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Ovarian Steroid Cell Tumor About a Case

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Abstract **Case Report**

Tumors of the sex cords and stroma of the ovary are relatively rare, accounting for around 8% of all primary ovarian tumors. Steroid cell tumors of the ovary are rare, accounting for less than 0.1% of all ovarian tumors. The majority of these tumors produce steroid hormones, and only 10-15% of patients are asymptomatic. Although steroid cell tumors are generally benign, there is a risk of malignant transformation. Surgery is the main treatment. We report a case of a steroid cell tumour of the left ovary discovered on anatomopathological examination of the left adnexectomy operative specimen in a 47-year-old hypertensive diabetic patient who clinically presented with virilism with severe hirsutism, muscular and clitoral hypertrophy and frontotemporal baldness, and biologically increased androgens without associated hypercorticism, and in whom an abdomino-pelvic scanner showed a left latero-uterine mass measuring 74x66x64 mm, pelvic MRI showed a polylobed left ovarian mass classified ORADS 5, measuring 53x65x87 mm. After histological confirmation of the diagnosis, the patient was taken back for total surgery with total hysterectomy, right adnexectomy, omentectomy, appendectomy, multiple peritoneal biopsies, with no signs of malignancy on histological examination, simple postoperative follow-up and our patient placed under surveillance.

Keywords: Sex cord and stromal tumors of the ovary; steroid cell tumors of the ovary; androgens; adnexectomy; anatomopathology.

INTRODUCTION

Ovarian tumors are subdivided into three main categories: epithelial tumors, germinal tumors, tumors of the gonadal stroma and sex cords, and other rare tumors.

Tumors of the sexual cords and stroma of the ovary are relatively rare, accounting for around 8% of all primary ovarian tumors. Their precise diagnosis is essential, as it determines the therapeutic approach.

Ovarian steroid cell tumors are part of the tumors of the sex cords and stroma of the ovary. They are ovarian neoplasms composed of steroid hormone-secreting cells and usually occur in adults. They are very rare, have steroid-secreting capacity and are most often virilizing. Only histopathological examination, with or without immunohistochemical study, can provide an accurate diagnosis. These tumors are generally benign, but the potential for malignancy is not negligible, and treatment is usually surgical. Once malignancy has been confirmed, complementary therapies are required.

CASE REPORT

47-year-old patient, gravida 4, para 4, type 2 diabetic on Glucophage, hypertensive on triple therapy, history of thyroidectomy for complicated goiter with tracheotomy, hospitalized in the endocrinology department for a clinical and biological hyperandrogenism syndrome, presenting with virilism associated with severe hirsutism, muscular and clitoral hypertrophy, and frontotemporal baldness. Biological workup shows increased androgens without associated hypercorticism: testosteronemia=12.75 ng/ml (14x normal), dehydroepiandrosterone sulfate (SDHEAS)=850 ng/ml, estradiol=173 pg/ml. Abdomino-pelvic scanner showed a heterogeneous, roughly oval, left latero-uterine mass measuring 74x66x64 mm, pelvic and latero-aortic adenopathies, no signs of peritoneal carcinosis. Pelvic MRI showed a roughly oval, polylobed left ovarian mass classified as ORADS 5, measuring 53x65x87 mm. Chest X-ray showed multiple nodular peri-hilar opacities. Chest scanner showed no suspicious-looking parenchymal

lesions, basal subpleural curvilinear condensations that were sequelae of COVID 19 infection.

An exploratory laparotomy was indicated. On exploration, a tumour measuring 8x7 cm was found over the left ovary; the right ovary was unremarkable. A major left adnexectomy of the mass was performed, the postoperative course was straightforward, and the operative specimen was sent for anatomopathological study, which showed a steroid cell tumour of the ovarian stroma associated with a stromal tumour contingent with kitten-ring cells, the ovarian capsule intact, and the tumour classified as stage IA.

The decision of the multidisciplinary coordination meeting (MCM) was to measure tumour markers: CA125=21,1; CA19-9= 9,2; ACE=1,77. After reassessment, the MCM decided to perform total cytoreduction surgery in addition to a left adnexectomy. By subumbilical median laparotomy, a total hysterectomy was performed, with right adnexectomy, omentectomy, appendectomy and multiple peritoneal biopsies. No lymph node dissection was performed. Surgical specimens were sent for anatomopathological study, which showed the absence of histological signs of malignancy on all specimens sent. Post-operative care was straightforward, and the patient was given a follow-up appointment.

