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 germcell tumor after dysgerminoma

- Introduction of chemotherapy has increased the survival rate to 94%
- In pregnancy, mortality and morbidity of both mother and baby increases due to technical, ethical and religious considerations
- The prognosis still remains unsatisfactory if untreated

Case Report

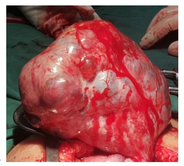
This is a case of a 26-year-old primigravida, on her 23rd week age of gestation (AOG) who presented with a rapidly enlarging ovarian mass.

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 4 cm and the left 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/ml) was within normal levels.

On 23rd weeks age of gestation with increasing bloatedness and abdominal pain, patient underwent Exploratory Laparotomy, Peritoneal Fluid Cytology, Right Oophorectomy, Left Oophorocystectomy. Intraoperative 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 grayish 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 biloculated, reddish, cystically enlarged measuring 6 x 5 x 4 cm containing, sebum and hair.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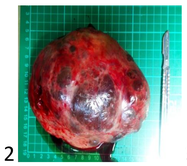
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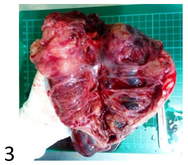
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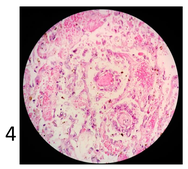
Histopathologic diagnosis of the right ovary was a Germ Cell Tumor consistent with a Yolk Sac Tumor (pic 4). The left ovary showed a Mature Cystic Teratoma. The peritoneal fluid cytology revealed Atypical Epithelial Cells (pic 5).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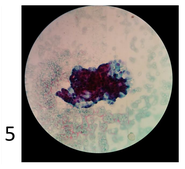
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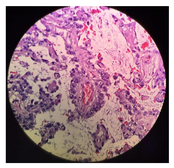
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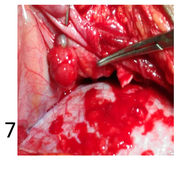
Patient was diagnosed as Gravida 1 Para 0 Pregnancy Uterine 23-24 weeks, breech, not in labor; Ovarian Carcinoma — Yolk Sac Tumor, right ovary (Stage 1C3); Mature Cystic Teratoma, Left ovary.

Post operation, chest X-ray showed normal results. A whole abdomen and pelvic Magnetic Resonance Imaging (MRI) without contrast was done which showed minimal hydronephrosis on the left, with single fetus in cephalic presentation. The liver, gallbladder, spleen, pancreas, adrenals and urinary bladder are unremarkable. No enlarged retroperitoneal and pelvic lymph nodes were noted. No ascites noted. Serum alpha feto protein (AFP) was increased at 260 IU/mL (0-5.8 IU/mL). The alkaline phosphatase level (ALP) was likewise, increased at 120.64.

Pelvic ultrasound revealed single live intrauterine pregnancy 25 weeks 1 day age of gestation by average sonar aging, and clear bilateral adnexae. The cervical length was within normal at 3.18cm with no signs of funneling.

Chemotherapy with Carboplatin was advised and offered, however, the patient and her husband opted to postpone this until after delivery due to its risk to the fetus. The patient was followed up every 3 weeks to observe both ovaries for any tumor recurrence and also for fetal growth monitoring.

At 38 weeks AOG, patient underwent Primary Cesarean Section followed by complete surgical staging and tumor debulking. Intraoperative findings showed numerous friable necrotic and hemorrhagic tumor implants scattered within the peritoneal cavity with variable measurements, the largest of which measured 8 x 6 x 4 cm. Residual tumor measured 3 x 2 x 0.5 cm (pic 7,8,9).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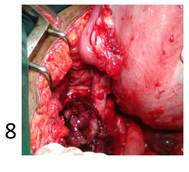
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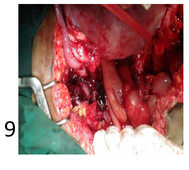
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Histopathology showed yolk sac tumor implants (pic 6) in the omentum, round ligament, bilateral adnexae, posterior uterus, posterior cul de sac, right and left pelvic side walls, left infundibulopelvic, bladder serosa, rectosigmoid, left paracolic 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 chemotherapy with Bleomycin, Etoposide and Cisplatin after 14 days post partum and no breast feeding during chemotherapy.

Discussion

RARE!

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:	Tumor marker:	Diagnosis and Staging:
Schiller-Duval bodies	Elevated serum alpha-fetoprotein (AFP) levels	• Ultrasound • Magnetic Resonance Imaging

Challenging treatment

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

Lack of incidence

TREAT ? **DILEMMA!** NOT TO TREAT ?

Early pregnancy →	TERMINATE
1st trimester →	HIGH RISK
>20 weeks →	DILEMMA

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 chemotherapy 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(2,4). 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. Delaying postoperative chemotherapy may increase the recurrence of the tumor but this same "therapeutic" chemotherapy may, on the other hand, be harmful to the fetus(2). 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.(1,2)

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