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Uterine Sarcoma: Retrospective Study on 10 Cases and Review of Literature

D Kasmi[1*], A Ahallat[2], A Abida[1], M Yousefi[3], S Bargach[3]
[1]Resident in department of gynecology obstetrics, cancer research and pregnancy at high risk, Maternity SOUISSI Rabat
[2]Resident in visceral surgery, national institute of oncology Rabat
[3]Professor in department of gynecology obstetrics, cancer research and pregnancy at high risk, Maternity SOUISSI Rabat

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***Corresponding author**
D Kasmi

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Abstract: Uterine sarcomas are malignant tumors with poor prognosis. The purpose is to clarify the epidemiological, diagnostic difficulties and therapeutic modalities and prognosis of uterine sarcomas. We analyse Ten cases of uterine sarcomas supported in Maternity SOUISSI Rabat (2011-2016). The average age is 55.8 years. Patients viewed had the clinical triad of bleeding, pain and / or pelvic mass. On the para-clinical, eight patients had an inconclusive ultrasound, one had a pelvic scan four was complete with MRI to show a pelvic mass. The realization of the biopsy endometrial curettage led to the diagnosis. All patients underwent a hysterectomy associated with lymph node dissection in one case. Histological analysis of surgical specimens objectified, 08 leiomyosarcoma, 01 endometrial stromal sarcoma, 01 mixed mullerian tumor. The classic clinical triad, reported by most authors, was found in the majority of patients. MRI and PET scan are the most successful exams. The diagnosis rests on hysterectomy's piece. For treatment, surgery with hysterectomy and adnexectomy with or without lymph node dissection followed by external radiation therapy is the gold standard. Indeed, uterine sarcomas are malignant tumors with poor prognosis whose diagnosis is primarily postoperative. Surgery with radiation therapy is the gold standard. Hope rests on new drugs that are being tested.
Keywords: uterine sarcomas, pelvic MRI, curettage biopsy, hysterectomy, bilateral adenexectomy.

INTRODUCTION

Uterine sarcomas are a rare entity, with a reserved prognosis. Pathologically, it is a heterogeneous group consisting of several histological types.

The anatomopathological diagnosis is made by endometrial biopsy either by hysteroscopy, biopic curettage or with the help of an endometrial biopsy pipelle but in some cases can only be done on the operative part.

The treatment is essentially surgical.

MATERIALS AND METHODS

Epidemiology

We have reported 10 cases diagnosed in our training between 2011 and 2016. The main age of our patients was 55.8 with extremes between 44 and 73 years, the average age of the LMS was 56.6 years and the SES was 48 years old. 7 out of 10 patients were postmenopausal or 70%. 2 in perimenopause and 1 unspecified.

Diagnosis

The functional signs are dominated by metrorrhagia (90%) followed by pelvic pain (20%) and abdominal-pelvic masses (30%), of which 2 patients had a mass delivered by the cervix, concerning the physical signs 70% of patients had an enlarged uterus.

Pelvic ultrasonography was performed on 8 patients objectifying either an intra-cavitary mass with increased uterus, or a heterogeneous endometrial thickening hypervascularized Doppler or cervical isthmic myoma. Pelvic CT was performed on one patient, indicating the presence of a uterine process.

The MRI complement was performed on 4 patients but it was not conclusive for the diagnosis of uterine sarcoma. Pathologically, the diagnosis was made by biopsy curettage of the endometrium in 04 patients, and by enucleation of the cervical mass in 2 patients.

The chest X-ray and abdominal CT, was performed as part of the extension assessment, which went back to normal in all patients.

