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Postmenopausal Ovarian Sertoli-Leydig Tumours: Case Report

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ABSTRACT

Sertoli and Leydig cell tumours are rare secretory tumours of the mesenchyme and sex cords. Diagnostic certainty is histological after surgery and there is no specific ultrasound sign despite the strong clinical presumption. The prognosis depends on the degree of cellular differentiation and the presence of heterologous elements. We report on the occurrence of a sertoli-leydig tumour in a 50-year-old postmenopausal woman. Poorly differentiated forms of Sertoli-Leydig tumours have a significant malignant potential. Treatment is surgical, with chemotherapy providing an interesting adjuvant. The prognosis after surgery is dominated by recurrence.

Keywords

Tumour, Sertoli-leydig, Ovary, Menopausal.

Introduction

Sertoli-Leydig tumours are defined by the WHO as tumours formed by Sertoli cells and Leydig cells in variable proportions, more or less associated with a primitive stroma and heterologous elements [1]. They are very rare, accounting for less than 0.2% of all ovarian tumours [2]. They belong to the group of mesenchymal and sex cord tumours. They frequently occur in young women, between the ages of 23 and 25, although it is not uncommon to see them in patients over the age of 45, and even 10% of these tumours occur after the menopause [2-4]. They are the main female secretory tumour responsible for virilisation. Diagnostic certainty is histological after surgery, and there is no specific ultrasound sign despite the strong clinical presumption. The prognosis depends on the degree of cellular differentiation and the presence of heterologous elements.

Patient and Observation

Mrs A.S., aged 50, menopausal for 4 years, consulted us with abdominal distension associated with a deterioration in general condition, which had been evolving for 2 months. Clinical examination revealed a distended abdomen with palpation of a large mass extending above the umbilicus. Pelvic MRI revealed a large median abdominopelvic mass measuring 18 x 12 cm, with a solid-cystic encapsulated appearance and a predominantly mutiloculated fluid component, with thick, irregular partitions enhanced by contrast medium, and a solid component enhanced by contrast medium (Figure 1). There is a moderate amount of peritoneal effusion. The CA 125 assay was 574.04U/ml. Exploratory laparotomy revealed a 17 cm left ovarian mass associated with a moderate amount of effusion. An adnexectomy was performed, with extemporaneous examination revealing a poorly differentiated invasive carcinoma, leading to total hysterectomy without adnexal preservation, with bilateral pelvic curage, omentectomy and peritoneal biopsy. On definitive anatomopathological examination: Histological image of the tumour at magnification 10 and 20 shows tumour proliferation in a solid mass of microcysts and tubules within an oedematous stroma. The tumour cells are basophytic with sparse cytoplasm and moderately atypical hyperchromatic nuclei (figure 3); The tumour cells express calretinin, wt1 and cytokeratin AE1/AE3 (Figure 3), giving a morphological appearance consistent with a Sertoli-leydig tumour of intermediate differentiation, located in the left ovary. The patient's follow-up was marked by a worsening of her general condition, with anorexia and weight loss. The patient died at 23 days post-op.

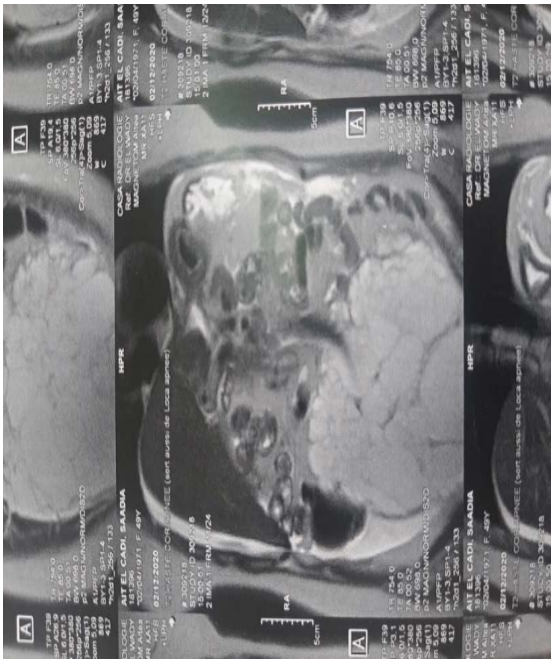

Figure 1: Abdominal-pelvic solid-cystic mass.

