Primary Nongestational Ovarian Choriocarcinoma Mistaken for Ectopic Pregnancy: A Case Report

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Abstract

Primary choriocarcinoma of the ovary is an extremely rare neoplasm. According to its histologic origin, it is classified as gestational or non-gestational. Non-gestational choriocarcinoma of the ovary is a germ cell tumor with a worse prognosis than gestational choriocarcinoma. Due to the rarity of the tumor, there is a lack of information on the clinico-pathologic features, diagnosis, and treatment. They pose diagnostic challenges in reproductive age group patient because of elevated human chorionic gonadotropin (HCG). we report a case of non-gestational or primary choriocarcinoma which is mistaken for an ectopic pregnancy clinically, resulting in delay in diagnosis and proper management.

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Introduction

Germ cell malignancies may represent up to 15% of ovarian cancers in Asian and African-American nations¹. Ovarian choriocarcinoma is a germ cells tumor, very rare malignant neoplasia, about 40 cases have been reported in medical literature². Ovarian choriocarcinoma may be gestational or non-gestational. The estimated incidence of gestational ovarian choriocarcinoma is 1 in 369 million pregnancies³. Non-gestational ovarian choriocarcinomas account ≤0.6% of all ovarian neoplasms; the pure type is extremely uncommon⁴. Due to the rarity of pure ovarian choriocarcinomas, information on the clinico-pathologic features, diagnosis, and therapeutic options is limited. Here we present a case of a pure ovarian choriocarcinoma, likely of non-gestational origin, and discuss the diagnosis and treatment together with a brief review of the literature.

Case Report

A 22- years-old married woman presented with amenorrhoea for 3 months with usual sign symptoms of pregnancy like nausea, vomiting, anorexia and weakness. Her pregnancy was confirmed by urinary pregnancy test. She also complained of lower abdominal pain which initially continuous and dull aching in nature without any radiation but last 3 days pain became severe. On examination patient was severely anaemic, her pulse 102 beat/min and BP 90/60 mm of hg. An ill-defined mass was present in hypogastrium & left iliac fossa. The Consistency of mass was doughy and size could not be delineated properly due to tenderness. Bimanual examination revealed, a soft and tender mass felt through left and posterior fornices, there was no cleavage between the mass and uterus. Our clinical diagnosis was chronic ectopic pregnancy. Investigation report shows- Beta HCG level-299 mIU/ml, USG shows bilateral adnexal mass in which left adnexal region a cystic mass and in right adnexal region a moderately complex mass (5.7x4.8) predominantly cystic having septation.

So as our clinical diagnosis was chronic ectopic pregnancy, decision for emergency laparotomy was taken. During laparotomy left ovary found completely replaced by red,lobulated and friable about 15x12x8 cm mass, which is completely covered with omentum and adherent with posterior surface of uterus.

Follicular cyst present in right ovary otherwise right ovary and tube were normal in appearance. Peritoneal washings were obtained for cytology. Left sided salpingo-ophorectomy was performed. On gross examination, it was red, haemorrhagic and angry looking but capsule of the

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tumour was intact. Microscopic examination revealed characteristic histological feature of pure choriocarcinoma with anaplastic cytotrophoblast and syncytiotrophoblast cells in clusters, small sheets and diffusely and large areas of hemorrhage and necrosis.

**Discussion**

Choriocarcinoma of the ovary is a rare and aggressive germ cell tumor. Primary choriocarcinoma of ovary could be gestational or non-gestational in origin. The distinction between the two is difficult, but necessary, as the non-gestational type has bad prognosis. Primary gestational choriocarcinoma arising from an ectopic ovarian pregnancy. Most non-gestational choriocarcinoma of ovary are arising from germ cell differentiating to trophoblastic component, occur in admixture with other germ cell tumours like teratoma, endodermal sinus tumor, embryonal carcinoma or dysgerminoma. The most common primary site of choriocarcinoma is intrauterine. Primary extra-uterine choriocarcinoma has been reported to occur in the fallopian tube, ovary, and elsewhere in the abdomen and pelvis. All types secret α-hCG. However, α-hCG levels are usually lower in non-gestational variants in comparison to gestational types. Monitoring of serum α-hCG can be useful method in evaluating response to therapy.

