

Primary Pure Ovarian Choriocarcinoma: A Case Report and Review of Literature

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Abstract: Primary ovarian choriocarcinoma is a rare tumour which could either be of gestational or nongestational origin. Differentiating one from the other is difficult but important for effective treatment because the nongestational type has a poor prognosis. We present a case of primary pure ovarian choriocarcinoma occurring in a nullipara with no pregnancy history.

Keywords: primary, ovarian, choriocarcinoma, gestational, nongestational.

I. Introduction

Primary choriocarcinoma occurring outside the uterus is very uncommon and is found mainly in the genital tract in patients with coincident or a preceding pregnancy.¹ The very rare primary ovarian choriocarcinoma can originate from an ovarian pregnancy (gestational type which is more common) or as a type of germ cell tumour (nongestational type)². Primary nongestational choriocarcinoma arises from germ cells of the ovary instead of the gestational tissues in the ovary³. Majority of these primary nongestational choriocarcinomas of the ovary occur with other germ cell tumours^{4&5}.

The pure variety of primary ovarian nongestational choriocarcinoma is an exceedingly rare tumour, and we report such in this article.

II. Case Report

Miss A.O, a 26 year old nulliparous secondary school leaver who had her last menstrual period on the 15th of May 2013, presented to our Gynaecological Emergency Unit on the 29th of May 2013, on account of abdominal swelling of one year duration and abdominal pain of 6 months duration. Abdominal swelling was insidious in onset but increased rapidly over the last 6 months prior to presentation. Abdominal pain was sudden in onset, initially localized at the right iliac fossa but became generalized over the last 3 weeks prior to presentation. Pain was burning in nature, non-colicky, radiating to the waist and upper thigh. It gradually increased in severity and was associated with dysuria and urinary frequency but there was no fever. There was associated weight loss but no history of cough or difficulty in breathing. There was no associated vaginal bleeding.

Her menarche was at 15 years of age. There was no family history of uterine fibroid, breast cancer or female genital tract malignancies.

At presentation, she was cachectic, in painful distress, not pale, not jaundiced or cyanosed. She was not febrile or dehydrated and had no pedal oedema. There was moderate non-localized abdominal tenderness with a palpable abdominopelvic mass of 22 weeks size with irregular outline.

The mass was cystic, mobile, and not attached to adjacent structures. A working diagnosis of right ovarian malignancy was entertained.

Full blood count and serum electrolyte, urea and creatinine were normal. Serum B-HCG was markedly elevated (105,000 mIU/ml). The liver enzymes were elevated and chest X-Ray showed normal study. Abdominopelvic ultrasound scan revealed a right multiloculated ovarian mass measuring 17 x 16cm with solid components. Uterus was sonographically normal. An intravenous urogram done showed right obstructive uropathy.

She subsequently had laparotomy and a right salpingo-oophorectomy. Intraoperative findings included a moderate hemorrhagic ascitis, grade II pelvic adhesions and a huge right ovarian mass with tiny seedlings on the peritoneum, liver, intestine and urinary bladder. The uterus and the fallopian tubes were grossly normal.

The tumour sent for histopathological evaluation measured 16 x 14 x 10cm. The cut surface was solid and haemorrhagic with few peripherally located cystic spaces. Also, two small pieces of tissue from the peritoneum was sent.



Figure 1: Cut surface of right ovarian mass showing solid tumour with areas of haemorrhage, necrosis and peripheral cystic spaces.

Microscopic examination showed proliferating trophoblastic cells within a wide background of haemorrhage and necrosis with moderate secondary inflammation. Mitotic figures were abundant with extensive vascular invasion. There was no evidence of other germ cell elements or co-existing tumours. Histologic sections from the peritoneal biopsies showed proliferation of malignant trophoblastic cells. A histopathologic diagnosis of primary ovarian choriocarcinoma with spread to the peritoneum was made.

