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*Case Report*

# Pure ovarian choriocarcinoma: report of two cases

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## Abstract

Pure primary ovarian choriocarcinoma is an extremely rare condition of gestational or nongestational origin. The possibility of gestational origin can be suspected by the presence of a corpus luteum of pregnancy but definite diagnosis would be based on genetic analysis. Here, we present two cases of pure ovarian choriocarcinoma in the forth decade of life with the possibility of gestational origin.

**KEYWORDS:** Gestational, Choriocarcinoma, Ovary.

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Germ cell malignancies may represent up to 15% of ovarian cancers in Asian and African-American nations, where epithelial ovarian cancers occur less commonly.[1]

Pure ovarian choriocarcinoma is extremely rare, with about 40 cases described in the English language articles.[2] Choriocarcinoma may be gestational or nongestational. The gestational type arises from an ovarian pregnancy or is of metastatic origin from uterine choriocarcinoma, whereas the nongestational type is an extremely rare germ cell neoplasm. To distinguish pure nongestational choriocarcinoma of the ovary from metastatic or primary gestational choriocarcinoma, DNA polymorphism analysis is a useful method.[3,4] The gestational variants are extremely sensitive to chemotherapy with overall cure rate approaching 98% in specialized centers.[5] The prognosis correlates with the bulk of tumor as well as the sites and number of metastases.[6] We present two cases of pure ovarian choriocarcinoma with possibility of gestational origin.

## Case Report

**Case 1:** A 31-year-old woman, with a gestational history of G9, P1, Ab8 and L1 was admitted to Mirza Koochak Khan Hospital, Tehran, Iran in June 2001 with the symptoms and signs of acute abdomen. She had 5 years secondary infertility of unknown reason following her last abortion and gave the history of a missed menstrual period as well as 50 days of spotting. The serum β-hCG level was more than 1000 mIU/ml. Pelvic sonography revealed enlarged right ovary and a 2.7 cm left ovarian cyst. No abnormal finding was detected in uterus. In view of an adnexal mass along with positive pregnancy test, a possibility of ectopic pregnancy was considered. A right salpingo-oophorectomy was carried out. Gross examination of right ovary revealed a 7×7×4.5 $cm^3$ mass with a hemorrhagic cut surface. Following thorough sampling of the ovarian mass (1 section per cm of the greatest dimension of the tumor), microscopic examination revealed characteristic histological fea-

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tures of choriocarcinoma, composed of bilaminar structures of cytotrophoblasts and syncytiotrophoblasts with severe nuclear atypia and mitotic figures admixed with necrotic tissue. A corpus luteum of pregnancy (Figure 1) with multiple colloid bodies and calcifications was also observed (Figures 2 & 3).The patient received Etoposide/ Metotrexate / Actinomycin D/ Cisplatin/ Etoposide (EMA-CE) regimen for treatment and serum β-hCG level decreased to 70 mIU/mL. She underwent second laparotomy for careful examination of tumoral involvement. Specimens from left ovary and omentum revealed no histologic features of malignancy. A corpus luteum cyst was detected in the left ovary. Computed tomographic scan revealed no brain or lung metastases. Serum alpha- fetoprotein (AFP) was in normal range. After the surgery, she took 4 courses of chemotherapy with EMA-CE regimen. The level of β-hCG decreased to below the cut-off value. There has been no evidence of tumor recurrence during seven years follow-up.

