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CASE REPORT

Pure ovarian choriocarcinoma - an aggressive and rare entity

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ABSTRACT

Pure ovarian choriocarcinomas are exceedingly rare cancers that develop from either the ovary's gestational tissue (ovarian ectopic pregnancy) or the ovary's pure germ cells. They are divided into two types: gestational ovarian choriocarcinoma (GCO) and non-gestational choriocarcinoma (NGC) based on their origin (NGCO). The presence of GCO is indicated by a history of amenorrhoea with sexual intercourse, very high serum beta hCG levels, and the appearance of choriocarcinoma with a corpus luteal cyst in histology, but the DNA polymorphism and presence of paternal DNA in the tumour clinches the diagnosis. Cytoreductive surgery and adjuvant chemotherapy are the mainstays of treatment for pure ovarian choriocarcinomas. Here we present a case of pure ovarian choriocarcinoma, likely of gestational origin.

Keywords: Choriocarcinoma, gestational, ovary, ovarian choriocarcinoma.

Pure ovarian choriocarcinomas are extremely rare malignancies that arise from either gestational tissue in ovary (ovarian ectopic) or from pure germ cells of the ovary. On the basis of origin, they are classified as gestational ovarian choriocarcinoma (GCO) and non-gestational chorio-carcinoma (NGCO). The reported incidence of GCO is 1 in 369 million pregnancies while for NGCO it is less than 0.6% of ovarian germ cell tumors.[1,2] Both the variants are seen in young women of reproductive age group. The two variants cannot be differentiated clinically or histopathologically. DNA polymorphism analysis is a useful modality to differentiate between the two for therapeutic and prognostic purposes.[3] The treatment of pure ovarian choriocarcinomas essentially comprises of cytoreductive surgery and adjuvant chemotherapy.[4] Here we present a case of pure ovarian choriocarcinoma, likely of gestational origin.

Case

A 30 year old female, G4P1L0A2 was admitted to BPS GMC (W), Khanpur Kalan, Sonepat, Haryana on 30th April, 2020 with amenorrhoea of 2.5 months, pain lower abdomen and multiple episodes of bleeding per vaginum for the last 6 days. She had one episode of syncopal attack on the day of admission. On physical examination, her PR=68/min, BP=100/60 mmHg, RR=14/min, SpO_2=99% on room air. Her pelvic examination revealed pulled up cervix, a tender mass of around 8-10 cm in right fornix and fullness with tenderness in left fornix. Her urine pregnancy test (UPT) was negative and all other hematological investigations were normal. Sonography revealed a normal sized uterus with a large heterogenous solid cystic mass lesion of size 8x8 cm^2 arising from right ovary with bulky left ovary with the probable diagnosis of right adnexal ectopic pregnancy. Serum beta hCG was sent for the patient inspite of negative UPT. Her serum beta hCG levels came out to be more than 5,00,000 mIU/mL and MRI pelvis was planned. Her MRI pelvis showed a right adenexal mass of 9.6x9.1x6.6 cm^3 in pelvis with hemorrhagic component, right hematosalpinx and bilateral bulky ovaries – possibility of right molar ectopic gestation with bilateral theca lutein cysts. With the provisional diagnosis of right molar ectopic pregnancy, a laparotomy was performed after written informed consent.

Intraoperatively, a 10x10 cm^2 lobulated, hemorrhagic ovarian mass was present on right side with right hematosalpinx attached to a rudimentary horn (figure 1). On

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the left side, a 6x6 cm^2 lobulated, hemorrhagic ovarian mass was seen with swollen fallopian tube as shown in figure 1. Approximately 300cc of hemoperitoneum was noted. All the other abdominal and pelvic viscera were normal on examination. A right salpingo-oophorectomy was done with rudimentary horn excision and omental biopsy was taken.

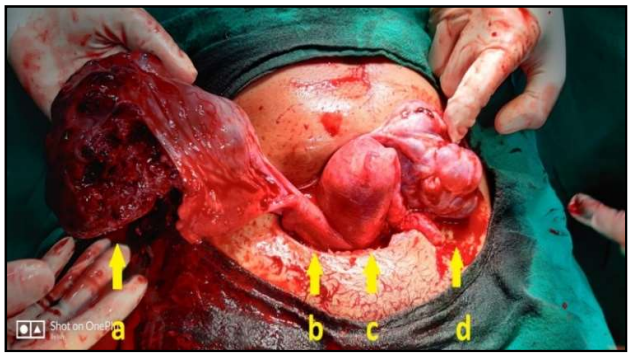

Figure 1: Intraoperative picture showing a) Right sided 10x10 cm lobulated, hemorrhagic ovarian mass with right hematosalpinx, b) rudimentary horn, c) unicornuate uterus, d) Left sided 6x6 cm lobulated, hemorrhagic ovarian mass with swollen fallopian tube.