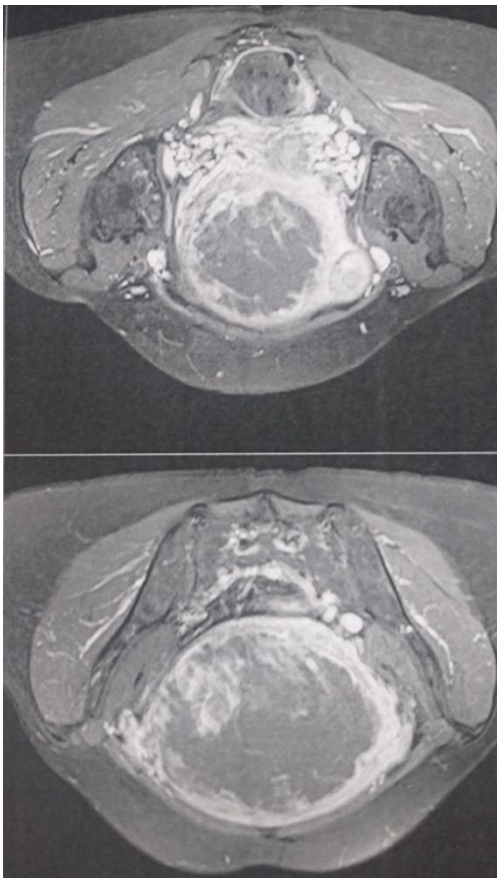

Fig-1: PELVIC RMI of q leiomyosarcoma

TREATMENT

Surgery

All our patients underwent total hysterectomy with bilateral adnexectomy associated, in 01 cases, with pelvic dissection where the diagnosis of Mullerian tumor was made intraoperatively. The anatomopathological's result of the operative specimens showed:

Mixed Mullerian tumor: 01 cases

Leiomyosrcoma: 08 (02 grade II and 01 grade III)

Stromal sarcoma: 01 case (grade I)

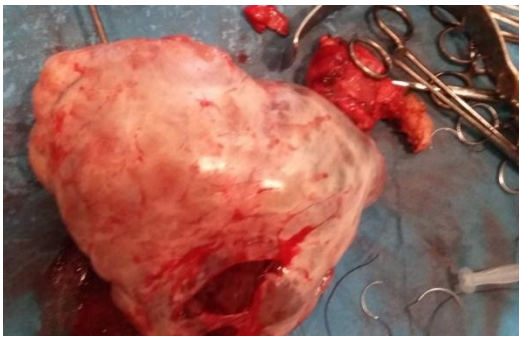

Fig-2: operating part of leiomyosarcoma

Radiotherapy

Realized in 04 patients postoperatively in front of a ganglionic invasion and / or a big tumor

Chemotherapy

03 of our patients received adjuvant chemotherapy.

EVOLUTION

After an average follow-up of 24 months, 04 patients were lost to follow-up, 03 died at intervals of 08, 09, and 10 months, 3 patients are always followed with good locoregional control.

DISCUSSION

Epidemiology

Impact

Uterine sarcomas are rare tumors, representing less than 3% of malignant tumors of the female genital tract and between 3 and 7% of malignant tumors of the uterine body.

Age

Uterine sarcoma can occur throughout life [1], between 24 and 95 years of age with an average age of 60.5 years for Olah *et al*. [2]; between 15 and 85 years old with a median of 54 years for L. Carvalho *et al*. in Portugal [3].

Risk factors

Some factors are discussed as risk factors for occurrence of uterine sarcoma: The black race; pauciprity; menopausal status; obesity and high blood pressure.

DIAGNOSIS

Diagnostic confirmation is often obtained by anatomopathological study of the operative specimen of a myomectomy or hysterectomy.

Clinic

Symptoms that may reveal uterine sarcoma are variable and nonspecific. The most frequently found signs are: 3.2.1.1 GENITAL HEMORRHAGIA

They are found in 90% of patients in the Olah series [2]; 70% of the patients of the Turkish series of D. Etiz [3]; 92% of our patients consulted during a genital hemorrhage.

Abdomino-pelvic mass

It is reported in 20% of the series of Olah [2], and 13% of the series of Etiz [3]. 30% of our patients had an abdominopelvic mass.

Pelvic abdomino pain

Often gravity type: This symptom is found in 20% of our patients, at a higher frequency than those of other series: Olah (27%) [2] and the series of Etiz (16%) [3].

Imaging

Ultrasound

Ultrasonography is weak, usually a heterogeneous non-specific, cystic and non-specific lesion of uterine sarcoma [4].

Tomodensitometry

It does not show a specific sign in favor of sarcoma, often it is a mass of hypodense aspect.

