LOW-GRADE ENDOMETRIAL STROMA SARCOMA DIAGNOSIS AND MANAGEMENT

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SUMMARY

Endometrial stroma sarcoma (ESS) is a rare histological entity, accounting for only 1-2% of endometrial malignancies. Diagnosis is based on morphological and immunohistochemical criteria, with increasing use of molecular biology (FISH, PCR or sequencing by RNA-seq, CGH array, etc.). It is important that the specimens are quickly sent to pathology in a fresh state for their treatment.

The pathology study concluded to the diagnosis of low grade ESS of malignancy, peritoneal location. In a woman over 40 years of age with imaging in favor of a single fibroid, it is necessary to fragment the piece in a suitable bag to avoid dissemination in the peritoneum. The objective of this study was to highlight the difficulty of evoking the diagnosis of ESS, especially in case of ectopic localization, to describe the clinical, radiological, histological and immunohistochemical aspects with a review of the literature.

KEYWORDS: Endometrial stroma sarcoma, Low-grade, Hysterectomy.

INTRODUCTION

Uterine sarcomas are rare tumors representing less than 3% of malignant tumors of the female genital tract, and their incidence is estimated to be between 4% and 9% of malignant tumors of the uterine body. Uterine sarcomas derive from the mesenchymal elements of the uterine body: smooth muscle and endometrial stroma or more rarely from mesenchymal elements unsuitable for the uterus (cartilage, bone, striated muscle). They can be associated with epithelial tumors giving rise to mixed tumors.
(carcinosarcoma and adenosarcoma). The classification adopted for these tumors is that of the WHO, the latest version of which dates from 2003.[2-4] This is a group of tumors, comprising different histological subtypes with tumors that may be of pure conjunctive origin such as leiomyosarcomas (LMS); the other subtypes are endometrial stroma sarcomas (ESS) and undifferentiated sarcomas. ESS are rare sarcomas and have a better prognosis than other uterine sarcomas with a 5-year survival of 57-70%. There are low-grade endometrial stroma sarcomas occurring in younger women, often with good prognostic hormone receptor expression, and undifferentiated endometrial stroma sarcomas occurring at a later age, with high mitotic activity and/or tumor necrosis, these are tumors with a very poor prognosis. The objective of this work is to clarify the clinical, diagnostic, therapeutic and prognosis difficulties of low-grade endometrial stromal sarcoma.

**Comment**

This is Mrs RH aged 48 years old, with no notable pathological history, G5P5(G1 is a cesarean delivery 20 years ago for acute fetal suffering, G2, G3, G4 and G5 are low route deliveries, with 5 live children. The patient has had menometrorrhagia with pelvic pain for 3 months, without any other associated signs.

General examination found a conscious patient, a pyretic, with normal blood pressure: 11/6 mmhg, slightly tachycardia at 100 beats per minute, discolored conjunctiva with cutaneous-mucosal pallor. Gynecological examination: Speculum inspection finds medium-abundant bleeding from the cervix with normal appearance and no vaginal lesions. Vaginal examination with abdominal palpation finds an enlarged uterus with no latero-uterine mass on the way to the umbilicus. The rest of the examination is without peculiarities. A complete check-up has been carried out which showed anemia at 8 g/dl the rest of the check-ups are correct. Ultrasound revealed an enlarged anterior uterus with irregular contours and the presence of a fibroid of the posterior wall measuring 6 cm in diameter classified as type 3 by FIGO. The patient is scheduled for myomectomy and transfusion of 2 red blood cells before surgery, and the piece is addressed to the anatomopathological study which was in favor of a low grade endometrial stroma sarcoma with smooth muscle differentiation (Figure 1,2,3,4,5). She was scheduled one week later for total hysterectomy with bilateral adnexectomy. The postoperative follow-up was simple and the patient was discharged with control in one month.
Figure 1: Tumor cells showed diffuse CD10 immunostaining,*100.

Figure 2: Tumor cells showed no H-Caldesmone immunoreactivity,*100.
Figure 3: Tumor showed diffuse smooth Muscle Actine immunostaining,*100fig.

Figure 4: The tumor is composed of diffuse infiltrates of small, nearly uniform cells with oval to spindle-shaped hyperchromatic nuclei (Hematoxylin ans Eosin,*100).
Figure 5: Tumor cells showed strong nuclear immunoreactivity for Estrogen Receptor,*100.

