more diagnostic difficulties.[17-19]

The radiological features of HLs are most often different than those observed in classical leiomyomas, and may resemble malignant neoplasms of the uterus.

Ludovisi et al has reported in a large multicenter study that ultrasound characteristics suggestive of malignancy are the inhomogeneous echogenicity, the absence of shadows and calcifications since calcifications are often found in benign leiomyomas, and presence of internal irregular cystic areas.[21]

When the adnexa are not seen in ultrasounds, the origin of the mass cannot be identified, and an MRI can be needed to help identify the organ of origin and the benign or malignant nature of the mass.[9]

Classical leiomyomas show intensity similar to normal myometrium on T1 weighted sequences.[21]

In leiomyomas showing degenerative changes, an increased T2 signal is often focally observed.[21] This anomaly in signal is thought to occur in large tumors that enlarge and outgrow their blood supply.[22, 23]

Thomassin-Naggara et al have discussed the role of MRI diffusion coefficient interpretation, and have shown that it can reach a 92.4% accuracy in distinguishing between benign and malignant uterine tumors.[24]

CT scan has a limited role in the diagnosis or staging of leiomyomas/sarcomas of the uterus.

Although imaging is very helpful, a final diagnosis is often not possible without a pathological examination, showing a massive accumulation of edema, dissecting benign smooth muscle fascicules with the so-called filigree pattern.[11] The blood vessels are often hyalinized. Accumulation of edema can produce many cystic cavities filled with a watery fluid and is responsible for the often-huge size of the tumor.[23] No nuclear or mitotic activity should be observed.[12,13]

Myxoid leiomyosarcoma is one of the main differential diagnoses that can have similar radiological features. On pathological evaluation however, this entity shows a gelatinous rather than edematous gross appearance with obvious infiltration. On microscopic appearance, the presence of nuclear pleomorphism and mitotic activity is the rule.

Other forms of degeneration include the myxoid form of leiomyoma, shows the presence of mucoid/gelatinous foci with cystic areas. A very high T2 signal and mild enhancement after contrast administration are typically observed on MRI.[22]

On the immunohistochemical level, in addition to the positivity for smooth muscle markers (muscle-specific actin, desmin, smooth muscle myosin heavy chain, smooth muscle actin), Griffin et al have described an overexpression of HMGA2 in HLs. This observation was associated to a HMGA2 rearrangement, identified by fluorescence in situ hybridization.[25]

On the therapeutic level, our research in the literature finds myomectomy and hysterectomy as two most used therapeutical methods.

Series by Clement et al have shown an uneventful clinical course in patients with HLs after surgical treatment.[13]

CONCLUSION

Very few cases of leiomyomas undergoing extensive hydropic changes have been reported in the literature. The diagnosis of this entity can be problematic since clinical and radiological features are often atypical. The pathological evaluation is the key of diagnosis, identifying a benign smooth muscle proliferation, dissected by an abundant edematous stroma.

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