



A Rare Case of Primary Nongestational Choriocarcinoma of the Ovary

KEYWORDS

Nongestational, Choriocarcinoma, Ovary, Chemotherapy.

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ABSTRACT *Pure primary ovarian choriocarcinoma is an extremely rare and aggressive tumor. It can be of gestational or non gestational in origin. The gestational type can arise from an ovarian pregnancy or can be of metastatic origin from uterine choriocarcinoma. The non gestational type is a very rare germ cell neoplasm. It is important to distinguish between two types of choriocarcinomas as non gestational origin is highly malignant and has worse prognosis than gestational type. But it is very difficult to differentiate by routine histological examination.*

Nongestational choriocarcinoma has been found to be resistant to single agent chemotherapy.

It occurs usually around 13 years of age and is mainly confined to females under 20.

INTRODUCTION

Pure primary ovarian choriocarcinoma accounts for less than 1% of ovarian tumors.¹ Shiromizu et al recently studied 467 ovarian germ cell tumors and found only one choriocarcinoma.² Gestational choriocarcinoma of the ovary can be primary, associated with ovarian pregnancy, or metastatic, arising from a primary gestational choriocarcinoma in the uterus. Non gestational choriocarcinoma of the ovary can be pure, but is more frequently associated with other germ cell tumors.³

Primary ovarian choriocarcinoma arising from germ cell is an extremely rare occurrence.⁴ Primary nongestational ovarian choriocarcinomas (NGCOs) are usually seen with other germ cell tumors.⁵ Pure primary nongestational ovarian choriocarcinoma of the ovary is extremely rare.⁶

Choriocarcinoma is a rapidly invasive, human chorionic gonadotropin (hCG) producing neoplasm which usually has an intrauterine location.⁷ Choriocarcinoma metastasize early and widely through both the venous and lymphatic systems.

Nongestational pure choriocarcinoma is so rare that the prognosis, chemo-sensitivity, and its genetic analysis have not been decided compared with that of gestational type. It is necessary, but difficult to distinguish nongestational choriocarcinoma from gestational choriocarcinoma except by DNA analysis.⁸

Case report :

A 20 year old unmarried girl came with the complains of pain abdomen since 6 months & bleeding per vagina since 15 days associated with generalized weakness to the Gynaecology OPD , S.V.S Medical College & Hospital. There is no past history of abortion / normal pregnancy. Abdominal examination revealed a diffuse, soft, non-tender mass of 20-22 week size.

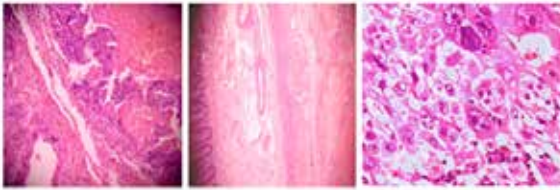
Ultrasound pelvis showed large ill defined, heterogeneously hyperechoic, soft tissue mass arising from right ovary measuring – 20x7 cm with large necrotic areas - Suggestive of solid right ovarian malignancy. The uterus was normal in size, shape and echotexture. No focal lesion was

seen in the uterus. Right fallopian tube and left adnexa visualized. Intrauterine or ectopic pregnancy was ruled out. USG appendix showed features of chronic appendicitis.

Exploratory laparotomy was done and specimens sent for histopathology. Gross showed multiple, large grey brown nodular masses and larger one measuring 9x9x2 cm. Cut section showed solid, necrotic and hemorrhagic areas. Also, received appendectomy specimen measuring 5x 1.5 cm.



H & E stained sections of the right ovarian mass revealed tumor area consisting of extensive areas of hemorrhage, necrosis and clusters of malignant trophoblastic cells with enlarged, hyperchromatic and bizarre nuclei. No other germ cell element was detected. Appendix biopsy showed metastatic deposits. In the immediate postoperative period serum b-hCG level was 66,580 mlu/ml .



(H & E – 10X) (H & E – 40X)

DISCUSSION

Ovarian germ cell tumors (OGCTs) arise primarily in young women between 10 and 30 years of age.¹⁰ In a review of the literature, the age range of the patients was seven months to 35 years with a mean of about 13 years.¹¹ Most ovarian choriocarcinomas are gestational in origin, and usually are metastases from uterine or tubal choriocarcinomas.⁵

Primary ovarian choriocarcinomas are rare aggressive tumors, and comprise only 1% of all ovarian tumors.¹² They can present in the pure form, or may occur in association with other germ cell tumors like teratoma, dysgerminoma or yolk sac tumor.¹³

Pure ovarian choriocarcinoma can be of gestational or nongestational type. Gestational ovarian choriocarcinomas occur in women of reproductive age, and are often associated with normal or molar pregnancies.⁷

It is important to distinguish between gestational and nongestational choriocarcinomas, since nongestational choriocarcinomas have been found to be resistant to single agent chemotherapy, have a worse prognosis & require aggressive combination chemotherapy.

The diagnostic criteria for nongestational choriocarcinoma which were first described by Saito et al¹⁴ are

1. Absence of disease in the uterine cavity
2. Pathological confirmation of disease
3. Exclusion of molar pregnancy
4. Exclusion of coexistence of intrauterine pregnancy

According to these criteria, the present case could be diagnosed as nongestational choriocarcinoma, but Jacobs et al¹⁵ stated that a nongestational type of ovarian choriocarcinoma can be diagnosed with certainty only in a patient who is sexually immature, unable to conceive, or has never had sexual intercourse. With such stringent criteria, it is difficult to diagnose a case of choriocarcinoma as nongestational.

Distinction of nongestational choriocarcinoma from gestational choriocarcinoma is impossible on histomorphology unless an evidence of pregnancy or other germ cell neoplasms is encountered. There are no ultrastructural or immunohistochemical features unique to either. DNA analysis for identification of paternal sequences can establish the diagnosis of gestational or nongestational choriocarcinoma.^{5, 16, 17}

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