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ENDOMETRIAL STROMA SARCOMA: A CASE REPORT AND REVIEW LITERATURE

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SUMMARY

Low-grade Endometrial Stromal Sarcoma (ESS) is a rare histological entity, accounting for 0.2% of all uterine malignancies, and approximately 7-25% of uterine sarcomas. Because of the absence of pathognomonic clinical and radiological signs, the diagnosis of SES is made retrospectively after analysis of the surgical specimen. The optimal treatment for this condition is still unclear: hysterectomy without adnexal preservation is the standard treatment. There is no consensus on adjuvant treatment. We report the case of a patient diagnosed with low-grade stromal sarcoma of the endometrial stroma in the gynecology department (Marternité les Orangers, RABAT). Based on this rare case and a review of the literature, we describe the epidemiological, diagnostic, therapeutic and prognostic features of this tumor.

KEYWORDS: The optimal treatment for this condition is still unclear: hysterectomy without adnexal preservation is the standard treatment.

INTRODUCTION

Sarcoma is a rare malignant tumor of the uterus. Its incidence is 1 to 2 cases per 100,000 women. The site of origin may be connective tissue, smooth muscle or endometrial stroma. The latter is an even rarer tumor, accounting for 7-25% of uterine sarcomas.[1,2] The World Health Organization (WHO) classifies endometrial stromal tumours into two categories: benign endometrial stromal nodules (BESN), which do not infiltrate the myometrium, and BES, which are characterized by myometrial infiltration. Based on cell morphology and number of mitoses, SES are classified as low- grade or undifferentiated (formerly high-grade) tumors.[3] In contrast to high-grade SES, the mean age of onset of low-grade SES is generally younger (45-55 years). Low-grade SES presents less frequent mitoses.

PATIENT AND OBSERVATION

Mrs W, aged 67, history of hypertension under antihypertensive treatment for 15 years, recently diagnosed diabetes under treatment, menopausal for 13 years. G6P6, 6EV/VB admitted for pelvic pain and low-grade metrorrhagia of 06 months duration, with no associated urinary signs. Clinical examination revealed an obese patient with a BMI of 36, uterus reaching the umbilicus Pelvic ultrasound: uterus increased in size by 16.66/16.80cm, intracavitary necrotic myometrial image measuring 10cm.

Pelvic MRI

Presence of a voluminous intrauterine tissue process involving the endometrium and occupying the entire uterine lumen, measuring 15 cm craniocaudal, 12 cm in diameter.

transverse. This is a largely necrotic process with a thickened wall that clearly takes up contrast medium and is responsible for massive invasion of the myometrium without invasion of the serosa.

Interstitial development measuring 36 mm in diameter Absence of significant pelvic adenopathy No visible pelvic peritoneal effusion. Respect for bladder and rectum.

No bone abnormalities Overall:
Large, suspicious-looking tissue process invading the entire endometrium and myometrium without invasion of the serosa, without adenopathy and without pelvic peritoneal effusion.

HSC + BE (1/6/23): Anapath examination: No endometrial mucosa, **No dysplasia or signs of malignancy**

the cervico uterine smear was free of abnormalities

The patient underwent an exploratory laparotomy, revealing a **large uterus of 22SA, hypervascularized** and approximately 10 cm long, with necrosis. A hysterectomy was performed with bilateral adnexectomy.

On pathological examination, macroscopy revealed a hysterectomy specimen with bilateral adnexectomy.

On cross-section of the body, presence of a tumoral lesion filling the entire lumen, reaching as far as the myometrium, whitish in appearance, renitent, with a yellowish friable zone measuring 14/13/19cm in the center. The friable zone represents 70% of the tumoral surface.

Cross-section of the left ovary showed a cyst with serous content measuring 1/0, 8/0, 3cm. Microscopically, the uterine body shows a proliferation of spindle cells, sometimes round, with high cellularity and diffuse architecture. Nuclei are ovoid, sometimes rounded and moderately atypical. cytoplasm is sparse and eosinophilic. mitotic figures are estimated at 3 mitoses per 10 fields.

