Sex Cord-Stromal Tumors of the Ovary: Clinical and Pathological Study of 13 Cases

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INTRODUCTION

Ovarian sex cord-stromal tumors are uncommon neoplasms that represent approximately 7% of all ovarian tumors. These tumors are subdivided as pure stromal tumors, pure sex cord tumors, or mixed sex cord-stromal tumors. They differ from the more frequent epithelial neoplasms via strong associations with hormone-mediated syndromes, presentation in a broad age range, and generally an early diagnosis with a good outcome.

OBJECTIVES

- ✓ Describe the clinical and imaging aspects to guide the diagnosis of ovarian sex cord tumors.
- ✓ Study the evolution and prognosis of these tumors after treatment.

METHODS

This is a descriptive study of all cases of malignant tumors of the Sertoli Leydig cell ovary diagnosed at the pathology laboratory of the University Hospital Farhat Hached in Sousse, Tunisia, since the creation of the center's cancer register over a 12-year period from 2004 to 2016.

RESULTS

- 13 cases were collected.
- Average age was 30 years [range 14 – 47]; 2 frequency peaks: ≤30 years (53.85%) and ≥50 years (6 cases or 46.15%)
- 7 patients were in genital activity; the others were menopausal.
- 2 women had a personal carcinological history: one was operated on a borderline tumor of the ovary and the other on a basal cell carcinoma of the face.
- Median consultation time was 6 months.
- Clinical presentation can be divided into 2 major signs:
 - endocrine signs present in 76.92% of cases (Table I),
 - and non-endocrine signs present in 53.84% of cases (Table II).

Table I: Endocrine signs in patients with sex cord tumors.

Endocrine Signs	Number
Hirsutism	3
Raucous voice	1
Clitoral hypertrophy	1
Abnormal bleeding	3
Irregularities of the menstrual cycle	3
Secondary Amenorrhea	1
Infertility	1

Table II: Non-Endocrine signs in patients with sex cord tumors.

Non-Endocrine Signs	Number
Abdominal and pelvic pain	6
Abdominopelvic mass	1
Increased abdominal volume	5
Alteration of the general condition	2
Fortuitous discovery	1

Pre-treatment Report:

1. Tumor markers:

- Cancer Antigen 125 (CA 125) was measured in 9 cases and was positive (>35 IU/ml) in only 3 patients.
- Alpha-feoto-protein (AFP) was measured in 7 patients and was negative in all cases (<10 ng/ml).
- The carcinoembryonic antigen (CEA) was measured only in 4 of our patients and was negative in all cases (<5ng /ml)

2. Hormonal study:

- Total testosterone was assayed only in 3 patients with signs of virilization; only one patient had a positive result (>0.90µg/l).
- Estradiol assay was also performed in 3 patients and was normal with respect to the cycle phase.
- Gonadotrophic chorionic β-hormone (β-HCG) was measured in 6 patients and all the results were also negative (<15 mIU/ml).

3. Pre-therapeutic imaging:

- Pelvic ultrasound was performed in all patients (Table III).
- Computed tomography (CT) was performed in 8 patients. It confirmed in most cases the existence of the tumor and allowed a local, regional and remote study of possible lesions.

Table III: Pelvic ultrasound data.

Characteristics	Number
Location	
Right ovary	3
Ovary Left	8
Two normal ovaries	1
Ultrasound appearance	
Anechoic	0
Echogenic	3
Heterogeneous	9
Doppler capture	3
Vegetation or partition	0
Endometrial thickening	1
Recto-uterine pouch	
Ascites	4
Nodes	0
Median tumor size	69 mm

Surgical Management:

- All patients underwent surgical exploration. It was performed in 6 patients by laparoscopic and 7 patients by laparotomy from the outset.

Table IV: Gestures made during surgery.

Gesture	Number
Peritoneal cytology	10
Cystectomy	2
Ovarian biopsy	3
Unilateral adnexectomy	6
Bilateral adnexectomy	6
Hysterectomy, bilateral adnexectomy, parietal biopsy, omentectomy and appendectomy	4
Hysterectomy and bilateral adnexectomy	3

Thus, at the end of the actions performed, we can conclude that **conservative treatment was performed in 46.17%** of cases while **radical treatment was performed in 53.83%** of cases.

FIGO Classification:

According to the FIGO classification, 10 patients were in stage I and 3 patients were in stage III.

Chemotherapy:

- None of the patients received neoadjuvant chemotherapy.
- 6 of our patients, all in the group of young patients, received adjuvant chemotherapy.

Evolution and Prognosis:

Four of our patients had metastasis during the course of development, and accounted for 30.76% of the cases. For overall survival, five of the 13 patients are still alive; representing an overall survival rate of 38.46%.

CONCLUSION

The rarity of sex cord-stromal tumors contributes to a low index of suspicion.