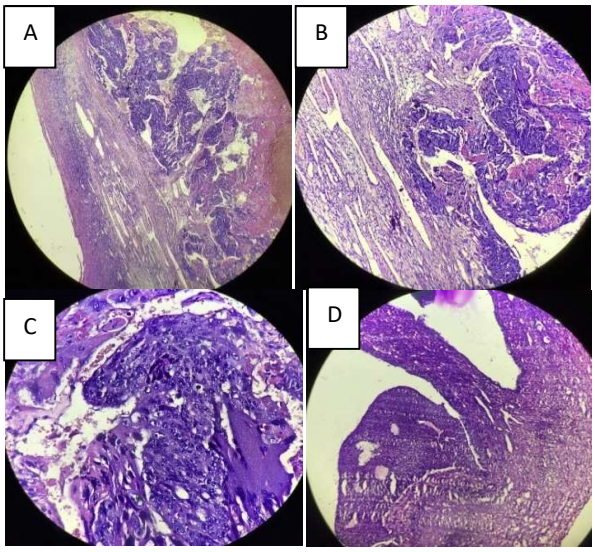

Figure 2: A) Microscopic picture showing sheets of tumor cells alongwith areas of hemorrhage and a rim of normal ovarian parenchyma (H&E 40x); B)&C) showing biphasic tumor cells (cytotrophoblasts and syncytiotrophoblasts) arranged in sheets and nests characteristic of choriocarcinoma (H&E 100x & 400x); D) showing part of corpus luteal cyst (H&E 100x).

Histopathological examination of tumor showed biphasic tumor cells (atypical cytotrophoblasts and syncytiotrophoblasts) arranged in sheets and nests characteristic of choriocarcinoma alongwith a corpus luteal cyst as depicted in figure 2. Specimen from omentum revealed presence of tumor cells and three lymph nodes present in omentum were found to be free from tumor. Hence, a final diagnosis of stage III A2 (FIGO) choriocarcinoma ovary was made and patient was planned for multiagent chemotherapy. Post operative day 7 serum beta hCG titre decreased to 15,000 mIU/mL. Now patient is under chemotherapy.

Discussion

Pure ovarian choriocarcinoma is a rare and aggressive tumor arising from gestational tissue (gestational choriocarcinoma) or pure germ cells of the ovary (non-gestational choriocarcinoma). The etiology of choriocarcinoma has been ascribed to four different sources: from maternal germ cells; from an ovarian pregnancy; from metastasis from a regressed or occult uterine primary; or in infants from metastasis from the placenta.[5] In our case, the origin was from ovarian pregnancy probably. The literature review showed the first case of NGCO was reported in 1985 and GCO in 1996. Nan Jia et al published their paper in 2017 and summarized 48 cases (including their own).[6]

The incidence of gestational choriocarcinoma is 1 in 369 million pregnancies.[1] In our institution, established in September 2011, the delivery rate is approximately 5,000 deliveries per year and it is the first case reported approximating to 1 in 45,000 deliveries. The patients usually present with non specific symptoms like acute abdomen and amennorhoea which can mimic other common conditions in young women like hemorrhagic ovarian cyst, tubo-ovarian abscess, ovarian torsion and ectopic pregnancy.[7] Our patient presented in the third decade of life, married and parous female with no live issue, with 2.5 months of amennorhoea and very high levels of serum beta hCG (more than 5,00,000 mIU/mL) with a negative urine pregnancy test. The reason for negative urine pregnancy test is that when too much hCG is present in a sample (usually above 5,00,000 mIU/mL), the antibodies may fail to bind as they are saturated and the test appears out to be negative.[8] The presence of a normal sized uterus with an adnexal mass and increased serum beta hCG levels alongwith radiographic findings pointed towards the probable diagnosis of right molar ectopic pregnancy.

As our patient was a parous female with no live issue, fertility preserving surgery (right salpingo-oophorectomy with omental biopsy) was performed with due consent. On HPE, it was found to be ovarian choriocarcinoma with presence of a corpus luteal cyst. H/O amennorhoea with

sexual intercourse, very high serum beta hCG levels, presence of choriocarcinoma with presence of a corpus luteal cyst in histopathology directed towards the presence of GCO but it is the DNA polymorphism and presence of paternal DNA in tumor that finally clinches the diagnosis.[9] Since DNA polymorphism analysis is an expensive and rarely available technique, we were unable to carry out in our patient. The differentiation of the two variants is important as the gestational type has a better prognostic outcome than the non-gestational one.[10]

The combination of cytoreductive surgery alongwith chemotherapy is the standard treatment.[6] Despite thorough research, there was found to be no association between the occurrence of ovarian choriocarcinoma and uterine anomalies. The patient's serum beta hCG levels decreased rapidly after surgery and she is under multiagent chemotherapy.

Conclusion

Ovarian choriocarcinoma is a very rare and aggressive tumor. In a female presenting with adnexal mass with very high serum beta hCG levels alongwith absent intrauterine trophoblastic disease, the possibility of ovarian choriocarcinoma should be kept in mind as early diagnosis and treatment play a major role in enhancing patient's prognosis.

Conflict of interest: None. **Disclaimer:** Nil.

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