**Case 2:** A 32-year-old woman with a gestational history of G3, P2, Ab1 and L2 was admitted to Mirza Koochak Khan Hospital, Tehran, Iran in September 2003 with nausea¸ vomiting and vaginal spotting. Sonography revealed a large necrotic mass in the left adnexa. Serum β-hCG level was 5500 mIU/mL and AFP level was within the normal range. A large necrotic mass in left ovary with extension to the posterior aspect of uterus was identified in laparotomy. Total abdominal hysterectomy and bilateral salpingo-oophorectomy, tumor debulkation and infracolic omentectomy were performed. On gross examination, there was a left ovarian mass measuring 13×11×10 $cm^3$ with lobulated and hemorrhagic-necrotic cut surface. Microscopic examination of the tumor demonstrated clusters of atypical cytotrophoblasts and syncytiotrophoblasts with mitotic figures (Figure 4) as well as a corpus luteum of pregnancy. There was no evidence of other germ cell tumors in multiple provided sections. The tumor was attached to the posterior surface of uterus. However, only serosal lining showed tumor extension. No tumoral involvement was identified in endomyometrium. Computed tomography of chest, abdomen and brain showed no abnormal findings. Since the initial impression of the clinician was more in favor of nongestational choriocarcinoma, the patient at first received 3 courses of Bleomycin/ Etoposide/ Platinum (BEP) regimen for 3 months. Due to incomplete response, the chemotherapy regimen was switched to EMA-CE. After 4 courses of new treatment, the serum β-hCG level reached to less than 5 mIU/mL. No recurrence has been identified during five years follow-up.