Parameters, adnexa and cervix free of tumor infiltration An immunohistochemical study showed:
Anti -CD10 antibody: positive and diffuse marking of tumor cells Anti-H caldesmone antibody: positive marking of tumor cells Anti-RE antibody: positive marking of tumor cells.
Anti-Cylin D1 antibody: positive labeling of rare tumor cells Anti-Actin antibody: positive labeling of tumor cells.
Conclusion: Morphological and immunophenotypic appearance consistent with low-grade endometrial stromal sarcoma with smooth muscle differentiation classified as PT1b (TNM classification) and FIGO stage IB.

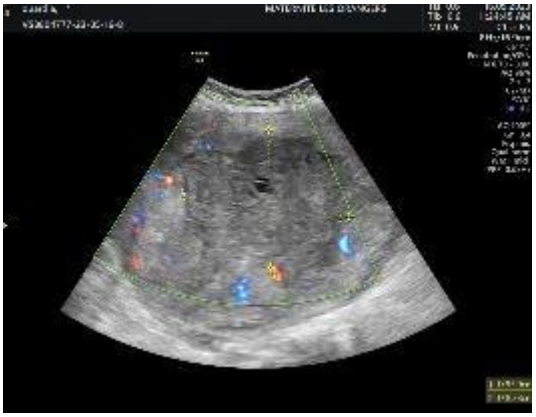
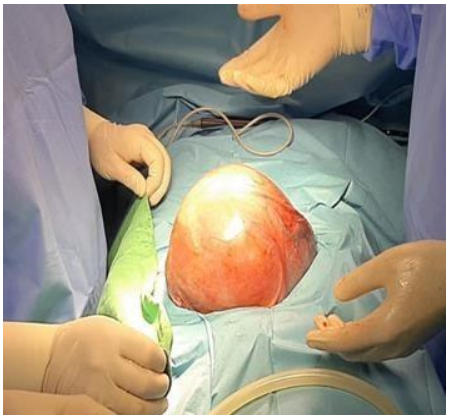


Figure 1: pelvic ultrasound showing a 10cm necrotic intracavitary myometrial image.



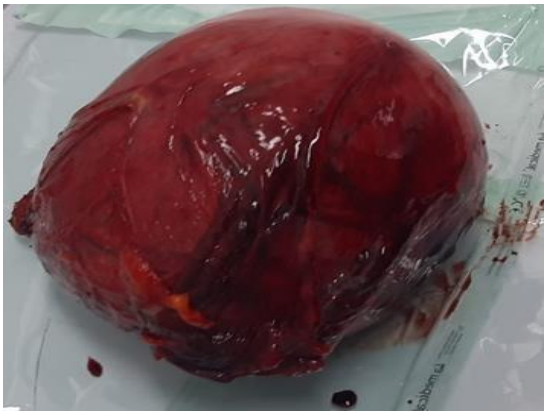


Figure 2 and 3: images showing the macrospic appearance of the mass Patient staffed at multidisciplinary consultation meetings: Decision to refer patient for chemotherapy.

DISCUSSION

Low-grade Endometrial Stromal Sarcoma (ESS) is a histological subtype of uterine sarcoma. Uterine sarcoma is a heterogeneous group of rare tumors of the uterine muscle and connective tissue.

Endometrial stromal sarcoma (ESS) accounts for 0.2% of all uterine malignancies, and approximately 7-25% of uterine sarcomas.[2,6] Annual incidence is 0.19 per 100,000 women.[7] ESS is the second most common type of uterine mesenchymal neoplasia after uterine leiomyosarcoma. According to the WHO classification published in 2014, low-grade SES is a subclass of Endometrial stromal tumors, along with benign endometrial stromal nodules, high-grade Endometrial sarcoma, and undifferentiated uterine sarcoma. Low-grade SES usually occurs in postmenopausal or peri-

menopausal women with an average age between 45 and 55.[8] Our patient was 67 years old. Rare cases of SES have been reported following administration of tamoxifen, estrogen or radiotherapy.[9,10] Obesity, type II diabetes and early menarche are also risk factors for low-grade SES.[11] Our patient presented with type 2 diabetes, hypertension and obesity. The clinical picture was polymorphous with no pathognomonic signs, dominated by metrorrhagia and chronic pelvic pain, sometimes associated with signs of compression such as dysuria or constipation.[12] Our patient had a history of low-grade SES.

