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NON GESTATIONAL CHORIOCARCINOMA OF THE OVARY IN A PERI-MENOPAUSAL WOMEN: A RARE CASE REPORT

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ABSTRACT

Non gestational choriocarcinoma is an extremely rare malignant germ cell tumor with incidence of less than 1% of all ovarian germ cell tumors. It carries a worse prognosis as compared to gestational choriocarcinoma (GCC). Given the rarity of the tumor, this disease is generally overlooked, which leads to delayed diagnosis and management. Here, we report a case of non-gestational ovarian choriocarcinoma in a perimenopausal women which is even more rare. A 39 yrs old P2L2 reported in emergency department with dull pain abdomen since last 20 days weeks with abdominopelvic mass. On ultrasound uterus was normal in size with two multicystic separate structures seen in pelvis suggestive of theca lutein cysts however, no intrauterine gestation sac seen. MRI Pelvis also revealed similar findings with presence of mild ascites. On further evaluation, she had high serum β-HCG and estradiol values. She was surgicaly explored in view of possibility of malignant germ cell tumor. Intraoperatively both ovarian masses were multicystic with solid areas on cut section and areas of hemorrhage and necrosis. On histopathological examination, it was confirmed as bilateral ovarian non gestational choriocarcinoma and she was started adjuvant chemotherapy as per protocol. So high index of suspicion and early intervention is the key in the management of non gestational choriocarcinoma for better prognosis.

INTRODUCTION

As per the Globocan 2018 Fact sheet, ovarian cancer was estimated to be the third most common cancer among Indian women and eighth overall, constituting 3.44% of all cancer cases[1]. It is also a leading cause of death from cancer in Indian women, with 3.34% of all cancer deaths in India in the same year. Regarding 5-year survival from ovarian cancer when diagnosed in Stage I is 94%, but only 15% diagnosed at advanced stage. Most (62%) of cases of ovarian malignancy are diagnosed at stage 3 and 4 when 5 year survival rate is only 28% [2].

Among ovarian tumors, tumor of epithelial origin are commonest. Tumors may arise from sex-cord stromal cells, mesenchymal cells and germ cells. Germ cell tumors commonly occur in the first two decades of life, but can also be seen in any age group[3]. Choriocarcinomas are rarest germ cell tumors, they can be gestational and non gestational. Non gestational choriocarcinomas differentiate toward trophoblastic structures. Only a few cases have been reported worldwide.

The prognosis of patients with choriocarcinoma of ovary is unfavorable, hence early diagnosis and early initiation of chemotherapy becomes all the more important. Due to vivid clinical presentation and less literature, diagnosis and treatment of choriocarcinoma of ovary is challenging. We are reporting a case of non gestational choriocarcinoma in perimenopausal age group which is very rare in this age group.

Case Report

A 39 years old P2L2 with previous two normal vaginal deliveries at term, presented in emergency department with complain of dull pain in lower abdomen on and off since last 20 days. She was stable, on per abdomen examination a mass around 24 weeks arising from the pelvis with cystic consistency, smooth surface and well defined margins and slightly tender on right side with restricted mobility and lower margins not reached. On per vaginal examination, cervix pointed forward with uterus appear anteverted and parous in size, a mass around 8×8cm filling entire pouch of douglas, cystic in consistency, slightly tender, fixed seems separate from uterus and the mass felt per abdominally. The mass felt on per abdominal examination, felt through the right and anterior fornix 15×15cm cystic in consistency, well defined, slightly tender with irregular surfaces and restricted mobility.

On ultrasound, uterus normal in size, no intrauterine gestation sac seen with two multicystic separate structures were seen in pelvis. Right sided multicystic structure with 825cc volume, measuring 7×6.5cm with peripheral vascularity and has solid area with central degeneration. Left sided multicystic lesion with similar characterstics and of 225cc volume. Ovaries were not seen separate from these multicystic lesions (figure 1). Possibility of bilateral theca lutein cysts was kept. A provisional diagnosis of gestational trophoblastic neoplasia was made.

