



## Ovarian Yolk Sac Tumor: A Case Report and Literature Review

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### Abstract

Yolk sac tumor, also called endodermal sinus tumor, is a rare ovarian neoplasm, representing the second ovarian germinal neoplasm subtype after dysgerminoma, it classically occurs in adolescent and young women. We report the case of a 24 years old woman treated in the Department of Radiation Oncology Mohammed VI university hospital of Marrakech, the tumor was revealed by a rapid clinical evolution involving: pelvic distension, intermittent and moderate pelvic pain, without menstrual cycle disorder nor altered general state, the patient had undergone a fertility-sparing surgery, Alpha Fetoprotein (AFP) blood level was 1,119 mg/ml at day 40 after surgery (*vs.* 52.330 mg/ml before surgery), the young woman had received 4 BEP cycles, normalization of AFP was obtained within 5 weeks after beginning chemotherapy, after 15 months of follow-up the young woman is still in remission.

**Keywords:** Yolk sac; Tumor-endodermal; Sinus; Tumor-ovary

### Introduction

Yolk sac tumor is a rare and aggressive malignancy, representing 20% of germinal tumors in ovary, characterized by its high chemo sensitivity the challenge is to ensure disease control without compromising fertility in adolescent and young women.

### Case Presentation

We report the case of a 24 years old woman, without notable neither personal nor familial antecedent, presenting 1 month before the first consultation with a rapid clinical evolution involving: pelvic distension with moderate intermittent pain, without hormonal disorders or general state alteration.

### Investigation

Pelvic imaging showed a heterogeneous latero-uterine process of 14 cm with small peritoneal outpouring (Figure 1). AFP level was high: 52.330 mg/ml, Beta-HCG and LDH levels were normal, imaging (computed tomography) showed no secondary distant lesion.

### Differential diagnosis

In front of an adnexal mass in a young woman the first diagnosis evoked was a germinal tumor; the elevated alpha fetoprotein blood level confirmed the diagnosis even before biopsy.

### Treatment

The young woman underwent a fertility sparing surgery (laparotomic right adnexectomy+omentectomy+appendectomy) as the diagnosis of endodermal tumor was set before surgery, surgical exploration described a well limited solido-cystic latero-uterine mass without ascites or regional infiltration, without per nor post-operative incident. Pathological analysis showed a solido-cystic mass of 1,300 grams, of 18 cm × 15 cm × 7 cm, with predominant solid component (80%), of white-gray color, firm consistency, with hemorrhage, necrosis zones and mucoid cysts. The different samples of the neoplasm revealed a malignant germinal proliferation organized in glandular-alveolar proliferation and anastomotic ducts bordered by cylindrical glycogen-rich cytoplasm cells, with myxoedematous stroma (Figure 2) and Schiller-Duval bodies: central vessel surrounded by atypical cylindrical cells (Figure 3). Epiploic tissue (12 cm × 10 cm) contained a friable white nodule of 1 cm, 5 cm showing the same tumoral proliferation that characterizes the solido-cystic mass, non-

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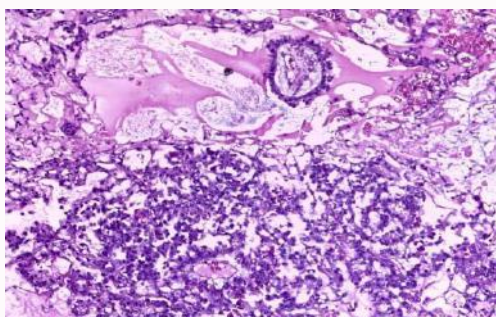
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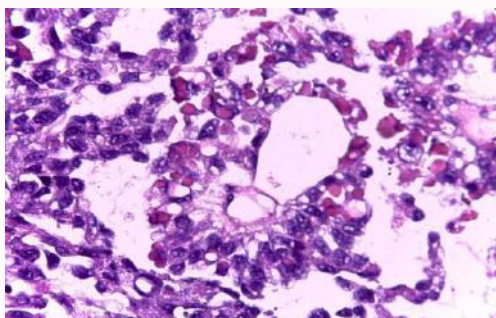
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**Figure 1:** Axial tomography sequence showing a 14 cm heterogeneous Pelvic mass.



**Figure 2:** Germinal proliferation in tubular pattern, with myxoedematous stroma (x10).



**Figure 3:** Schiller-Duval body (x40).

specific endo-appendicitis was noted. Post-operative imaging showed no residue or distant lesion, respiratory functional exploration was normal, alpha fetoprotein level was 1,994 mg/ml at J40 after surgery, LDH and HCG levels were normal.