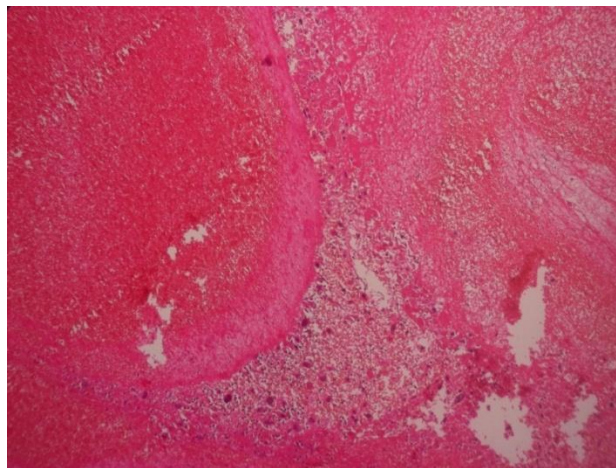


Figure 2: Proliferating trophoblastic cells within a wide background of haemorrhage and necrosis (H&E x 100).

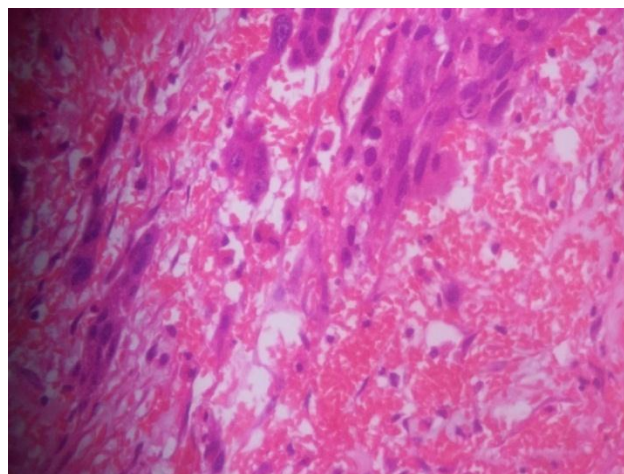


Figure 3: ovarian choriocarcinoma showing characteristic biphasic pattern of proliferating cytotrophoblasts and syncytiotrophoblastic cells (H&E x 400).

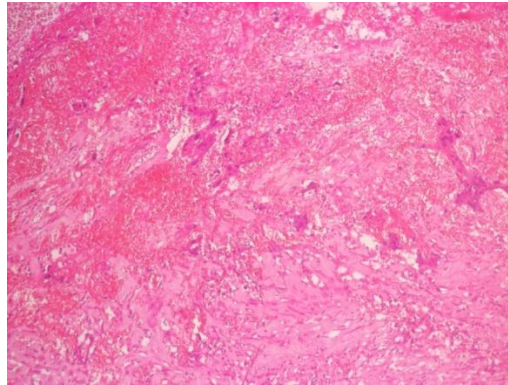


Figure 4: Malignant biphasic trophoblastic cells scattered within a haemorrhagic ovarian stroma with moderate reactive chronic inflammation (H&E x 100).

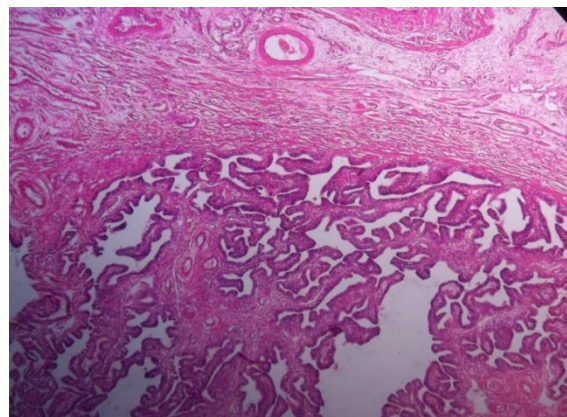


Figure 5: Histologic section of right fallopian tube showing normal histology with absence of tumour cells (H&E x 40).

Post operative recovery was uneventful and patient was discharged home on the 8th day post operation with referral for appropriate adjuvant chemotherapy. On follow up however, it was discovered that she did not travel for chemotherapy due to financial constraints. She was later reported to have died within one month of discharge from our hospital.

III. DISCUSSION

Primary Ovarian Choriocarcinoma is a very rare tumor and may be of gestational or nongestational origin^{1&4}. They are not only rare but aggressive tumours, comprising about 1% of all ovarian tumours⁶. Gestational ovarian choriocarcinoma are often associated with normal or molar pregnancies, ovarian pregnancy or from intratubal primary disease^{4,7&8}. The estimated incidence of primary ovarian gestational choriocarcinoma is 1 in 369 million Pregnancies⁹.