Her urinary pregnancy test was positive and rest of tumor markers were as follows:

TUMOR MARKER	VALUE
Beta HCG	1,66,044mIU/ml
CA 125	163U/ml
ALPHA Fetoprotein	2.5ng/ml
LDH	407U/L
CEA	1.1ng/ml
Serum estradiol	5708pg/ml
CA19-9	7.83U/ml

WHO prognostic score[4] was 10 considering term antecedent pregnancy, intergestation gap of more than 13 months and β-hcg value more than one lakh. She was further investigated keeping the possibility of gestational choreocarcinoma.

HRCT chest was normal. MRI Pelvis revealed bilaterally enlarged ovaries. On right side, ovarian solid cystic lesion with intense post contrast enhancement and central hemorrhage, measuring 6.8×9.8×10.5cm with multiple large ovarian follicles and functional cysts in both ovaries with presence of mild ascites. Considering the high Beta-HCG and serum estradiol values possibility of malignant germ cell tumor was made and decision for exploration taken.

Considering the possibility of malignant germ cell tumor she underwent exploratory laparotomy proceed surgergical

staging with infracolic omentectomy. Intra operatively, there was approximately 200cc of hemoperitoneum, uterus was enlarged to 8 weeks size with bilateral ovarian masses. Right ovarian mass measuring 15×15cm size bosselated, angry looking with right fallopian tube stretching over the mass along with adhered omentum. There was a rent around 1×1cm on the surface with haemorrhagic fluid oozing through it. Left ovarian mass was 8×8cm with 2 rent present over it both 1×1cm of size (figure 2). On cut section uterocervical length was 9cm endometrium and myometrium were normal. Cervix was healthy. Right ovarian mass was multicystic with solid areas and with areas of hemorrhage and necrosis. Left ovarian mass was also multicystic with solid areas and hemorrhage (figure 3). Omentum was granular and nodules present at multiple sites less than 1cm size, however, peritoneum was healthy.

Postopeartive period was uneventful and she was discharged on day ten. She returned to OPD with histopathology report showing bilateral ovarian non gestational choriocarcinoma with no metastasis deposits on omentum. Patient was referred to oncology department for adjuvant chemotherapy.

DISCUSSION

Choriocarcinoma can be gestational or nongestational, according to pathogenetic origin. Gestational choriocarcinomas mostly occur in women of reproductive age, usually over 12 months following a preceding pregnancy. Gestational choriocarcinoma contains components of the paternal allele that confirm it as gestational in origin. Sixty percent of gestational trophoblastic neoplasias follow a molar pregnancy, 30% follow abortions, and 10% follow ectopic or term pregnancies.[5] In contrast, nongestational choriocarcinomas are exclusively derived from the individual in which they arise. It has been most commonly observed in the ovaries. Extra genital choriocarcinoma is similar to germ cell tumors and has a tendency to distribute along the central axis of the body. Non gestational choriocarcinoma has been reported in males, children, and postmenopausal women. However, the exact pathogenesis of these tumors remains unknown. There are several hypotheses that attempt to explain their origin: One hypothesis assumes that retained totipotent germ cells have an abnormal migration during embryonic development, fail to undergo apoptosis, and subsequently transform into choriocarcinoma. Another hypothesis suggests dedifferentiation or metaplasia of adult tissue cells into cancer cells.[6,7]

However, it is still debatable to diagnose whether a case is gestational or non gestational choriocarcinoma based on histopathologic features alone. Recently, DNA polymorphism analyses have been used to differentiate gestational (presence of both paternal and maternal alleles in equal amounts of tumor cells) from non gestational chorio-carcinoma (maternal allele only)[8]. Fisher et al. pioneered the use of DNA analysis to identify a nongestational choriocarcinoma that was initially diagnosed as gestational choriocarcinoma on the basis of evaluation of the clinical history and presentation[9].

