Ovarian Yolk Sac Tumor: A Case Report and Literature Review

Eddaoualline H1*, Sami H1, Rais H2, Belbaraka R3, El Omrani A1 and Khouchani M1

1Department of Radiation Oncology, Mohammed VI Polytechnic University, Morocco
2Department of Pathology, Mohammed VI Polytechnic University, Morocco
3Department of Medical Oncology, Mohammed VI Polytechnic University, Morocco

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, intermittent and moderate 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.

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 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.

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+om entectomy+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-
Axial tomography sequence showing a 14 cm heterogeneous
Germinal proliferation in tubular pattern, with myxoedematous
chemo-sensitivity, possibility of fertility sparing surgery rather
in an earlier stage (70 to 80 at stage I), a better prognosis, a high
occurrence at younger age (women of 18 to 24 years old), a diagnosis
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, 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.

**References**