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Leiomyosarcoma of broad ligament – a rare case report

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Abstract: *Leiomyosarcomas (LMS) represent a minority of uterine malignancies, comprising merely 1-2% of the total cases. Those of the broad ligament are extremely rare, and highly malignant gynecological tumor. The prognosis of this aggressive tumor is poor. Very few cases are reported in the literature. Ultrasound and MRI help to diagnosis this type of tumors in order to decide the right therapeutic strategy but confirmatory diagnosis need a microscopy examination and immunohistochemical studies. The lake of a therapeutic consensus makes the management of these tumors very complicated especially when it occurs in a young woman with a pregnancy desire. In this article, we report an uncommon case of broad ligament leiomyosarcoma on a young Moroccan woman treated by total hysterectomy with right ovary conservation. Throughout, we highlight the presurgical diagnosis criteria and our experience in a conservatory treatment with an ovary treatment patient consent.*

Keywords: leiomyosarcoma, total hysterectomy, magnetic resonance imaging, broad ligament

Introduction.

Leiomyosarcomas (LMS) represent a minority of uterine malignancies, comprising merely 1-2% of the total cases [1]. However, they constitute 70% of uterine cancer related deaths [2]. LMS are considered the most frequent histological form of uterine sarcomas. Those of the broad ligament are very rare, rapidly progressive and highly malignant gynecological tumor**.** Moreover, they are coupled with poor prognosis, with the survival proportion for five years oscillates between 18.8% to 68%. The clinical diagnosis is very difficult because symptoms cannot be differentiated from common uterine leiomyomas, which is considered the most common tumors of the uterine myometrium. Furthermore, a significant number of leiomyosarcomas are only detected after surgery has been performed under the assumption of treating leiomyomas.[3-5].

Because of its rarity and atypical localization, we present a case of a young woman, that has undergone the management of a primary broad ligament LMS in our hospital.

Case report.

A 30-year-old woman, presented to our hospital with complaints of pelvic heaviness associated with dysmenorrhea and menometrorrhagia for the last 7 months. She had no significant personal or family history and she had used no medications. The general examination found a patient in tolerable general condition, and normal blood pressure, afebrile. Physical examination revealed an enormous palpable and movable mass, measuring almost 13 cm, reaching 2 cm below the umbilicus with no lymph-nodes. The gynecological examination was normal. The laboratory initial workup showed iron deficiency anemia at level 9 g/dl of Hemoglobin, and tumor marker CA-125 was within normal limits.

Transabdominal ultrasonography revealed a heterogeneous left latero-uterine mass, with septations, cystic and solid areas, measuring 09×10 cm. Its origin was not determined by ultrasound, so we performed a pelvi-abdominal MRI.

The sagittal and coronal T2-weighted image in MRI after injection of gadolinium revealed a bulky left latero-uterine mass, well-defined and heterogeneous with two components (tissue and fluid) measuring 103*98*115 mm, in iso signal T2, the fleshy portion is significantly enhanced after injection of gadolinium, which is in hyper signal diffusion with a low ADC (figure 1.a and 1.b). The cervix is lateralized to the left, normal in appearance. Ovaries are in normal size. Absence of hydrosalpinx. The lesion is highly suspected of malignity, with an abdominal exophytic component. In view of the clinical symptomatology and the results of the imaging, we indicated a surgical treatment.

A conservative treatment was decided because the patient had a desire of a pregnancy and she refused a radical treatment (hysterectomy). At intraoperative investigation we found a left sided uterine mass approximately 15cm in diameter, most of which is located in the left broad ligament (Fig 2.a). The bilateral fallopian tubes and right ovary were macroscopically normal, in contrary the left ovary was very adherent to the mass. The patient underwent a left salpingo-ophorectomy with uterine mass total resection (Fig 2.b). Peritoneal cytology was performed.

Histopathologic examination revealed spindle cell proliferation with elongated hyperchromatic nuclei and eosinophilic cytoplasm. The tumor cells were exhibiting prominent cellular atypia. There were some mitotic figures (Fig 3.a) and some areas of necrosis. Numerous multinucleate tumor giant cells were seen (Fig 3.b). The histological features confirmed the diagnosis of broad ligament leiomyosarcoma. Fallopian tube and left ovary were substantially normal.