Figure 6: Tumor cells showed strong nuclear immunoreactivity for Progesterone Receptor,*100.
DISCUSSION

Sarcomas of the endometrial stroma are formed by a malignant proliferation of cells reminiscent of the endometrial stroma in its proliferative phase. Depending on the histological grade, a distinction is made between undifferentiated sarcoma (formerly known as high-grade SES) and low-grade SES. In fact, the symptomatology often mimics that of fibroids, with a predominance of menometrorrhagia (45-86%) and pelvic pain (20-50%). Several authors have been interested in identifying the clinical features that make it possible to distinguish uterine sarcomas from fibroids before surgery. The conservative attitude often adopted in the management of fibroids can lead to a fragmentation of the tumor, which has a negative impact on the evolution.[5] Schwartz et al[6] recommend close and regular monitoring of the largest myoma during conservative treatment of a polymyomatous uterus.

A. Hassini et al[2] emphasizes the importance of regular monitoring of all women with uterine fibroids as a component of sarcoma screening. They recommend that this diagnosis should be considered in the face of rapid increase in fibroid volume, especially in postmenopausal patients, in the face of recurrent myomas that recur several times within a short period of time, or in the face of recurrent cervical necrosis.
Both imaging and clinical practice are non-specific\cite{7}; Indeed, the ultrasound appearance of uterine sarcoma is no different from that of fibroma. Nevertheless, the appearance of the fibroma as reworked and the rapid increase in the size of a fibroid should make the diagnosis suspicious, also Pelvic CT scan is similarly non-specific; Magnetic resonance imaging can help guide the diagnosis when conventional imaging suggests an atypical uterine mass. It also allows a local extension assessment to be carried out and is the first examination to be requested to distinguish between a recurrence and post-therapeutic changes.

Diagnosis is based on morphological, immunohistochemical and even molecular biology criteria. In the event that molecular biology analyses are required, it is important that the samples are quickly sent to pathology in a fresh state.

There is no specific functional symptomatology for uterine sarcomas (metrorrhagia, pelvic pain). Quite often, a rapid increase in a fibroid or necrotic tumor delivered by the cervix can be noted. Risk factors for sarcoma are: history of tamoxifen use, pelvic radiotherapy, any metrorrhagia, especially after menopause, requires exploration.

For this type of tumor there is a low sensitivity of the endometrial biopsy because it only allows an abrasion of the endometrial mucosa, not allowing the diagnosis of sarcomas deriving from the uterine muscle or mesenchyma (except if invasion of the associated endometrium). From a radiological point of view, there is currently no examination to guide us towards the nature of the disease and the histological type; it is therefore essentially used to assess the extent of the disease.

The diagnosis is often made a posteriori on a specimen removed for suspected fibroma, for example; reoperation must be discussed again after an extension imaging work-up and discussion at a multidisciplinary consultation meeting; in the absence of macroscopic lymph node invasion, there is no indication of lymph node removal. The consensual treatment of localized sarcomas remains surgery and includes a total colpohysterectomy with bilateral adnexectomy by laparotomy; the vaginal route must be avoided as it does not allow peritoneal staging.

Surgery is the first step in the management of uterine sarcomas. Laparotomy is the preferred approach because it reduces the risk of tumor fragmentation and peritoneal or vaginal spread. The reference surgical intervention in the case of a tumor that is, a priori, limited to the uterine body is total hysterectomy with bilateral adnexectomy.\cite{8,9} For advanced stages surgery is discussed, however, hysterectomy with bilateral adnexectomy appears to be indicated when technically feasible.
All the published series on the place of adjuvant radiotherapy in the treatment of uterine sarcomas have concluded that adjuvant radiotherapy is beneficial in terms of local control, but not in terms of overall survival. Nevertheless, the reduction in the number of pelvic recurrences, often accompanied by pain, may justify the prescription of adjuvant radiotherapy, particularly for tumors of high histological grade. The value of chemotherapy remains uncertain.

CONCLUSION
Endometrial stroma sarcomas are rare tumors, representing about 0.2% of all genital tract tumors. In practice, diagnostic problems are mostly encountered in recurrences or metastatic lesions, which can occur more than 10 years after the initial removal of the lesion and can then easily be confused with other sarcomas. This distinction is extremely important from a clinical point of view as LG ESS, even if metastatic, is a lesion with a good prognosis, normally responding favorably to hormone therapy.

REFERENCE