MRI

The characteristic MRI pattern of uterine sarcomas is a T1-weighted hypersignal and an intense or moderate heterogeneous signal in T2-weighted sequence.

ANATOMOPATHOLOGY

Biopsy guided by hysteroscopy and / or biopic curettage allows diagnosis only in case of endometrial involvement [5].

Several cases of uterine sarcomas have been diagnosed following hysteroscopic resection of the endometrium for treatment of bleeding after failure of medical treatment [6]. In other cases, the histological confirmation of uterine sarcomas is made during a hysterectomy or myomectomy for uterine fibroids. In our series, the anatomopathological diagnosis was made preoperatively by biopsy curettage of the endometrium in 04 cases (40%) and cervical mass enucleation in 02 cases, for the rest the diagnosis was made on the hysterectomy specimen.

Extending balance sheet

Uterine sarcoma is known to have a haematogenous swarm, so it is essentially pneumophile and hepatophilic. When an epithelial component is associated, swarming is essentially lymphophilic, so extension can be assessed by thoracoabdominal CT and pelvic MRI.

TREATMENT

Methods

Surgery

The purpose of this surgery is to perform an excision of the uterine tumor without breaking it up. If the diagnosis is strongly suspected or known before the initial surgical procedure, the chosen approach must allow exploration of the entire abdominopelvic cavity and excision of the uterus in monobloc to reduce peritoneal dissemination. or early postoperative vaginal, that is related to fragmentation of the part during vaginal extraction; the recommended route is therefore a median and not a pfannentiel or vaginal [7].

Surgical modalities: Exploration of the entire abdominopelvic cavity in search of locoregional or general extension. Collection of suspicious elements and peritoneal cytology are required [8].

Total hysterectomy with bilateral adnexectomy is the standard procedure [9]. It has been performed in all our patients, associated with lymph node dissection in some cases.

Radiotherapy

Postoperative external radiotherapy is the most widely used therapeutic sequence, the volume and the dose depend on the operative and anatomopathological accounts, in fact one finds in the literature a

heterogeneity of the results of the retrospective studies concerning the benefit of the radiotherapy and one can however retain a significantly lower local relapse rate for patients treated by surgery and postoperative radiotherapy, compared with that observed after surgery alone [10].

Additional chemotherapy

Adjuvant chemotherapy is not considered a standard in uterine sarcomas.

Gene therapy

Several molecules, such as celiciclib (CYC202; r-roscovitine), TNP-470 have been tested alone or in combination with other therapeutics, with disparate results. Promising results have been obtained with imatinibe mesylate, but further research is needed to confirm them [11].

Hormone therapy

Several hormones, such as anti-estrogens, progesterone, antiprogeterones and GNRH analogues, have been proposed in various publications without consensus being possible since they all relate to isolated cases or small series [12].

INDICATIONS

For uterine sarcomas operable from the outset, surgical excision followed by complementary radiotherapy represents the current standard for locoregional treatment. Lymphadenectomy performed; when there is an associated carcinomatous component, it concerns the latero-pelvic relays and possibly the lumbar-aortic region when they are invaded or in front of the presence of pelvic lymphadenopathy visible on imaging and / or palpable intraoperatively. For non-operable cases, there is no standard attitude. However, palliative radiotherapy may be offered to patients with active not operable tumors. When there is a recurrence of sarcoma in the metastatic form pulmonary or hepatic accessible to a surgical resection, surgery of excision (hepatectomy or pulmonary resection) must be discussed.

RESULTS

Prognostic factors

Clinical factors: Age and menopausal status seem to play an important role in the evolution of this tumoral pathology.

Histological factors: the histological type which seems to be a real prognostic factor in comparison with the grade and degree of cell differentiation.

PROGNOSTIC

The prognosis of uterine sarcomas is pejorative since 5-year survival is of the order of 30% in all stages (14). Only the subgroup of low grade endometrial stroma sarcomas has prolonged survival.

CONCLUSION

The clinical and imaging of uterine sarcomas have little contribution to diagnosis. The diagnosis of these tumors is most often post-hoc on a surgical piece. The postoperative external radiotherapy surgery association remains a gold standard. New therapies are being evaluated in hopes of improving the prognosis of patients.

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