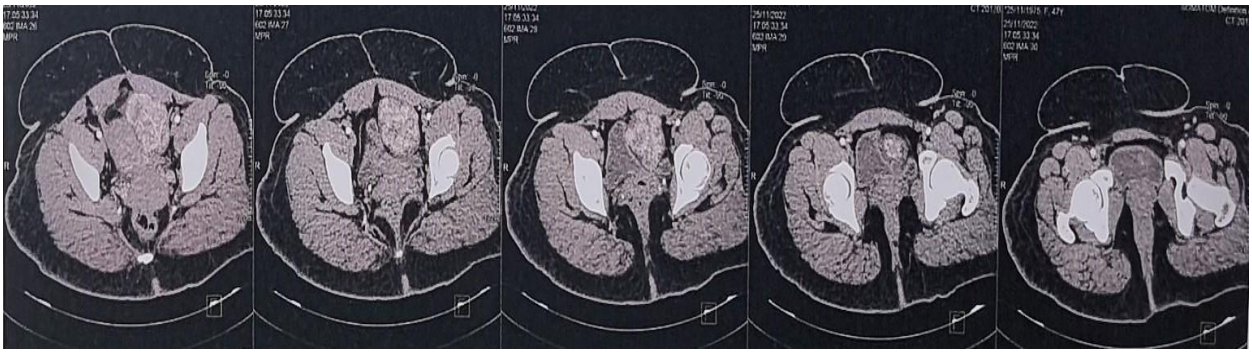

Figure 1: Abdominal and pelvic scanner showed a roughly oval, heterogeneous left latero-uterine mass measuring 74x66x64 mm

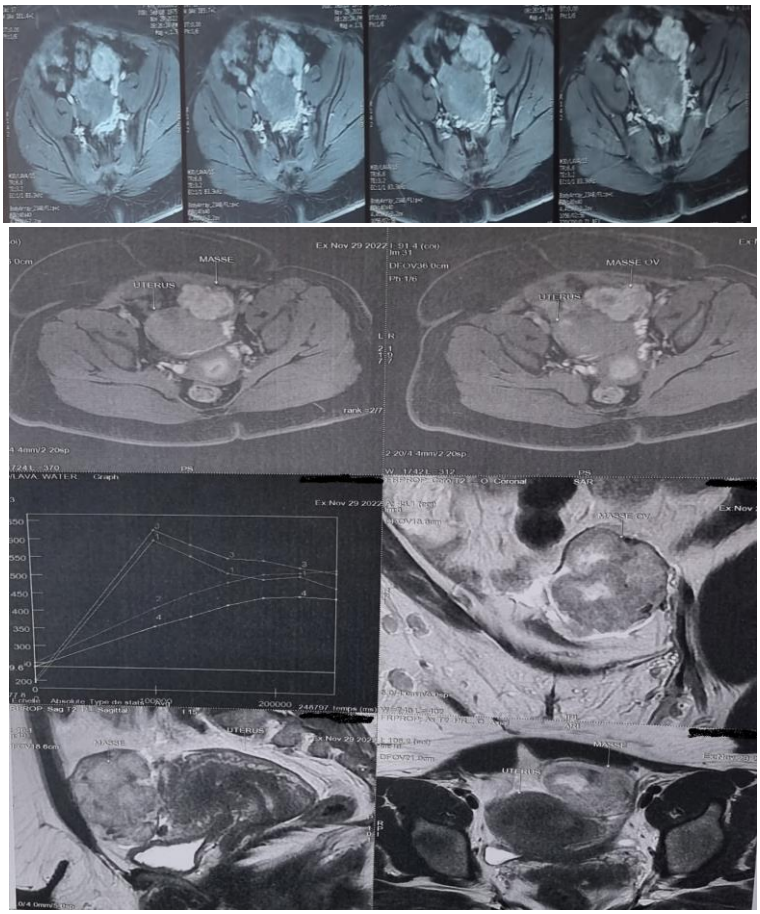

Figure 2: Pelvic MRI showing a roughly oval, polylobed left ovarian mass classified ORADS 5, measuring 53x65x87 mm