There are no distinctive ultrastructural or immunohistochemical differences between gestational and non-gestational choriocarcinomas. Diagnosis of pure non-gestational choriocarcinoma is uncertain without the DNA analysis. Genetic analysis must be a useful tool in determining the origin of the choriocarcinoma. However, a search for paternal DNA in tumor allows a definite distinction between gestational and non-gestational types. Tumors with gestational origin have paternal genomic structure while non-gestational tumors have genomes of only maternal origin without any alleles from paternal origin. Since DNA polymorphism analysis techniques are always expensive and not generally available in all medical centers, the application is limited. For this very reason, we could not perform molecular genetic analysis on the tumor from our patient.

The diagnosis of primary extra-uterine choriocarcinoma is challenging because the clinical symptoms are often nonspecific and can mimic other, more common conditions that occur in young women, such as a hemorrhagic ovarian cyst, tubo-ovarian abscess, ovarian torsion, and ectopic pregnancy. It is particularly challenging to differentiate choriocarcinoma from the more common ectopic pregnancy in a young woman who has a positive result of a pregnancy test, pelvic pain, and an adnexal mass. In our patient, the imaging findings, the clinical signs and symptoms, and the rarity of choriocarcinoma mistakenly favored the initial diagnosis of ectopic pregnancy. Even after initial diagnosis, the patient’s presentation with vaginal bleeding continued to favor primary intrauterine choriocarcinoma metastatic to the adnexa rather than primary extra-uterine.
ovarian choriocarcinoma. The rarity of the true diagnosis led to emergency laparotomy followed by left sided salphingo–oophorectomy. It delays actual diagnosis until histopathology report available and administering appropriate chemotherapy. Saito et al. first described the diagnostic criteria for NGCO in 1963. These include absence of disease in the uterine cavity, pathological confirmation of disease, and exclusion of molar pregnancy and of intrauterine pregnancy. All the criteria were fulfilled in this case. Owing to the rarity of the condition, little information on therapeutic options is available.

To date, no definitive treatment modality has been established for pure ovarian choriocarcinomas due to the low incidence. Thus, pure ovarian choriocarcinomas are generally treated by the same protocols used for ovarian germ-cell tumors and gestational trophoblastic disease. The treatment options of malignant germ cell tumour depend on surgico-pathological staging. The definitive surgical treatment is total abdominal hysterectomy and bilateral salpingectomy with omentectomy and peritoneal biopsies. For certain authors, the distinction between gestational and nongestational choriocarcinoma does not seem essential because the treatment would depend essentially on the stage of the tumor, without consideration of its gestational or non-gestational character. Then according to the histopathological type, adjuvant chemotherapy will be associated. Goswami et al. reported 29 cases in 2001, mentioned that pure ovarian choriocarcinoma responds well to the combination of surgical ablation and post-operative chemotherapy; the rate of survival in 2 years was 81% with the patients having received an adjuvant chemotherapy against 28% with the patients having not had this type of treatment. Non metastatic gestational choriocarcinomas respond effectively to methotrexate. On the other hand, nongestational tumors react less well to the methotrexate and require multiple agent chemotherapy. BEP chemotherapy is the current regimen for germ cell tumors of the ovary. Thus, our patient underwent BEP protocol of chemotherapy for a presumed primary pure non-gestational choriocarcinoma. The patient had a good response to the BEP regimen with a satisfactory decrease in the serum â-hCG level. There was no evidence of recurrence or metastasis after a 6-month follow-up.

In conclusion, our case provides evidence that conservative surgery in combination with post-operative adjuvant chemotherapy may be an alternate therapeutic strategy for pure ovarian choriocarcinomas of younger patient. Pure ovarian choriocarcinoma is aggressive with a high risk of metastasis, close follow-up with serum â-hCG and imaging examinations is essential.

References