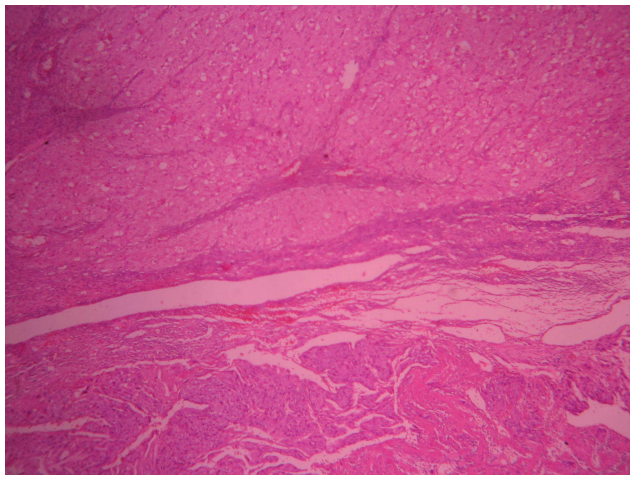

**Figure 1.** Corpus luteum of pregnancy adjacent to the tumor (40 x)

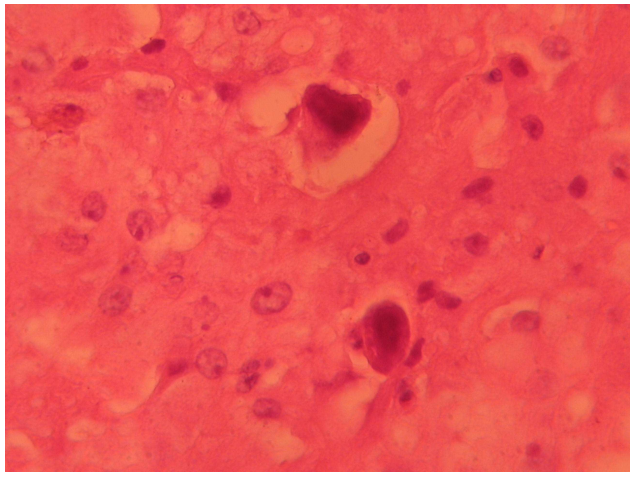

**Figure 2.** Calcifications in the corpus luteum of pregnancy (40 x)

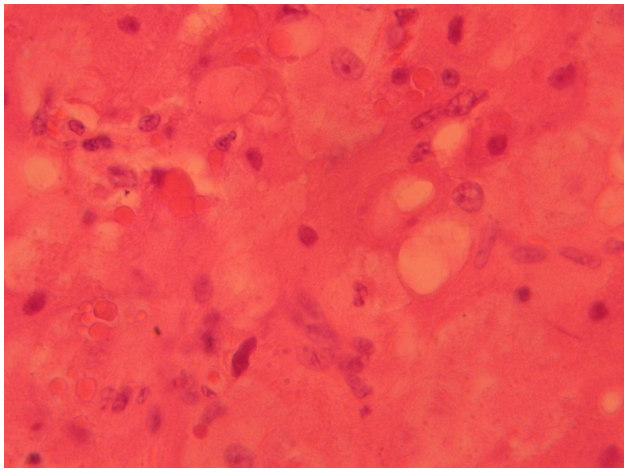

**Figure 3.** Hyaline droplets in the corpus luteum of pregnancy (400 x)

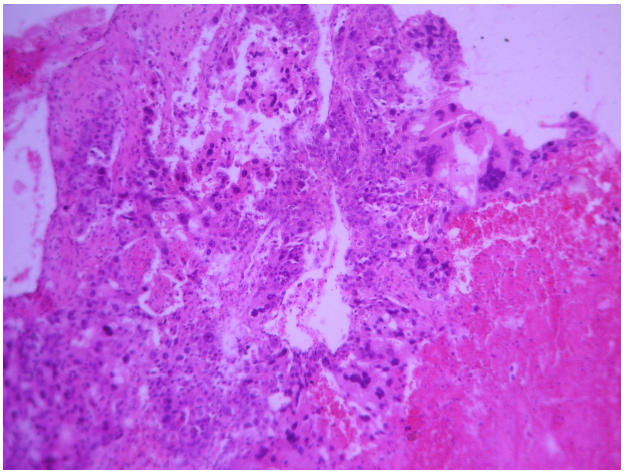

**Figure 4.** Typical admixture of bilaminar structure of cytotrophoblasts and syncytiotrophoblasts (40 x)

## Discussion

Choriocarcinoma of the ovary is a rare and aggressive tumor arising from germ cells, constituting less than 5% of all ovarian malignancies in Western countries.[1] Ovarian choriocarcinoma can be divided into three groups: as a metastatic gestational choriocarcinoma due to a regressed or occult primary gestational choriocarcinoma in other parts of the genital tract (mostly uterine corpus); as a primary gestational choriocarcinoma arising from an ectopic ovarian pregnancy, and as a nongestational germ cell tumor differentiating to trophoblastic components.[2] All types secret β-hCG. However, β-hCG levels are usually lower in nongestational variants in comparison to gestational types.[7] Monitoring of serum β-hCG can be a useful method in evaluating response to therapy. Serum β-hCG elevation leads to isosexsual pseudopuberty in premenarchal patients, while patients in reproductive age usually present with menstrual abnormalities (mainly amenorrhea).[1] Pure nongestational ovarian choriocarcinomas are extremely rare and highly malignant tumors frequently metastasizing through the lymphatics with intra-abdominal spread.[8,9] Non-gestational type choriocarcinoma involves the patients with average age of 13 and is largely confined to females under 20. The presence of a well developed corpus luteum of pregnancy adjacent to the tumor may be indicative of a gestational origin.[8] However, a search for paternal DNA in tumor allows a definite distinction between gestational and nongestational types. Tumors with gestational origin have paternal genomic structure while nongestational tumors have genomes of only maternal origin without any alleles from paternal origin.[2] Since DNA polymorphism analysis is not a generally available laboratory technique, we couldn't perform it on our cases.

Gestational ovarian choriocarcinomas have a better prognosis than their nongestational counterparts.[10,11] However some studies suggest that surgical stage of pure ovarian choriocarcinoma may be more important determinant of clinical outcome compared to being of gestational or nongestational type.[12]

The two presented patients were in the forth decade of life and showed no elements of other germ cell tumors but well developed corpus luteum of pregnancy with hyaline globules and calcifications adjacent to tumor. This finding is in favor of the gestational origin of the tumors. However the possibility of pronounced corpus luteum due to high levels of β-hCG can not be excluded.

## Conflict of Interests

Authors have no conflict of interests.

## Authors' Contributions

NIM carried out the design, histological diagnosis and writing of manuscript. SS did the histological diagnosis and writing of manuscript. NS and PRM wrote and edited the manuscript. ZE and FY did the clinical diagnosis, treatment and follow-up of patients. All authors have read and approved the content of the manuscript.

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