The case was discussed in a gynecological oncology meeting (RCP), with the decision to carry out a radical treatment and conservation of the right ovary considering her young age. The patient gave her consent for total hysterectomy.
A second surgery consisting on total hysterectomy has been realized and right ovary conservation. The definitive result of the histopathological exam confirmed the diagnosis of LMS broad ligament. The extension workup was negative.
Patient was called upon for follow up; all routine investigations were done and found to be normal. No abnormality was detected on gynecological examination and computed tomography (CT) imaging was also normal. Patient again visited the hospital for follow-up after one year of surgery and no abnormality was detected. After 18 months of follow up, CT imaging was done and was found to be normal; patient now is found to be in good health.

Discussion.

Leiomyosarcoma is the most prevalent subtype of uterine sarcoma. Nevertheless, it represents only 1 to 2% of uterine malignancies [6,7]. A big portion of this percentage appears de novo, while only 0,2% of cases result from sarcomatous transformation in a benign leiomyoma [3]. Gardner et al. define broad ligament leiomyosarcoma as tumors on or in the broad ligament, entirely distinct from the uterus or ovary [8].

The vast majority of leiomyosarcomas patients are women over 40 years old (the overall average age of cases is 60 years, but very rare cases of young women are founded), which contrast our case. Uterine LMS are generally accompanied with irregular vaginal bleeding, palpable pelvic mass and pelvic pain [7]. The broad ligament LMS are clinically manifested by pelvic pain and increased abdominal volume.

Regularly, the pain is a uterine colic accompanies with the passage of blood clots from the vagina, and the intensity of pain could be explained by the short period between the appearance of symptoms and the diagnosis [9]. However, depending on literature reviews, scarcely 0.27–2.6% of women, diagnosed with 'rapidly growing uterus', were confirmed as having LMS [10]. On rare occasions, initial symptoms may involve tumor rupture (manifesting by hemoperitoneum), extrauterine extension, or the presence of metastases [7].
The various researches have analyzed the contribution of B-mode ultrasound and color Doppler in the preoperative diagnosis of uterine sarcomas. Szabo and *al.* compared the indexes and the color Doppler of 129 patients without any differentiating characteristics between fibroids, leiomyosarcomas and carcinosarcomas. Aviram and *al.* showed on a series of 111 patients that there were no morphological differences between the three types of lesions [11].
The MRI delivers better morphologic information on soft tissue intensity than computed tomography. Consequently, MRI might be a crucial tool in preoperative differentiation and defining the suitable treatment [9].
Leiomyosarcomas usually appear, on MRI, as a large infiltrating myometrial mass of heterogeneous hypo intensity on T1-weighted images, with irregular and undefined sides. On T2-weighted images, intermediate-to-high signal intensity is usually noticed, with an excessive central intensity revealing a large necrosis (existed in more than 50% of cases). Furthermore, Hemorrhage is prevalent and centers of calcifications may exist. After contrast administration, they present premature heterogeneous improvement because of the aforesaid zones of necrosis and hemorrhage.
Some authors have highlighted that irregular margins, necrosis, and rapid growth are the most notable characteristics indicating malignancy [3]. Nevertheless, no case has been diagnosed before surgery and the final diagnosis of leiomyosarcoma is based on microscopic examination, supported by IHC studies [6].
Utilizing diffusion-weighted imaging (DWI), malignant lesions can be identified as hyperintense areas with distinct tissue contrast, facilitating quantitative measurements of Apparent Diffusion Coefficient (ADC) values [3]. Tamai et *al.* has claimed important differences in the average ADC values ($\times 10^{-3}$ mm^2/s) of sarcomas 1.17 ± 0.15, which were lower than those of the normal myometrium (1.62±0.11) and degenerated leiomyomas (1.70 ± 0.11), without any overlap [12,13].

The gold standard of management practices of this rare tumor is the same of uterine leiomyosarcoma based on radical surgery type total hysterectomy and bilateral salpingoophorectomy [14].

Based on the success of the primary resection, the recovery rates for these patients vary between 20 and 60%. Consequently, the primary surgery is very important to increase the chances of recovery [15].

