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HIGH-GRADE ENDOMETRIAL STROMAL SARCOMA :A CASE REPORT

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ABSTRACT Uterine sarcomas are tumors of poor prognosis characterized by a great histopathological heterogeneity. High endometrial stromal sarcomas (ESS) occurring at a late age, with high mitotic activity and / or tumor necrosis, are very poorly prognostic tumors.

If it is accepted that the standard treatment of uterine sarcomas is surgical, the place of adjuvant treatment remains controversial.

We report a case of a high-grade ESS referred to initially on MRI and diagnosed on pathological examination of a hysterectomy performed in a 64-year-old postmenopausal woman with postmenopausal bleeding.

KEYWORDS : High-grade, Endometrial Stromal Sarcoma, Surgery

INTRODUCTION:

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. It is a group of tumors, comprising different histological subtypes with tumors that may be of pure conjunctive origin such as leiomyosarcoma (LMS) (60%); the other subtypes are endometrial stromal sarcomas (ESS) and undifferentiated sarcomas (US). High-grade ESS occurring at late age, with high mitotic activity and / or tumor necrosis, are very poorly prognostic tumors.

OBSERVATION :

It is a patient aged 64 years, having 6 vaginal childbirths, of low socio-economic level, cholecystectomized, without a family history neoplastic, menopausal 8 years ago, took the oral contraception during 2 years in total, without notion of taking hormonal treatment substitute for menopause. She was admitted to our hospital in November 2016 for postmenopausal bleeding.

The clinical examination found a patient in poor general condition, discolored conjunctiva, abdominal palpation found a painless firm pelvic mass and on gynecological examination the cervix was dilated with a tumoral process delivered by the cervix, the vaginal touch was found the uterus increased in size with a mass of firm firmness of the uterus measuring 26 cm in height and 20 cm of width in the belly of douglas pouch. The rest of the general examination was peculiar.

The hemogram revealed hypochromic microcytic anemia at 5.6 g/ dl. The rest of the preoperative checkup was without abnormalities. The patient received 3 red blood cells preoperatively.

Pelvic Magnetic resonance imaging (MRI) showed a large lesion process measuring 20 × 14 × 11 cm, occupying the entire uterine cavity to the isthmic region, with a heterogeneous T1 and T2 signal containing tissue, hemorrhagic and necrotic zones infiltrating the myometrium of more than 50%, respecting the peripheral serosa without exceeding it, raising heterogeneously after injection of gadollinium, making the appearance of a uterine sarcoma infiltrating more than 50% of the myometrium with presence of bilateral external iliac adenopathies.

At surgical exploration the uterus was enlarged in volume measuring 30 × 18 × 12 cm seat of a intracavitary process of 17 × 14 cm. The 2 annexes, right and left, appear healthy. A total hysterectomy with bilateral oophorectomy was performed. The postoperative course was simple.

The histological study was in favor of an undifferentiated high-grade endometrial stroma sarcoma of diffuse fusocellular architecture, developing at the expense of the uterine wall. This tumor is made of atypical cells, elongated with nuclei hyperchromatic nuclei showing many mitotic figures. The mitotic activity is estimated at about 18 mitoses / 10champs with rare foci of necrosis.

The immunohistochemical study showed the expression of anti-CD10 +, the antibodies AML, H-caldesmone and the hormonal receptors are negative, the Ki67 = 20%. The cervix, annexes and parameters are healthy. Peritoneal cytology was haematic without cells suspected of malignancy.

DISCUSSION:

-Introduction :

Endometrial stromal sarcoma represents a rare subtype of uterine mesenchymal neoplasm and less than 10% of uterine sarcomas

-Classification :

The majority of uterine sarcomas are leiomyosarcomas (60%), with a myxoid form and an epithelioid variety, often voluminous tumors, whose survival is 25 to 40% at 5 years irrespective of stage.

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- ESS and undifferentiated sarcomas are rare. ESS is a better prognosis than other uterine sarcomas with a 5-year survival of 57-70%.
- Low-grade endometrial stromal sarcomas are distinguished in younger women with often good-prognosis hormone receptor expression in contrast of undifferentiated endometrial stromal sarcomas occurring at a later age, with high mitotic activity and / or tumor necrosis.

US are tumors of very poor prognosis. The pathological definitions were reviewed: ESSs were previously low grade sarcomas and undifferentiated sarcomas were undifferentiated high grade sarcomas.

In 2014, WHO reclassified the endometrial stromal tumors (ESTs) on the basis of immunohistochemistry and molecular findings into 4 subtypes: endometrial stromal nodule (ESN), low grade ESS (LGESS), high grade ESS (HGESS) and undifferentiated uterine sarcoma (UUS) [1]. This was based on the finding that at molecular level both LGESS and HGESS exhibit relatively simple karyotype whereas defined chromosomal rearrangement is lacking in UUS.