The patient received 4 cycles of BEP protocol (Bleomycin, Etoposide, Cisplatin) with 2 episodes of grade IV neutropenia, AFP level decreased to 54 mg/ml after the second cycle of chemotherapy, normalization was obtained within 5 weeks, after a follow-up of 28 months, the patient did recuperate a regular menstrual cycle 4 months after the last chemotherapy course, imaging shows no suspect image, with normal alpha fetoprotein level, clinical surveillance does not reveal any persistent chemotherapy-related toxicity.

## Discussion

Ovarian germinal neoplasms differs from adenocarcinomas: occurrence at younger age (women of 18 to 24 years old), a diagnosis in an earlier stage (70 to 80 at stage I), a better prognosis, a high chemo-sensitivity, possibility of fertility sparing surgery rather than

radical one, and presence of specific tumor markers (AFP in yolk sac tumors). The classification of international federation of gynecology is the same for non-epithelial neoplasms of ovary. Yolk sac tumor is a non dysgerminoma malignancy arising from endodermal sinus, most often unilateral with a diameter of 5 cm to 50 cm, the typical clinical presentation is a rapid abdomino-pelvic distension, pain is the main revealing symptom and could sometimes lead to urgent surgery specially in case of ovarian torsion [1], other symptoms could include: pelvic masse, metrorrhagia, ascites, fever, symptoms related to infection or rupture of tumoral mass. Evolution is longtime loco-regional, in very advanced stages hepatic metastasis are frequent, lymph node metastasis and pulmonary ones are present in 62% and 41% respectively [2]. Imaging, even if not specific, classically shows a hyper vascularized solido-cystic image with intra-tumoral hemorrhage zones and heterogeneous enhancement after administration of contrast product [3]. Ultrasound is an important tool; it allows diagnosis, characterizes the adnexal mass, and shows eventual ascites or hepatic metastasis. CT scan permits detection of carcinosis and adenopathy even though lymph node involvement is rare in this type of germinal malignancy [4], magnetic resonance imaging shows the hyper-vascularized and hemorrhagic character of the mass.

Alpha fetoprotein is a specific marker, the association of an adnexal mass and an elevated AFP level is specific of a vitelline component, permitting to raise diagnosis with quasi-certainty even before histological proof, and therefore to orientate surgical procedure in young women [5]. The typical histological aspect is a clear cell proliferation organized in network generally of micro-cystic aspect. Schiller-Duval bodies are cellular structures that resemble fetal glomerulus, pathognomonic of endodermal tissue. Surgery, before, treated this neoplasm as adenocarcinoma (extensive surgery : hysterectomy, adnexectomy, omentectomy and lymphadenectomy), it's no more the case since 1976, when it was proven that adnexectomy was equivalent to extensive surgery in patients with stage I yolk sac tumors, the studies including series of ovarian endodermal tumors had shown equivalent results after adnexectomy compared to more aggressive surgery [4,6]. Systemic lymphadenectomy does not seem to improve the five-year survival rate [4,7-9].

Chemotherapy has dramatically changed the prognosis of these malignancies; the five-year survival rate has increased from 14% to nearly 90% [6], BEP protocol extrapolated from the treatment of testicular germ cell tumors has shown equivalent efficacy to PVB protocol (Cisplatin, Vinblastine, Bleomycin) with less toxicity [10], various studies has proven the efficacy of BEP protocol in ovarian germ cells tumors with a five-year survival rate of 94% (all stages considered) [4,7,11]. In a recent study including 84 cases of yolk sac tumors [12], the overall survival and event free survival was significantly influenced by: presence or not of ascites at diagnosis, stage, type of surgery and time to AFP normalization (before or after 42 days). The national comprehensive cancer network recommends, after surgical resection, 3 to 4 BEP cycles (4 cycles if poor prognosis factors: residue, metastatic stage, high post-operative AFP level).

A case series study of 52 cases of yolk sac tumor [13], evaluating the long-term fertility results: among 40 patients who underwent conservative surgery, 39 did recuperate a regular menstrual cycle after chemotherapy, one patient had intermittent ovarian dysfunction under second line chemotherapy for relapse, the average time to cycle recuperation after BEP protocol was 5 months, pregnancy was

achieved in 12 of 16 patients who attempted conception. The national comprehensive cancer network (2016) recommends in patients who achieved complete clinical response a surveillance of AFP every 2 to 4 months during 2 years, to detect eventual recurrence, imaging could be considered since many case reports suggest that patients who have received chemotherapy for germ cell tumors may later present with growing teratoma syndrome.

## Learning Points

Ovarian yolk sac tumor is the second germinal malignancy after dysgerminoma. Every ovarian mass should be assessed with germ cell tumor markers, to distinguish epithelial and non-epithelial malignancies in purpose to allow fertility sparing surgery. The standard of care is a fertility sparing surgery with adjuvant chemotherapy.

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