Resection of the ovaries and lymph node dissection have always been a debated topic since metastases of these organs rarely occurs and are often related to intra-abdominal disease [7]. Apparently, the role of bilateral salpingo-oophorectomy, for younger women with normal ovaries, is unclear. In parallel, a recent study has claimed that the preservation of the ovary is not associated with poor oncological outcomes [16]. Moreover, it is possible to preserve the ovary for women in perimenopause having early phase LMS, in order to avoid the morbidity related to iatrogenic menopause [7]. We decided to follow the same therapeutic strategy in our young patient and respected her right ovary.

The positive surgical margin noticeably contributes in the risk of local relapse (LR) for patients having uterine LMS. On the other hand, the pelvic RT decreases this danger and leads to an enhancement in overall survival (OS). In parallel, surgery optimization and RT can lead to a significant reduction in important diseases and deaths secondary to pelvic LR. Despite the crucial role of RT

in the enhancement of local control and OS of patients having LMS, a significant portion of survival patients appear to have distant metastases (DM) afterwards. Hence, innovative and efficient approaches shall be invented to fight against systemic metastases [12]. Without surgery, the role of radiation therapy and chemotherapy is restricted. According to new studies; 55% of patients, with recurrent or surgically unresectable uterine LMS, respond positively to the association of gemcitabine and docetaxel [16]. It seems that Adjuvant radiotherapy (RT) has a limited clinical value for women with uterine LMS resection in early or advanced phase [17]. Our patient had a low-grade tumor and did not receive any adjuvant therapy.

Conclusion.

LMS of broad ligament is a rare and aggressive tumor. Ultrasound and MRI help to diagnosis this type of tumors in order to decide the right therapeutic strategy but confirmatory diagnosis need a microscopy examination and immunohistochemical studies. The gold standard of management practices is based on total hysterectomy and bilateral salpingoophorectomy. A conservation ovary can be realized in early phase LMS to avoid the morbidity related to iatrogenic menopause.

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Conflict of interests.
Authors declare no conflict of interests.

Figures.

