

MYOMETRIAL LEIOMYOSARCOMA - CASE REPORT

JOÃO AFONSO ROSA JÚNIOR, LORENA RODRIGUES NAVES MARTINS SOARES, RAFAELA GIL PIMENTEL, FRANCISCO MAUAD FILHO, AUGUSTO CÉSAR GARCIA SAAB BENEDETI, FERNANDO MARUM MAUAD

ABSTRACT

INTRODUCTION: Leiomyosarcomas are reported between 3% to 7% of all uterine cancers, usually occurring in women over 40 years of age. Furthermore, there are case reports in the literature on young women and even children.

CASE REPORT: The present report addresses a severe and rare case of myometrial leiomyosarcoma in a 32-year-old woman.

DISCUSSION: Tumor staging is the single most important prognostic factor. If we focus on the differentiation between leiomyoma and leiomyosarcoma through two-dimensional and color Doppler ultrasound, it is demonstrated that the detection of hypervascularization combined with other ultrasound findings suspicious of uterine smooth muscle tumors requires an additional diagnostic evaluation before starting treatment.

CONCLUSION: The importance of early diagnosis is noted and it is emphasized that the investigation and follow-up of uterine nodular images, following the recommendation of FEBRASGO, is directly related to the improvement of the prognosis in this population.

KEYWORDS: LEIOMYOSARCOMA, MYOMETRIUM, YOUNG WOMAN, MALIGNANT NEOPLASM, ENDOVAGINAL ULTRASOUND.

INTRODUCTION

Leiomyosarcoma has become the second most common subtype (first is carcinosarcoma) of uterine sarcomas, accounting for only 1% to 2% of these malignancies. Approximately 1:800 uterine smooth muscle tumors are leiomyosarcoma¹⁻⁴.

They are rare, only 13% among uterine sarcomas (when studying the cervix, endometrium and myometrium) in women, and represent 3% to 7% of all uterine malignancies. Although it usually occurs in women over 40 years of age, there are cases in the literature described in young women and even in children⁵.

Leiomyosarcomas have particular evolutionary characteristics: aggressiveness, with hasty dissemination and poor prognosis¹. It is also inferred about the origin of leiomyosarcomas: mesenchymal tissue and malignant degeneration of a leiomyoma².

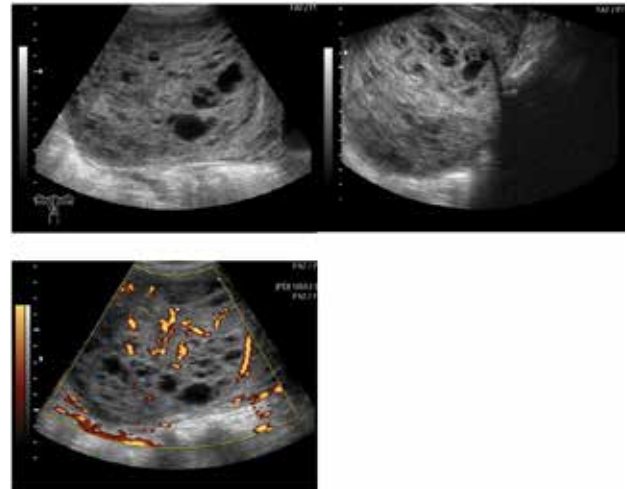
Physical manifestations are commonly vague: genital bleeding, increased uterine volume, and gastrointestinal and urinary complaints. The use of diagnostic imaging evaluation is not accurate to elucidate cases, but rather optimizes the tracking and follow-up of nodular image, suggestive of leiomyosarcoma.

OBJECTIVE

To address a severe and rare case of myometrial leiomyosarcoma in a young woman.

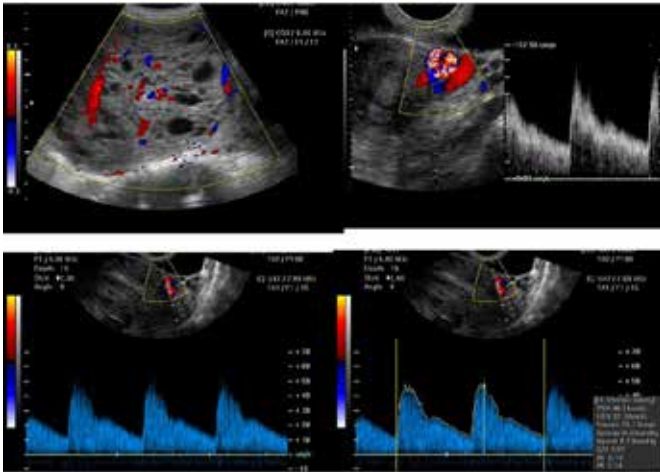
CASE REPORT

A 32-year-old married patient (G1 P1), reporting a two-month menstrual delay, currently with mild vaginal bleeding. Presenting a hemogram with anisocytosis and moderate hypochromia, normal urogram, grade I colpocytology and other normal exams. On ultrasound examination, a nodular image with heterogeneity and intermingled cystic areas is observed in the myometrial region, with the presence of anarchic vascular flow on the Doppler study (figure 1-4).



Figures 1-3: Echographic study demonstrating myometrial nodular image with intermingled cystic areas and anarchic flow on Doppler study.





Figures 4 – Echographic study demonstrating vascularization of nodular image on Doppler study, showing high-resistance vessels.

In view of the reported complaints, in the case of a patient of reproductive age, the clinical thought, why not be a trophoblastic disease? A study with B-hCG was performed, with a negative result.