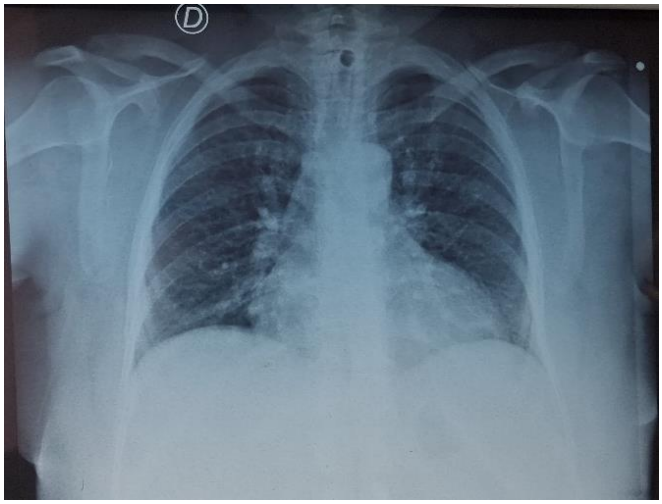

Figure 3: Chest x-ray showing multiple nodular peri-hilar opacities. No suspicious-looking parenchymal lesions on chest scanner

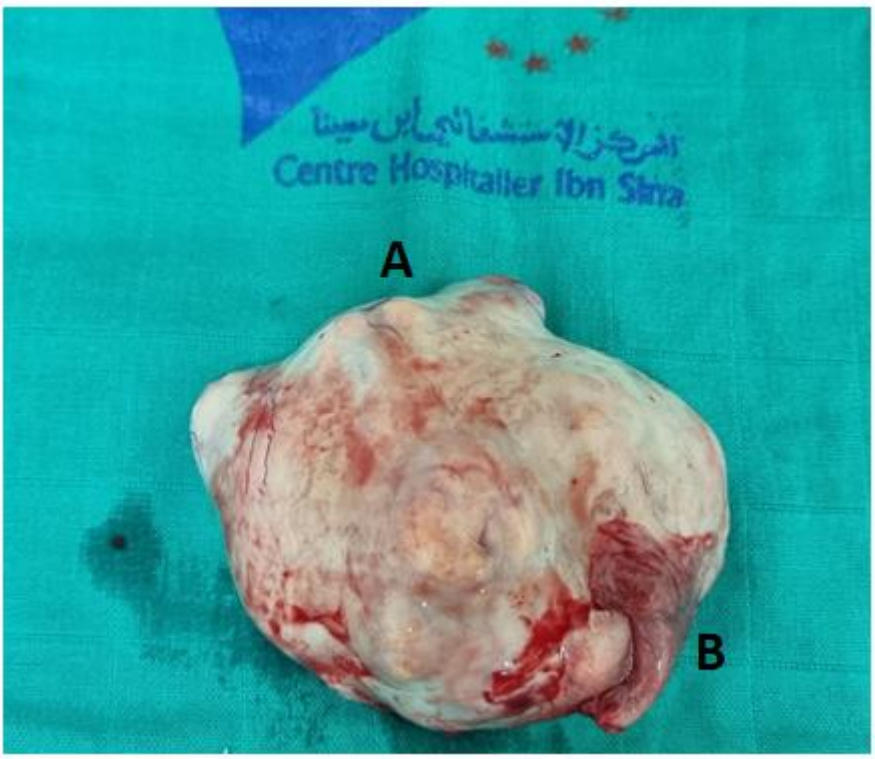

Figure 4: Left adnexectomy specimen sent for anatomopathological study: 8x7 cm polylobed mass over the left ovary (A), left fallopian tube (B)

DISCUSSION

Ovarian tumors fall into three main categories: epithelial tumors (55%), germ cell tumors (30%), tumors of the gonadal stroma and sex cords (8%), and other rarer tumors. Ovarian cancer accounts for 3,7% of all female cancers (6th rank) [1].