Non gestational choriocarcinoma is an extremely rare malignant germ cell tumour with incidence of less than 1% of all ovarian germ cell tumors, it is prevalent among women of reproductive age. The clinical symptoms of the disease are normally non-specific and may include abdominal pain and vaginal bleeding. These symptoms mimic the presentations of other more common conditions occurring in young women, such as tubo-ovarian abscess and ectopic pregnancy.[10] Therefore, misdiagnosis is very high, as in this case, symptoms, clinical signs, and ultrasonographic findings, led to the diagnosis of ectopic pregnancy. The diagnosis is usually established post-operatively, after histopathological examination of the removed ovarian tumour. The tumour is composed of two cell lines of cytotrophoblast and syncytiotrophoblast that secrete beta-HCG, causing positive UPT results in the absence of pregnancy. The tumour has a high tendency to metastasise early via hematogenous dissemination, to the lung (80%), pelvis (20%), liver (10%), and other rare sites including gastrointestinal tract, spleen, and kidney. Therefore, prompt diagnosis and early initiation of therapy are important factors for a better prognosis.

CONCLUSION

Non gestational choriocarcinoma is a rare cancer however, despite its rarity, it must be considered as one of the differential diagnosis in women with adnexal mass and increased serum β-hcg. Early detection and appropriate management may improve the patient's outcome.

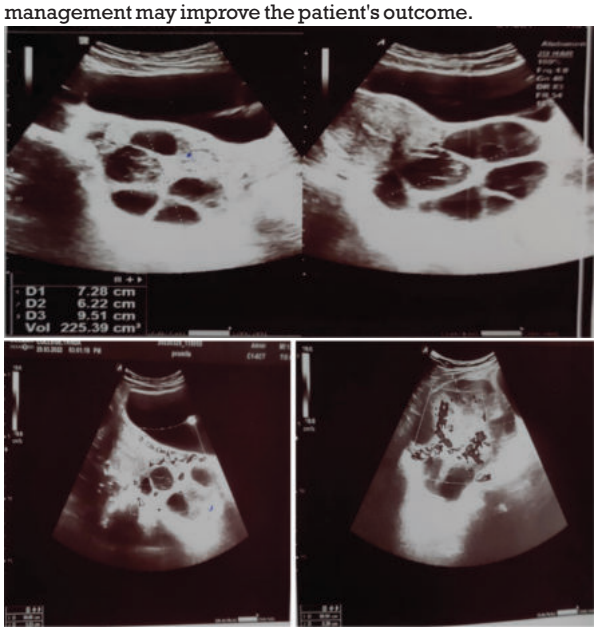

Figure 1: USG And Doppler Findings Of Bilateral Ovarian Masses

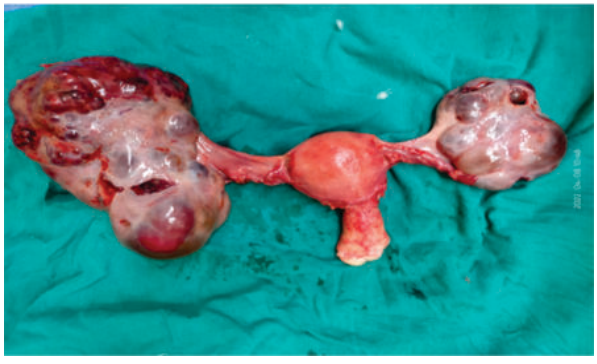

Figure 2: Uterus And Bilateral Ovarian Masses – Gross Finding

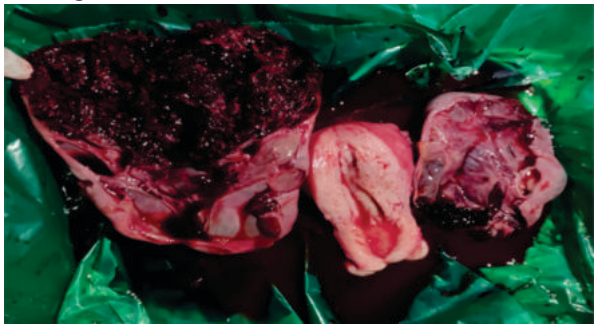

Figure 3: Uterus And Bilateral Ovarian Masses – Cut Section

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