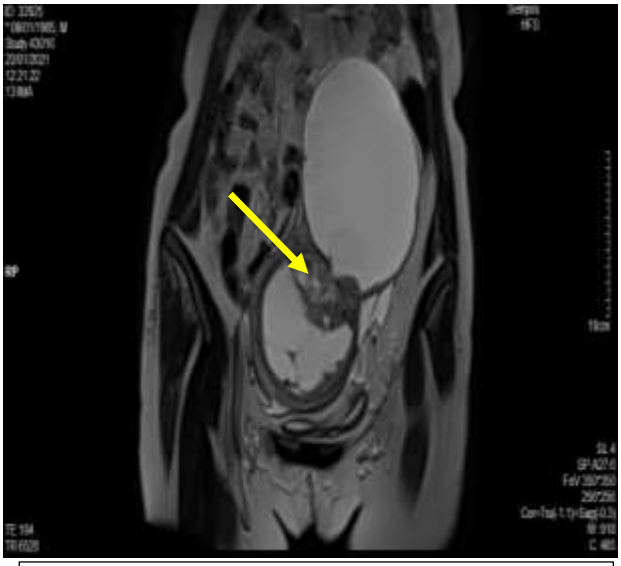

Figure 1.a: coronal T2-weighted image in MRI after injection of gadolinium

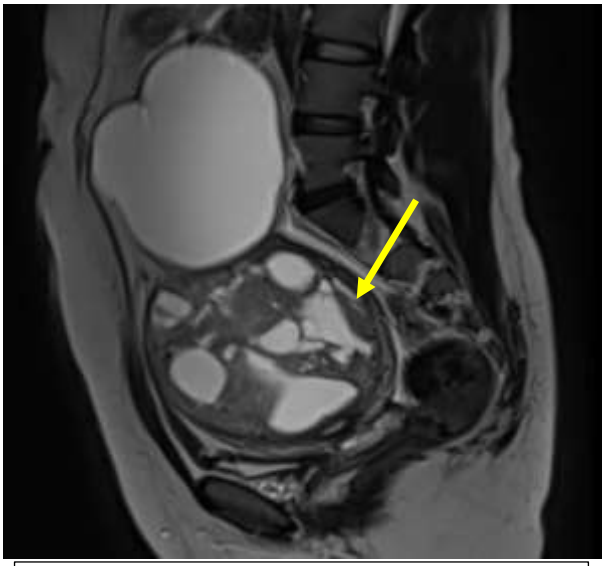

Figure 1.b: sagittal T2-weighted image in MRI after injection of gadolinium

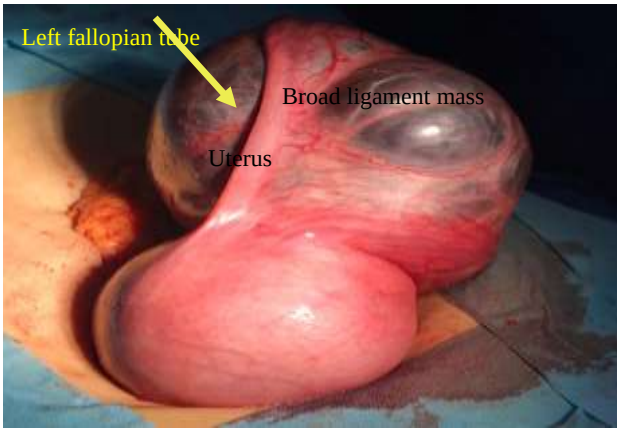

Figure 2.a: left sided uterine mass approximately 15cm in diameter, most of which is located in the left broad ligament

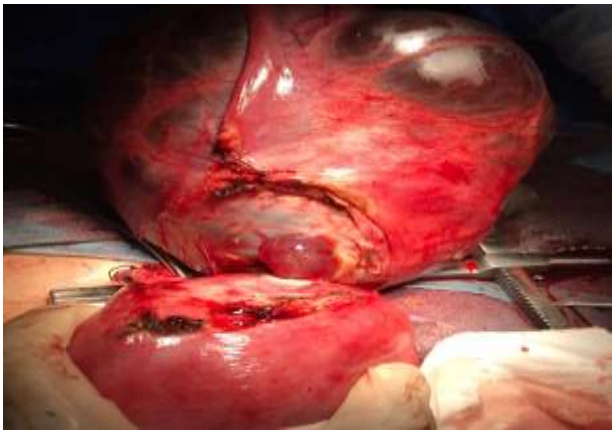

Figure 2.b: left salpingo oophorectomy with uterine mass total resection

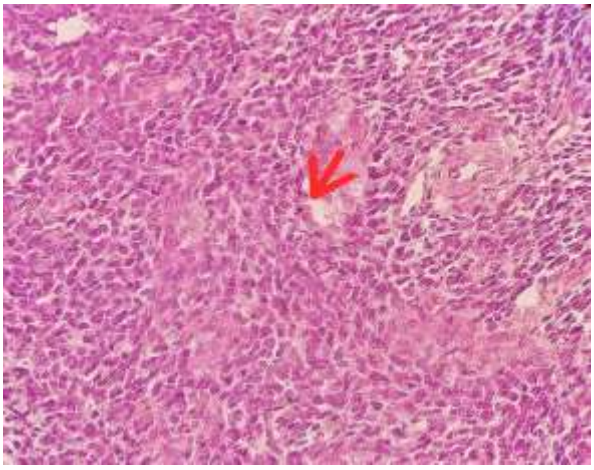

Figure 3.a HE * 20: Spindle-shaped tumor cells are equipped with atypical hyperchromatic nuclei with the presence of a few mitoses (red arrow)

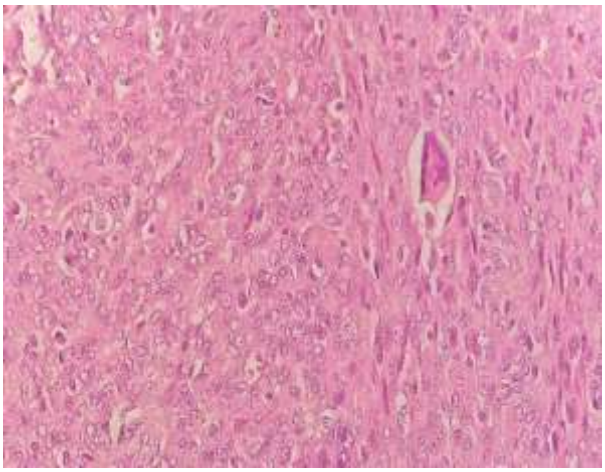

Figure 3.b HE* 20: Tumor proliferation showing atypical pleomorphic multinucleated cells

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