Tumors of the sex cords and stroma of the ovary are relatively rare, accounting for around 8% of all primary ovarian tumors and 7% of all ovarian cancer tumors. Most are low-grade. The majority of stromal tumors are diagnosed in women over 50, but they can also occur in adolescents and young women. They may be estrogenic or androgenic. Some are characterized by an element of female differentiation (granulosa and fibrothecal group tumors) or male differentiation (Sertoli or Sertoli-Leydig cell tumors). Some are made up of steroid cells: stromal luteoma, Leydig cell tumor, hilar tumor [2]. These tumors develop at the expense of cells belonging to the supporting tissue of the ovaries, and cover a wide range of heterogeneous lesions characterized by their great morphological diversity. These cells produce sex hormones such as estrogen, progesterone and androgens. Stromal tumors often over-secrete these hormones. Accurate diagnosis of these tumors is fundamental, as it determines the therapeutic

approach. The development of medical imaging techniques in recent years has facilitated the diagnosis of ovarian tumours. However, only histopathological examination, with or without immunohistochemical study, can provide an accurate diagnosis. Because of their rarity and morphological diversity, these tumors pose a number of diagnostic, therapeutic and prognostic challenges.

Steroid cell tumors include stromal luteoma, Leydig cell tumors and unspecified steroid cell tumors. Only the latter have malignant potential, so metastatic [3]. Clinically, they may be accompanied by signs of virilization or manifestations of hyperoestrogenism. Stage, age, tumour size, presence of necrosis, nuclear atypia and number of mitoses have been reported to have an impact on patient survival [4].

Non-specific steroid cell ovarian tumours account for 60% of all steroid cell ovarian tumours and less than 0.1% of all ovarian tumours [5].

Steroid cell tumours occur at any age, with an average age at diagnosis of 47. In 50% of cases, they are virilizing tumors. They may be non-secreting (25%), cause hyperoestrogenism (10%) [5] or, more rarely, Cushing's syndrome. They are malignant in 25 to 43% of cases [5,6]. They are made up of cells similar to those that secrete steroid hormones. Stromal luteoma and Leydig cell tumors are clinically benign. Approximately one-third of steroid cell tumors without other indications are clinically malignant.

In terms of imaging, the usually small size of these tumours means that they may not be detected by ultrasound or CT scan [7]. Color Doppler, pelvic angioscanner or dynamic MRI appear useful for unmasking the major hypervascularization of these tumors [8].

Steroid cell tumors account for less than 0.1% of ovarian tumors [9]. They are generally benign, although the potential for malignancy is non-negligible in the presence of well-defined histological criteria. Good ovarian imaging is essential in the assessment of hyperandrogenism without associated adrenal abnormalities. Treatment is mainly surgical. Complementary therapies are required once malignancy has been confirmed.

Given that 70% of patients have stage I disease (overall 5-year survival 95%), surgery is the most important therapeutic weapon. Its role is essential, but no precise recommendation can be given as to its modalities : removal of all lesions present remains the basis of treatment. Finally, the prolonged natural history of the disease argues in favor of iterative surgery in the event of recurrence.

Published studies provide no decisive evidence in favor of treatment with radiotherapy, the volume to be irradiated is not defined (it is not obvious that total abdominal radiotherapy will be necessary), and in the publications the doses used, when reported, are highly variable.

Chemotherapy is proposed for tumors with a poor prognosis and in relapse, although no scientific data have been published [4]. Chemosensitivity is evidenced by the numerous responses observed in palliative situations: brief response to alkylating agents, frequent response to adriamycin-bleomycin, actinomycin-fluorouracil-cyclophosphamide and cisplatin-based combinations ; the highest response rate is 80% with the cisplatin- etoposide-bleomycin (BEP) combination, protocol identical to that used for testicular tumours.

In our case, the tumour was confined to the left ovary with stage IA, and left adnexectomy was performed. After confirming the diagnosis of ovarian tumour on anatomopathology, with the presence of a contingent of stromal tumour with kitten-ring cells, we decided to carry out total surgery with total hysterectomy, right adnexectomy, omentectomy, appendectomy, multiple peritoneal biopsies, with the anatomopathological result of the absence of histological signs of malignancy on all surgical specimens studied.

CONCLUSION

Ovarian steroid cell tumors are very rare tumors that belong to the tumors of the sex cords and ovarian stroma, they are seen in adult women and are most often virilizing as in our case described, they are generally benign but the risk of malignant transformation is possible, the diagnosis is confirmed by histopathology, treatment is most often surgical.

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