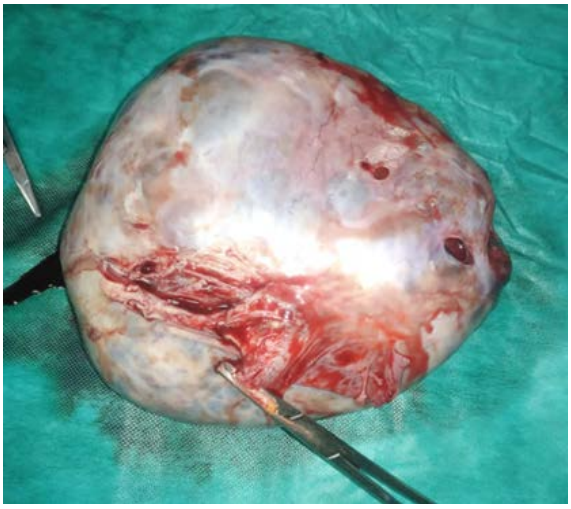

Figure 2: Left adnexectomy specimen.

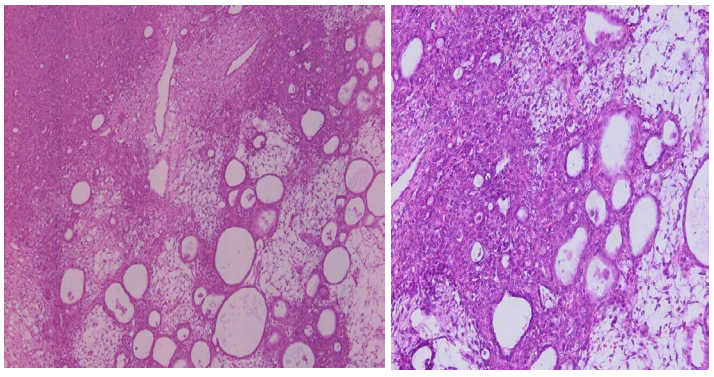

Figure 3: Histological image of the tumour at magnification 10 and 20 showing tumour proliferation in the form of solid masses, microcysts and tubules within an oedematous stroma.

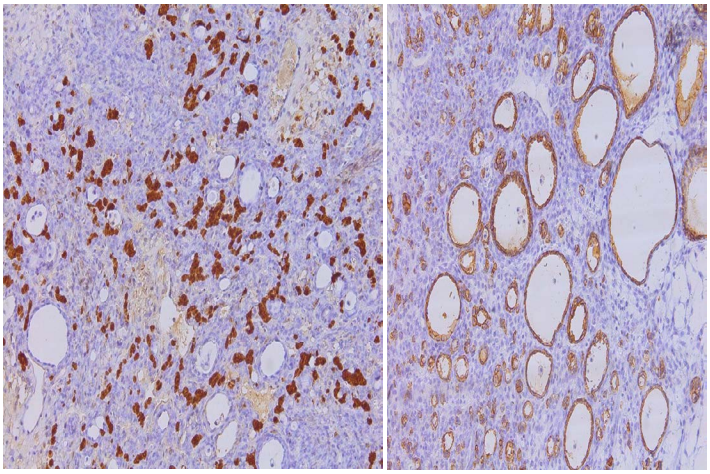

Figure 4: Tumour cells express calretinin, wt1 and cytokeratin AE1/AE3.