Another diagnostic possibility would be adenomyosis due to the following findings: globose uterus, intermingling myometrial cysts, loss or irregularity of the endometrial-myometrial junctional zone and translesional vascularization.

The patient was followed up and investigated for a myometrial mass and then referred for surgery (figure 5). The anatomopathological result showed leiomyosarcoma.



Figures 5 – Anatomical part of the total hysterectomy.

DISCUSSION

The World Health Organization (WHO) classifies cancer as one of the leading causes of death in the world. There is evidence that they have multifactorial triggers, including risk factors such as physical inactivity, advanced age, obesity and smoking, associated with these, the use of coloring substances, hormone therapy, menopause and genetic abnormalities that influence the cellular evolutionary cycle and somatic cell

metabolism.

The vascular behavior in leiomyosarcoma should be considered as a fibromuscular neoplasm, because it presents a variable pattern depending on the evolution of the tumor. On Doppler velocimetry, alternating vascular resistivity may be observed depending on perivascular fibrosis. The same vascular behavior can occur with leiomyomas, mainly due to the degenerative processes that they may be subject to⁸.

It should be remembered that myometrial nodular pathology is not always a leiomyoma, and according to FEBRASGO's recommendation, these findings should be followed up with 1) quarterly ultrasound reassessment in the 1st year, and 2) an increase in nodular volume above 25% quarterly suggests leiomyosarcoma.

Despite controversies, most myometrial leiomyosarcomas are not associated with pre-existing leiomyomas and there is no biological evidence associating leiomyosarcomas with benign smooth muscle tumors.

Preoperative imaging tests such as ultrasound and PET scan, using current resources, are incapable of differentiating benign and malignant tumors in smooth muscle, for such magnetic resonance could bring greater subsidies, however, without scientific evidence.

Two main ways of evaluating the malignant potential of leiomyosarcomas are observed, which stand out: tumor size and mitotic index. However, the absolute minimum criteria for malignancy are still not well elucidated.⁵

In an attempt to stage the tumor lesion, as well as assess its prognosis, the following variables were evaluated: patient age, peritoneal cytological findings, cell type, mitotic index of sarcomatous elements, grade of mixed mesodermal tumor sarcoma (MMT), size and location tumor size, depth of myometrial invasion (MMT only), lymphatic-vascular space involvement, adnexal spread, lymph node metastasis, site of recurrence, and adverse effects of surgery.⁶

After studying all factors in a control group, it was noticed that the most effective surgical treatment, except for metastatic lesions, would be extrafascial hysterectomy with bilateral salpingo-oophorectomy. In cases of MMT, selective pelvic and aortic lymphadenectomy may be helpful in determining the need for postoperative therapy. It is a procedure that can be performed without significant morbidity by trained surgeons on properly selected patients.⁶

It was also analyzed for lymph node positivity, strongly suggesting the use of uterine factors to select patients at high risk of recurrence to be included in subsequent adjuvant treatment studies, in a clear attempt to identify a truly effective adjuvant therapy.⁶

CONCLUSION

Despite the low prevalence of leiomyosarcoma in young women and given the aforementioned differential diagnoses, as well as the ultrasound findings, irregular contours and cystic degenerations, the importance of early diagnosis is highlighted and it is emphasized that the investigation and follow-up of uterine nodular images, following the rec-

ommendation of FEBRASGO, is directly related to the improvement of the prognosis in this population.

Therefore, it is concluded that the gold standard for the definitive diagnosis remains the anatomopathological one.

REFERENCES

- 1- Duarte G. Patologia benigna do corpo uterino e das tubas. In: Halbe HW (Ed.). Tratado de Ginecologia. São Paulo: Roca. 1987; p. 710-734.
- 2- Kurjak A, Shalan H, Kupesic S, Predanic M, Zalud I, Breyer B, Jukic S. Transvaginal color Doppler sonography in assessment of pelvic tumor vascularity. *Ultrasound Obstet Gynecol.* 1993; 3: 137-154.
- 3- Exacoustos C, Romanini ME, Amadio A, Amoroso C, Szabolcs B, Zupi E, Arduini D. Can gray-scale and color Doppler sonography differentiate between uterine leiomyosarcoma and leiomyoma? *J Clin Ultrasound.* 2007; 35(8): 449-457.
- 4- Giuntoli RL 2nd, Metzinger DS, DiMarco CS, Cha SS, Sloan JA, Keeney GL, Gostout BS. Retrospective review of 208 patients with leiomyosarcoma of the uterus: prognostic indicators, surgical management, and adjuvant therapy. *Gynecol Oncol.* 2003; 89(3): 460-469.
- 5- Lack EE. Leiomyosarcomas in childhood: a clinical and pathologic study of 10 cases. *Pediatr Pathol.* Boston, Estados Unidos da América. 1986; 6:181-197.
- 6- Major FJ, Blessing JA, Silverberg SG, Morrow CP, Creasman WT, Currie JL, Yordan E, Brady MF. Prognostic factors in early-stage uterine sarcoma. A Gynecologic Oncology Group study. *Cancer.* 1993 15; 71(4): 1702-1709.
- 7- P. J. Figo. Staging for uterine sarcomas. *Int J Gynecol Obstet.* Barcelona, Espanha. Elsevier. 2009; 104(3): 177-178.
- 8- Durand-Réville M, Dufour P, Vinatier D, Martin de Lassalle E, Lucot JP, Monnier JC, Lecomte-Houcke M, Puech F. Uterine leiomyosarcomas: a surprising pathology. Review of the literature. Six case reports. *J Gynecol Obstet Biol Reprod.* 1996; 25: 710-715.