Our patient was a nullipara, presenting in the reproductive age with abdominal mass and abdominal swelling similar to the presentation pattern for this disease as documented in other reports. Apart from being a nullipara, her last menstrual period was on the 15th of May, 2013 and both physical examination and abdominopelvic ultrasound scan revealed no other tumour in the uterus or fallopian tubes. We therefore believe that the index case with a confirmatory histopathological diagnosis showing no other germ cell elements was a pure form of nongestational choriocarcinoma.

She had no fever on presentation and the laboratory investigations did not suggest associated urinary tract infection, however, the urinary frequency and dysuria may be explained by the presence of bladder seedlings and the pressure from the massively enlarged right ovary. Also, liver metastasis may explain the elevated liver enzymes as was seen at laparotomy.

Nongestational primary choriocarcinoma of the ovary can be pure but more frequently occurs as a component of a mixed germ cell tumour like teratoma, dysgerminoma, or endodermal sinus tumour^{10,11&12}. Pure choriocarcinomas of the ovary are extremely rare tumours¹³.

Saito et al,¹⁴ first described the diagnostic criteria for nongestational choriocarcinoma to include the following; 1. Absence of disease in the uterine cavity. 2. Confirmation of the disease by histopathological examination. 3. Exclusion of molar pregnancy. 4. Exclusion of intrauterine pregnancy. The index case satisfied

these requirements completely and is diagnosed nongestational ovarian choriocarcinoma. We are also aware of the ascertainment by Jacobs et al¹⁵ that a nongestational ovarian choriocarcinoma can be diagnosed unequivocally only in a sexually immature patient or who has been unable to conceive or has never had sexual intercourse. The index case was a nullipara and therefore satisfies Jacobs requirement also.

In the series by Axe et al,⁴ the primary treatment for all the six cases was unilateral oophorectomy followed by adjuvant chemotherapy as indicated. The two patients that had extended surgeries were based on review after the initial treatment. The index case being a nullipara who needed to preserve her fertility had initial unilateral salpingo-oophorectomy and was for adjuvant chemotherapy thereafter. Gestational choriocarcinoma responds well to single-agent chemotherapy with methotrexate or Actinomycin D. For those cases like ours with distant metastasis to the liver or central nervous system, pre-treatment serum B-HCG levels of more than 40,000 mIU/ml, post term gestational trophoblastic disease or a period of more than four months since pregnancy, triple-agent chemotherapy as been advocated. However patients treated by salpingo-oophorectomy with or without single-agent chemotherapy also achieved long-term survival^{15&16}.

The index case ran an aggressive course with documented peritoneal, intestinal, bladder and liver tumour seedlings similar to what is seen in other reports¹³. Gestational ovarian choriocarcinomas have better prognosis than their non gestational counterparts^{4,11,17,18&19}. Furthermore, prognostically, pure choriocarcinomas are often lethal unlike the mixed types in which good survival rates have been obtained⁴. Overall, unlike placenta choriocarcinoma, ovarian choriocarcinomas are generally unresponsive to chemotherapy and are often lethal¹³.

While the clinical characteristics and histomorphology of both tumour types are identical, the distinction of gestational and nongestational ovarian choriocarcinoma is very difficult but necessary for effective treatment and prognostication^{3,11&18}. Histomorphological distinction is possible when unequivocal evidence of pregnancy or other germ cell neoplasms are present. There are no specific or diagnostic ultrastructural or immunohistochemical features for either. Currently, DNA polymorphism analysis for identification of paternal sequences is used to diagnose or confirm gestational or non gestational choriocarcinoma^{3,5,20,21}. Available literatures show that only a few cases of nongestational choriocarcinoma have been diagnosed by DNA polymorphism analysis. Since in our centre and environment such facility is not yet available, the index case was diagnosed as a nongestational type of ovarian choriocarcinoma based on histomorphologic study.

IV. Conclusion

Pure ovarian nongestational choriocarcinomas are extremely rare and very lethal tumours that require early diagnosis and aggressive treatment by surgery followed by adequate adjuvant chemotherapy.

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