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Ovarian Adult Granulosa Cell Tumor: Report of 3 Cases

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Abstract

Granulosa tumors are rare malignant tumors of the ovary. There are two histological types: the adult form (95%) and the juvenile form (5%). They often occur in the adult population with extreme ages ranging from 40 to 70 years. The clinical signs are very variable with a slow evolution and a risk of recurrence that can go up to 30-40 years after the diagnosis. The radiological diagnosis is based on ultrasound data and magnetic resonance image (MRI). Radical surgical treatment is the basis of their therapeutic management.

Keywords: Granulosa tumors; Ovary; Diagnosis

Introduction

Granulosa tumors (TGO) are a rare entity of malignant ovarian tumors (5%). They are hormone-secreting and give rise to granulomatous and thecal cells of the ovary, thus forming part of the group of tumors of the sex cords and the stroma of the ovary [1]. The adult form is the most common tracing a very typical clinical pathological profile with a slow progression and recurrence that occurs long after the date of onset of the disease. The management is essentially surgical when the diagnosis is early [1].

Case Reports

Case report 1

Mrs. M.R, 57-year-old married and mother of 6 living children delivered vaginally, menopaused for 9 years and who has for 2 years postmenopausal metrorrhagia of average abundance associated with chronic pelvic pain. The clinical examination found the presence of bleeding from the endocervix with an enlarged uterus at two across two fingers above the pubic symphysis and a right lateral uterine mass with a separating furrow with the uterus. Pelvic ultrasound showed a heterogeneous and vascularized right lateral posterior uterine image of 8 cm with a cystic solido-cystic double doppler (Figure 1) with heterogeneous 51 mm endometrial thickening. Hysterosonography: 2 intracavitary polyps at the posterior wall of 21 and 13 mm.

Pelvic MRI: A right ovarian mass of irregular contours with a fleshy and cystic double component, bounded by a hyposignal T2 border whose hinge component is in T1 isosignal, discrete T2 hypersignal exhibiting a restriction of diffusion, is weakly enhanced heterogeneously after injection of the 77/46 mm contrast medium. Irregular endometrial thickening. CA125: 17 U/ML (Figure 2).

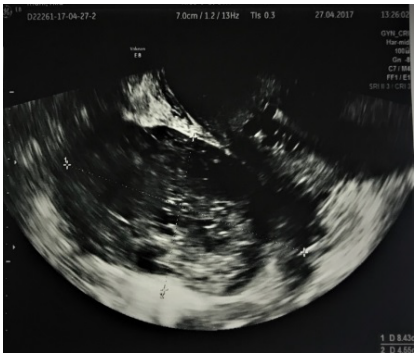

Figure 1: Ultrasound image of the ovarian tumor with heterogeneous ecogeneity.

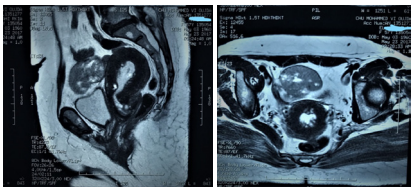

Figure 2: Images of MRI showing tumor process and endometrial thickening.

The patient initially benefited from an endometrial biopsy by surgical hysteroscopy whose anatomopathological result is in favor of a simple hyperplastic endometrial polyp without atypia. Then, in the second stage of a straight anexectomy, the anatomopathological result returned to a tumor of the adult granulosa of the solid type, with capsular rupture with right horn and peritoneal cytology free from any tumor infiltration. The patient received a surgical supplement: total hysterectomy with left adnexectomy and biopsy of the left and right parietocolic aligners and omentectomy. Of which the anatomopathological result showed secondary locations in the cervix, uterine serosa and epiploon of an adult granulosa tumor. Patient currently benefits from her chemotherapy treatments

Case report 2

Mrs N.F 81-year-old married with no children, menopausal since 25 years, diabetic under ADO hypertensive under medical treatment. Who has for 3 months a progressive increase in abdominal volume with chronic constipation and urinary incontinence. The abdominal examination found a 20/15 cm abdominal mass fixed deep down through the fingers below the xiphoidal appendix, and the gynecological examination found filling of the vaginal sacs with the mass. Pelvic ultrasound revealed a compartmentalized cystic mass with endocystic vegetations of 22/15 cm (Figure 3), the uterus not visible. Pelvic MRI showed a suspected ovarian tumoral process (Figure 4) with compression of both ureters and uretero hydro-nephrosis bilaterally. Biologically, the patient had functional renal failure and a CA 125: 9 U/ml.

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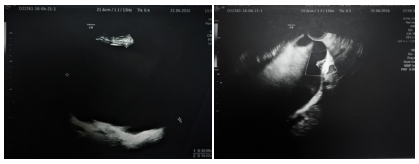

Figure 3: Two ultrasound images showing a voluminous image of cystic tone with an endocystic vascularized Doppler bud.

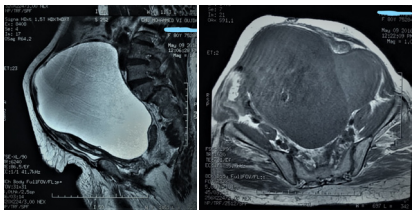

Figure 4: two pelvic MRI images showing suspicious abdominal pelvic tumor.