In our case, the symptomatology consisted of pelvic pain associated with metrorrhagia. Ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI) cannot be used to diagnose low- grade SES, due to the absence of specific signs known to date. The diagnosis is often made retrospectively after anatomopathological analysis of the surgical specimen.[13]

The reference treatment for low-grade SES is surgical, and consists of a hysterectomy. by laparotomy, in order to avoid fragmentation during vaginal resection.[14] Bilateral annexectomy is justified in view of the hormone-dependent nature of these tumours.

Several studies have described a higher recurrence rate when the ovaries were preserved in non- menopausal women.[15,16] Pelvic and para-aortic lymph node involvement does not appear to influence prognosis.

Lymphadenectomy Reduces the number of pelvic recurrences, without reducing intra-abdominal localization and distant metastases.[7] However, nearly 10% of patients who undergo who did not undergo lymphadenectomy had lymph node metastases, according to Chan et al.[15,18,19] Lymphadenectomy does not improve survival.

Medroxyprogesterone, GnRH analogues and antiaromatases appear to be effective as adjuvant medical therapy, due to the expression of hormone receptors and aromatases by low-grade SES.[19,20] Postoperative radiotherapy appears to improve only locoregional control of the disease, without modifying the overall survival rate.

The indication must be determined on the basis of side effects.[21]

Recurrences are estimated at 10-20% of cases, of which 40% are local (vagina or lesser pelvis), and 60% distant (intra-abdominal or lung).[22,23] The time to recurrence is 5.4 to 9.3 years for stages I and II, and only for stages III and IV.[23,24] There are also forms of late recurrence, described 10 to 30 years after treatment.[24,25] Surgery, radiotherapy and chemotherapy can be used for locoregional and distant recurrence, but only surgery with complete resection can improve cure rates and overall survival. Surgery for metastases should also be discussed.[14]

CONCLUSION

Like other uterine sarcomas, low-grade SES is a very rare tumor entity. Hysterectomy or bilateral adnexectomy is the first-line treatment. The benefits of lymphadenectomy or tumor cytoreduction are unclear. Medroxyprogesterone and aromatase inhibitor therapies are under discussion as adjuvant treatments. Radiotherapy enables locoregional control, and must be carefully indicated in view of side effects. Surgical revision is also the treatment of choice in the event of recurrence. Other therapeutic options need to be evaluated to improve management.

REFERENCES

1. Ashraf-Ganjoei T, Behtash N, Shariat M, Mosavi A. Low grade endometrial stromal sarcoma of uterine corpus, a clinicopathological and survey study in 14 cases. World J Surg Oncol, August 9, 2006; 4: 50.
2. Hosh M, Antar S, Nazzal A, Warda M, Gibreel A, Refky B. Uterine Sarcoma: Analysis of 13,089 Cases Based on Surveillance, Epidemiology, and End Results Database. Int J Gynecol Cancer Off J Int Gynecol Cancer Soc., Jul 2016; 26(6): 1098-104.
3. F L, Jj T, Z A, A G, C L. [Uterine sarcomas: clinical and histopathological aspects. Report on 15 cases]. Gynecol Obstet Fertil, June 5, 2008; 36(6): 628-35.
4. Endometrial Stromal Sarcomas: A Review of Potential Prognost...: Advances in Anatomic Pathology [Internet]. [cited 25 Apr 2021]. Available from: https://journals.lww.com/anatomicpathology/Abstract/2010/03000/Endometrial_Stromal_Sarcomas A_Revie w_of.3.aspx
5. Park J-Y, Park S-K, Kim D-Y, Kim J-H, Kim Y-M, Kim Y-T, et al. The impact of tumor morcellation duringsurgery on the prognosis of patients with apparently early uterine leiomyosarcoma. Gynecol Oncol, August 2011; 122(2): 255-9.
6. An Evaluation of Prognostic Factors in Uterine Carcinosarcoma - ScienceDirect [Internet]. [cited 25 Apr 2021].
7. Available at: https://www.sciencedirect.com/science/article/abs/pii/S0090825897948757
8. Harlow BL, Weiss NS, Lofton S. The epidemiology of sarcomas of the uterus. J Natl Cancer Inst., March 1986; 76(3): 399-402.
9. Matsuura Y, Yasunaga K, Kuroki H, Inagaki H, Kashimura M. Low-grade endometrial stromal sarcoma recurring with multiple bone and lung metastases: report of a case. Gynecol Oncol., March 2004; 92(3): 995-8.
10. Beer TW, Buchanan R, Buckley CH. Uterine stromal sarcoma following tamoxifen treatment. J Clin Pathol, June 1995; 48(6): 596.