Discussion

Sertoli and Leydig cell tumours are rare, accounting for 0.2% of all ovarian cancers [2]. These tumours are derived from the mesenchyme and sex cords and include all phases of embryonic development of the testis, from the diffuse stromal and undifferentiated cord aspect to the well-differentiated sertoli tube [1]. Depending on the variable proportions of sertoli and leydigial elements, these tumours are classified into 3 groups: benign, well-differentiated forms, 60% of which are secretory, intermediately differentiated forms (immature sertoli cells) and poorly differentiated, sarcomatoid or retiniform forms [1]. These tumours are almost exclusively unilateral and confined to the ovary, with approximately 10% of cases presenting with ovarian rupture and 4% with ascites [5]. Familial forms of these types of tumour have been described [6]. Clinically, the average age of onset is around 25 years [2,5], but cases in postmenopausal women are not uncommon, as in our patient. Symptoms are not specific. They may include a tumour syndrome, pelvic pain or menstrual disorders such as secondary amenorrhoea. Signs of virilisation such as hirsutism, hoarseness of the voice, clitoral hypertrophy, defeminisation of the silhouette and changes in psycho-sexual behaviour can be found in 30 to 50% of cases. They vary greatly in size, up to 35 cm, with an average size of 12-14 cm [7].

On ultrasound, Sertoli-Leydig cell tumours appear as heterogeneous vascularised tissue masses with solid areas; in pure Sertoli cell forms, they are frequently multiloculated, associated with anechoic fluid areas [8]. Macroscopically, SLI are usually purely solid, with a smooth outer appearance. On cross-section, the solid areas have a yellowish appearance, with a soft, fleshy consistency. In large tumours, there is often an associated cystic contingent, in which case the external appearance remains smooth but lobulated. Cysts are multilocular with a clear fluid content. Their walls appear more rigid than in benign or borderline epithelial tumours; there may be areas of haemorrhage and necrotic tissue [9].

Histological examination confirms the diagnosis and enables the grade to be defined according to the varying proportions of

sertolitic and leydigric elements and their degree of differentiation. Well-differentiated tumours are those with pure Sertoli cells, which take on the appearance of the prepubertal testis, and Leydig cells which develop in the hilum, and mixed tumours, consisting of Sertoli cells separated by clusters of Leydig cells. Tumours with intermediate differentiation are made up of immature Sertoli tubes, associated with a stroma made up of Leydig cells, in a minority proportion. Poorly differentiated forms are made up of spindle-shaped cells with a pseudosarcomatous, retiniform appearance, reminiscent of the rete testis. Heterologous elements may be found in the latter two forms (bone tissue, cartilage, gastrointestinal epithelium, hepatocytes) [10].

The management of this type of tumour has not yet been codified, with each centre having its own practices [10]. Generally, in well-differentiated forms, considered to have a good prognosis, unilateral oophorectomy is recommended in order to preserve fertility. Moderately and poorly differentiated forms are often diagnosed at an early stage and can also be treated to preserve fertility; if the patient is post-menopausal or if the tumour is extensive, more radical treatment with bilateral adnexectomy and hysterectomy is indicated. The value of adjuvant treatment has not yet been clearly established due to the paucity of studies to date. It is often used in advanced forms, poorly differentiated forms or if there is a heterologous contingent [11].

The prognosis for well-differentiated forms is excellent, with a five-year survival rate of 100%; for moderately to poorly differentiated forms, five-year survival drops to 80%. When the tumour remains localised to the ovary, five-year survival is 95%, while for metastatic forms it is close to 0%. Recurrence and asynchronous metastases are rare, but have been described, with recurrence generally occurring 2 to 3 years after surgery [11].

Conclusion

Non-epithelial tumours of the ovary derived from the mesenchyme and sex cords are rare, those involving Sertoli-Leydig cells the most so. They are characterised by signs of virilisation of ovarian origin. Poorly differentiated forms have a significant malignant potential. Treatment is surgical, with chemotherapy an interesting adjuvant. The prognosis after surgery remains guarded, dominated by recurrence.

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