A Case Report: Pregnancy with Ovarian Yolk Sac Tumor

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Background
Yolk sac tumor, known as Endodermal Sinus Tumor (EST); 1 in 15,000 to 32,000 pregnancies.
- In the United States — 0.048/100,000 women
- 62.7% were diagnosed in the first and second trimester
- Peak incidence: in adolescents to young women
- Very rare, aggressive, rapidly growing tumor
- 2nd most common germ cell tumor after dysgerminoma
- Introduction of chemotherapy has increased the survival rate to 94%
- In pregnancy, mortality and morbidity of both mother and baby increase due to technical, ethical, and religious considerations
- The prognosis still remains unsatisfactory if untreated

Case Report
This is a case of a 26-year-old primigravida, on her 23rd week age of gestation (AOG) who presented with a rapidly enlarging ovarian mass.

Patient started to have irregular menstrual cycles 5 years prior to consultation. Ultrasound findings showed bilateral ovarian cysts suggestive of dermoid cysts that measured 3 cm in diameter each. One year prior, with persistence of symptoms, bilateral ovarian cysts increased in size with the right measuring 5 cm. Patient opted to do monitoring however she was lost to follow up.

Four months prior, on her first prenatal check up at 8 weeks age of gestation (AOG), bilateral ovarian cysts were still visualized on transvaginal examination with no change in size.

On her 12 weeks AOG until 20 weeks AOG, patient complained of bloatedness associated with right flank pain. A transabdominal ultrasound showed the left ovary with no sonographic change, still measuring 5 cm. The right adnexa, however, was noted to grow to 15 x 14 x 9 cm, predominantly cystic with septations and with solid components. Only minimal color flow enhancement was noted. Sassone Score of the right ovarian new growth was more than 10. So malignancy was considered, although tumor marker CA-125 (34.74 u/mI) was within normal levels.

On 23rd week age of gestation with increasing bloatedness and abdominal pain, patient underwent Exploratory Laparotomy, Peritoneal Fluid Cytology, Right Oophorectomy, Left Oophorocystectomy. Intraperoperative findings revealed an enlarged cystic right ovary with irregular contour, ovoid, bosselated, grey to red measuring 17 x 13 x 10cm (pic 1.2).

On cut section, the mass had greyish glistening surface with a thick wall (pic 3). It showed areas of hemorrhage and necrosis. Mucin was noted within some locules. During the dissection, there was rupture of the right ovary which contained chocolate-like fluid and a separate locule with hemorrhagic fluid. The left ovary was bilocular, reddish, cystically enlarged measuring 6 x 5 x 4 cm containing, sebum and hair.

Histopathological diagnosis of the right ovary was a Germ Cell Tumor consistent with a Yolk Sac Tumor (pic 4). The left ovary showed a Mature Cystic Teratoma. The peritoneal fluid cytology revealed Atypical Epithelial Cells (pic 5).

Histopathology showed yolk sac tumor implants (pic 6) in the omentum, round ligament, bilateral adnexae, posterior uterus, posterior cul de sac, right and left pelvic side walls, left infundibulopelvic, bladder serosa, rectosigmoid, left paracolic and anterior abdominal wall. The right fallopian tube and placenta were negative for tumor.

Post-operative diagnosis was G1P1 (1001) Pregnancy Uterine delivered term live baby male, Yolk Sac Tumor Stage 3C with tumor persistence and progression. Baby delivered with Apgar Score 9,9, birth weight of 2722, maturity index 37 weeks.

Patient was then advised to start chemotheraphy with Bleomycin, Etoposide and Cisplatin after 14 days post partum and no breast feeding during chemotherapy.

Discussion

There is no available local data in the Philippines.

Symptoms:
1. Abdominal enlargement
2. Abdominal distention
3. Abdominal pain
4. A lower abdominal or pelvic mass

In most cases, EST is detected late in pregnancy due to the accompanying physiological changes of pregnancy(1).

Hallmark:
- Schiller-Duval bodies

Tumor marker:
- Elevated serum alpha-fetoprotein (AFP) levels

Diagnosis and Staging:
- Ultrasound
- Magnetic Resonance Imaging

Challenging treatment

Radical surgery followed by chemotherapy in 2nd trimester could achieve remission and rescue of fetus

Lack of incidence

For fetus >20 weeks, the dilemma is the possible effect of chemotherapy on the fetus vs the risk to the mother of disease progression if chemotherapy is delayed(2). Malignant germ cell tumor of the ovary is extremely responsive to chemotherapy. If recurrence occurs during pregnancy, subsequent chemotherapy would likely be able to save the mother.

In our case, however, the patient and relatives decided to postpone chemotheraphy because of concerns regarding the risk to the baby. It was very fortunate that the baby is physically healthy and with no known congenital anomaly.

Prognosis
If left untreated, yolk sac tumors can be fatal. However, with proper treatment the prognosis is very good. It is emphasized that surgical intervention with adequate staging remains the foundation of ovarian cancer treatment, even during pregnancy. However, the decision to perform conservative or radical surgery for pregnant women relies on histology, degree of invasion and metastasis, patient’s reproductive history and desire for fertility preservation. Under certain circumstances, the adequate staging of primary surgery is personalized. The priority is the health and wellbeing of both mother and fetus, as well as the quality of patient’s life. Five-year OS and event-free survival were, respectively, 94% and 90%(3).

Summary
Yolk sac tumor is an aggressive but a curable disease if diagnosed early and appropriate treatment has been instituted. Tumor debulking surgery with adjuvant combination chemotherapy is the treatment of choice(4). Yolk sac tumor is a rare tumor encountered in everyday practice and it is even rarer when associated with pregnancy. Therapeutic strategies for malignant tumors in the second trimester always pose an important dilemma. Delayed postoperative chemotherapy may increase the recurrence of the tumor but this same “therapeutic” chemotherapy may, on the other hand, be harmful to the fetus(5). Indeed, religious, ethical and economic factors should be considered along with thorough discussion with the patient and her relatives regarding the risks and hazards of the disease, investigations, and the treatment options available.(6,7)

References
1. Otake, M., Matsumata-Tohno, M. Yolk Sac Tumor in Pregnancy: A Case Report. JSS Journal of Obstetrical and Medical Sciences (JOMS-JSS), Volume 14, Issue 2, pp 16-20