11. Meredith RF, Eisert DR, Kaka Z, Hodgson SE, Johnston GA, Boutselis JG. An excess of uterine sarcomas after pelvic irradiation. Cancer, Nov 1, 1986; 58(9): 2003-7.
12. Hoang L, Chiang S, Lee C-H. Endometrial stromal sarcomas and related neoplasms: new developments and diagnostic considerations. Pathology (Phila). feb 2018; 50(2): 162-77.
13. Thiel FC, Halmen S. Low-Grade Endometrial Stromal Sarcoma - a Review. Oncol Res Treat., 2018; 41(11): 687-92.
14. Amant F, Coosemans A, Debiec-Rychter M, Timmerman D, Vergote I. Clinical management of uterine sarcomas. Lancet Oncol, Dec 2009; 10(12): 1188-98.
15. Stadsvold JL, Molpus KL, Baker JJ, Michael K, Remmenga SW. Conservative management of a myxoid endometrial stromal sarcoma in a 16-year-old nulliparous woman. Gynecol Oncol, 1 Oct 2005; 99(1): 243-5.
16. Li N, Wu L-Y, Zhang H-T, An J-S, Li X-G, Ma S-K. Treatment options in stage I endometrial stromal sarcoma: a retrospective analysis of 53 cases. Gynecol Oncol, Feb 2008; 108(2): 306-11.
17. Amant F, De Knijf A, Van Calster B, Leunen K, Neven P, Berteloot P, et al. Clinical study investigating the role of lymphadenectomy, surgical castration and adjuvant hormonal treatment in endometrial stromal sarcoma. Br J Cancer, Nov 5, 2007; 97(9): 1194-9.
18. Köhler G, Evert M. Uterine Sarkome und Mischtumoren: Handbuch und Bildatlas zur Diagnostik und Therapie. Walter de Gruyter, 2009. 209 p.
19. Mesenchymal tumors of the uterus. I. A clinical and pathological study of 53 endometrial stromal tumors - Norris - 1966 - Cancer - Wiley Online Library [Internet]. [cited 25 Apr 2021]. Disponible sur: https://acsjournals.onlinelibrary.wiley.com/doi/abs/10.1002/1097-0142(196606)19:6%3C755::AIDCNCR2820190604% 3E3.0.CO;2-U
20. Thomas MB, Keeney GL, Podratz KC, Dowdy SC. Endometrial stromal sarcoma: treatment and patterns of recurrence. Int J Gynecol Cancer Off J Int Gynecol Cancer Soc., Feb 2009; 19(2): 253-6.
21. Chu MC, Mor G, Lim C, Zheng W, Parkash V, Schwartz PE. Low-grade endometrial stromal sarcoma: hormonal aspects. Gynecol Oncol, Jul 2003; 90(1): 170-6.
22. Denschlag D, Thiel FC, Ackermann S, Harter P, Juhasz-Boess I, Mallmann P, et al. Sarcoma of the Uterus.