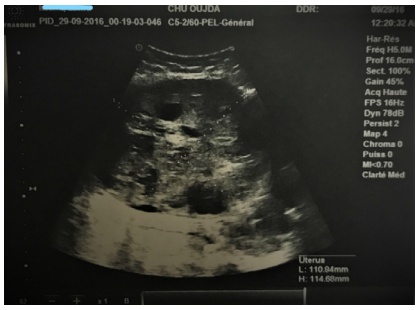

Figure 5: Ultrasound image showing ovarian tumor with heterogeneous and dual component.

The patient had a left nephrostomy and an exploratory Laparotomy showing a cystic mass adhering to the intestinal loops and the sigmoid whose origin was not identified. The patient had a cystectomy with anatomopathological findings diagnosing an adult granulosa tumor and negative peritoneal cytology. The patient was a candidate for adjuvant chemotherapy, but given her advanced age and the comorbidity field, clinical and radiological surveillance was initiated with good progression without signs of recurrence or distant metastasis for a year.

Case report 3

Mrs. K.Z 52-year-old mother of 2 living and perimenopausal children who presents chronic pelvic pain type gravity with an increase in the abdominal volume evolving since 6 months. At the Clinical Examination: presence of a pelvic abdominal mass arriving at the umbilicus. Vaginal touch: left lateral uterine mass with separation furrow with uterus. Pelvic ultrasonography demonstrated a left ventricular posterior uterine image of 11 cm with heterogeneous tissue Doppler suggestive of ovarian tumor or sub serous fibroid (Figure 5). The CA 125 was negative.

Pelvic MRI showed a suspicious suprarum and uterine right uterine mass of well-defined ovarian origin with regular heterogeneous contours in T1 and T2 containing multiple cystic zones and delimited by a hypo-signal capsule with restriction of diffusion, it is enhanced by intense and heterogeneous manner with injection of Gado, measuring 15 cm long axis. The patient had left adnexectomy, peritoneal cytology; right ovary biopsy and epiploic biopsy. Pathological examination returned to a left ovarian tumoral process evoking a tumor of the sex cords. The immunohistochemical study diagnosed an adult granulosa tumor. A radical surgical complement has been realized. The suites were simple, currently under clinical supervision.

Discussion

Tumors of the ovarian granulosa (OGT) are rare, their incidence is 1.3 cases/100,000/year variable, with an average of 53 years for the adult form [1,2]. Clinical symptomatology is not specific for these tumors, but is most often manifested by an increase in abdominal volume with diffuse abdominal pelvic pain sometimes associated with cycle disorders or postmenopausal metrorrhagia. Their hyper-estrogenic character explains the appearance of endocrine manifestations and their association with other estrogen-dependent pathologies such as endometrial hyperplasia with or without atypia [2], as shown in our first clinical case.

Ultrasonography shows a large cystic or mixed tumor partitioned with intra tumor necrotic sites giving it its often unilateral heterogeneous character usually without calcifications with sometimes ascites. On pelvic MRI, these masses are hypersignal in T1 testifying to the presence of haemorrhagic changes, and in intermediate signal in T2 with alternating solid and cystic spaces responsible for their spongy appearance. Metastases are generally less frequent with the following sites of preference: the peritoneum and the liver [2,3]

The cornerstone of GTO treatment is total hysterectomy with bilateral adnexectomy with staging of the pathology consisting of omentectomy and peritoneal exploration with cytology and multiple peritoneal biopsies. Conservative treatment, based on unilateral adnexectomy with abdominal cavity exploration and endometrial biopsy, is proposed in young women with a desire for pregnancy as well as early stages. Ganglion dissection is not recommended as the risk of lymphatic invasion is infrequent [2,4].

Macroscopically they are bulky tumors up to 30 cm long axis of solido-cystic appearance necrotic in places, usually unilateral. The histological diagnosis for the adult form is based on the presence of "Call exner" bodies and coffee bean cores, with a histochemical immune profile characterized by the extracellular expression of a number of markers such as vimentin, CD99, smooth actin and inhibin [2].

Radiotherapy has not been effective until recently. Chemotherapy will have a place in advanced metastatic stages or in case of recurrence. In the literature hormone therapy is needed more and more to slow the evolution of the disease. Many studies discuss the benefits of anti-aromatases, Luteinising Hormone Releasing Hormone (LHRH) analogues, and progestins. [4]

The evolution of this pathology depends on the date of diagnosis and the treatment instituted, it is frequently favorable with long periods of remission interspersed with recurrences in the medium and long term. Overall survival and no recurrence, in the literature, is 84% to 94% at 10 years for the early stage. The prognosis varies according to the patient's age of tumor size, distant metastases, the histochemical immunity profile, the number of mitoses and the presence of a mutation in the *P53* gene. The serum tumor markers have a place in the prognosis and the therapeutic follow-up, the most frequently dosed are the inhibin which is negative after the surgical excision of the mass and whose increase testifies to the recurrence. And AMH: anti-Mullerian hormone that represents a marker of therapeutic efficacy [2,3].

Conclusion

Granulosa tumors of the ovary are malignant and rare. Early

diagnosis allows conservative management with less metastatic risk. The follow-up of this pathology must be prolonged given the risk of recurrence in the long term [1].

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