-The diagnosis :

Diagnosis is based on morphological, immunohistochemical and even molecular biology criteria.

In the event that molecular biology tests are necessary, it is important that the samples are sent quickly for freezing within 15 to 30 minutes.

ESS is a tumor that occur in middle-aged women (mean age 45 years) with genital hemorrhage. ESSs represent 10 to 15% of uterine sarcomas, in peri and postmenopausal women (42 to 58 years, ¾ <50 years), with possible association with hormone therapy, such as tamoxifen.

the clinical feature is Vaginal haemorrhage, sometimes pelvic or abdominal pain, the tumor can protrude to the cervical os, the ectopic extension is in 1/3 of cases at the time of diagnosis.

There is a well-defined benign or stromal nodule form and an infiltrating malignant form in which there is a low grade with little mitosis or endolymphatic stromal malformation and a high grade form or stromal sarcoma.

Macroscopically, all these stromal tumors have a soft yellow-orange appearance, they can protrude through the cervical os, the diagnosis is mainly on curettage.

Histologically, it is a monomorphic proliferation of small cells surrounded by reticulin fibers, condensing around small vessels, small foci of hyalinization, scattered foam cells and pseudo epithelial clusters. The marked vascularity typical of these tumors can lead to the erroneous diagnosis of hemangiopericytoma especially in metastatic locations.

These cells are RE + RP + and respond to progestins.

In tumors of endometrial stroma: there is the role of chromosomal rearrangements including JAZF1 / JJAZ1 following t (7; 17) (p15; q21)[2]

Stromal sarcoma of high grade or undifferentiated form [3] (WHO):

It has the same intravascular extension as the low-grade form of which it is differentiated by a high mitotic activity> 10 mitoses / 1.7 mm^2 (although this criterion is not absolute, high-grade forms have few mitoses and some low grades are proliferative), atypia and high cellularity[4].

Macroscopically: there is diffuse growth occupying the entire endometrial surface with a soft, polypoid mass with frequent rearrangements.

Histologically: there is a more pronounced polymorphism with fusiform or polygonal cells, nuclear and mitotic atypies, high cell density, aggressive invasion of the myometrium, necrosis, haemorrhage, irregular pleomorphic vascularization.

Genetically: t (7; 17) (p15; q21) is common in the stromal nodule and low grade ESS, rarer in the high grade ESS.

Endometrial stromal sarcoma is heterogeneous, with one half associated with rearrangement of JAZF1, possibility of further rearrangement t (10; 17) (q22; p13) with YWHAE-FAM22A / B fusion transcript, comparison with rearrangement cases of JAZF1. The YWHAE - FAM22A / B + cases are high grade with small nested cells, high number of mitoses, large irregularly shaped nuclei, focal necrosis. The less aggressive component is common to fusiform cells with fibrous stroma / fibromyxoid, which can be found in metastases, which RE / RP +, CD10 + in contrast to round cells.

JAZF1 cases do not have nests, less mitoses, small nuclei. The stadium is less advanced.

-The prognosis:

The prognosis is poor with pelvic recurrence and systemic metastases especially in the lung, death is common within 2 years, survival at 5 years ranging from 48 to 10% [5].

Size and extra-uterine extension are important prognostic factors, tumors smaller than 4 cm and those limited to the uterus rarely recur (at stage I the prognosis is similar to the low grade form). Ploidy seems to have a prognostic impact.

Some high-grade sarcomas exhibit marked polymorphism without clear vascularization and correspond to poorly differentiated sarcomas of aggressive behavior. The answer is common to progestins [6].

-Treatment :

Surgery including hysterectomy and bilateral salpingo-oopherectomy (BSO) is considered as the initial standard management [7]. The prognostic significance of lymph node metastasis and role of lymphadenectomy is debatable [8].

The optimal adjuvant treatment is still unclear [9]. Weitmann et al. proposed the use of adjuvant RT as most effective treatment, however randomized trial failed to show benefit of adjuvant RT in stage I or II sarcoma [10]. In ESS, there is very little data to support the use of chemotherapy. Our data also revealed the need for adjuvant treatment especially in high grade and advanced stage ESS. However, due to small number of patients conclusion regarding the optimal adjuvant treatment could not be drawn [10].

CONCLUSION:

Even if role of optimal adjuvant treatment with chemotherapy, radiotherapy and hormone therapy is yet undefined, Complete staging surgery is associated with improved Disease Free Survival. Resection of recurrent disease is associated with survival advantage.

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