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PURE NON-GESTATIONAL CHORIOCARCINOMA OF OVARY, A RARE GYNAECOLOGICAL EMERGENCY – A CASE REPORT AND REVIEW OF CURRENT TREATMENT OPTIONS

Oncology

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ABSTRACT

Non-Gestational Ovarian Choriocarcinoma is an extremely rare ovarian tumour, with an incidence of less than 0.6% of malignant ovarian germ cell tumours. Due to the age of presentation and increased beta HCG levels and non-specific clinical symptoms and signs, misdiagnosis is very high. We present a case of pure NGOC in an 11-year-old girl who was referred to our hospital as a case of ruptured ectopic pregnancy, as patient presented with hemoperitoneum and a positive urine pregnancy test. The patient was immediately taken for life-saving laparotomy due to ovarian mass with massive hemoperitoneum. Histopathological evaluation revealed a pure non-gestational choriocarcinoma, these rare aggressive tumours with a poor prognosis. Early diagnosis and differentiation from gestational variety can help improve prognosis. Further research is needed to find novel treatment modalities to improve survival rates.

KEYWORDS

Pure Ovarian Choriocarcinoma, Acute Abdomen, Hemoperitoneum, Non-Gestational Choriocarcinoma, Ovarian Mass, Human Chorionic Gonadotropin

INTRODUCTION

Choriocarcinoma are ovarian germ cell tumours that can be either gestational or non-gestational. Non-Gestational Ovarian Choriocarcinoma (NGOC) is an extremely rare ovarian tumour, with an incidence of less than 0.6% of malignant ovarian germ cell tumours (1). Due to the age of presentation and increased beta HCG levels, as well as nonspecific clinical symptoms and signs, misdiagnosis is very high (2). It is very important to differentiate gestational from non-gestational choriocarcinoma, as not only the prognosis but also the treatment varies. A pure non-gestational choriocarcinoma behaves very aggressively and does not respond to the usual chemotherapy regimen. We present this case report in view of its rare occurrence and also discuss the presentation and histopathological findings of this tumor.

Case Report

A 11 year old girl was referred from peripheral health centre to the emergency department of our hospital with acute abdomen and hemoperitoneum, along with a positive urine pregnancy test. A possible diagnosis of ruptured ectopic pregnancy was made at referral. The patient had attained menarche one year prior, with irregular cycles and was unsure of her last menstrual period. On examination, the patient appeared very pale, with a pulse rate of 103/min and blood pressure of 100/60 mmHg. Abdominal examination revealed lower abdominal distension up to the umbilicus, with generalized tenderness and guarding. Ultrasound of the abdomen and pelvis showed a large pelvic-abdominal complex mass of 15*12 CMS arising likely from right adnexa, with non-homogenous echoes and central large anechoic component suggestive of necrosis, along with peripheral vascularity. The uterus was displaced to the left side with normal endometrial thickness. The right kidney showed hydronephrosis due to pressure from the mass, and moderate ascites with an area of increased echogenicity in sub-hepatic region (hemoperitoneum). Ovarian tumours markers showed markedly elevated beta HCG of 173333 U/L, LDH was 349 (120-300), CA-125 was 49 (0-35), while all other ovarian tumour markers were normal. The patient's mother was informed about the likelihood of an ovarian pathology, and the patient was taken for life-saving emergency laparotomy. Intra-operatively, there was a large highly vascular right adnexal mass measuring 20*20 CMS, adherent to the posterior aspect of uterus, right pelvic sidewall, and sigmoid colon, with the capsule of the mass already breached. A right-sided salpingo-oophorectomy was performed, with an estimated blood loss of 2 Liters. Patient was adequately resuscitated and shifted to high-dependency unit. The gross ovarian specimen measured 14 cm at its maximum dimension and was congested. The capsular aspect was bosselated and breached at multiple sites. Microscopic examination revealed a choriocarcinoma. The tumour comprised atypical cytotrophoblasts displaying amphophilic cytoplasm and pleomorphic nuclei with prominent nucleoli, as well as multinucleated syncytiotrophoblasts (figure-1). Extensive areas of haemorrhage (figure-2), necrosis, and frequent atypical mitotic figures were noted. The PD-L1 test performed at a tertiary oncology centre was positive, with 95% of tumour cells showing a positive immune reaction. Post–operative recovery was good, and the patient was discharged on 4th post-operative day. She was referred to the oncology centre for further management, where she was found to have stage 4 choriocarcinoma. She was started on chemotherapy, but succumbed to life within a year from diagnosis.

DISCUSSION

Choriocarcinoma are ovarian germ cell tumours that can be either gestational or non-gestational. Non-Gestational Ovarian Choriocarcinoma (NGOC) is an extremely rare ovarian tumour, with an incidence of less than 0.6% of malignant ovarian germ cell tumours (1). NGOC is frequently associated with other germ cell tumours such as immature teratoma, endodermal sinus tumour, embryonal carcinoma and dysgerminoma(3). Pure NGOC are much rare than mixed variety tumours, with an incidence rate of $1/(3.8 \times 10^8)$ (4). Edward et al in their in their peer-reviewed literature done in 2023 reported only 57 cases since 1995 (5).

Generally, NGOC has been clinically diagnosed in female patients who are sexually immature, unable to conceive, or have never had sexual intercourse (6). In our case, the patient was 11 years old. Ovarian germ cell tumours commonly occur in the first two decades of life but can also be seen in any age group (7).

Due to the age of presentation and increased beta HCG levels, as well as non-specific clinical and para clinical symptoms and signs, misdiagnosis of this rare tumour is very high (2). In our case, the patient was referred as a case of ruptured ectopic. In a literature review done by Ao Xue et al, 10 of the 35 cases were misdiagnosed, of which 7 were misdiagnosed as ectopic pregnancies (8). In general, HCG in ectopic pregnancies rarely exceeds 10 kU/L, and the probability of HCG levels exceeding 40 kU/L during ectopic pregnancies (excluding live foetuses) is extremely limited (9). With levels exceeding 40KU/L, a differential of choriocarcinoma with ectopic should be considered. In our case, pt had a beta HCG of 1,73,333 U/L at presentation. Beta HCG is elevated in all cases, and few cases have elevated LDH and CA-125; in our case, the latter two were marginally elevated.

The clinical manifestation of this malignancy is non-specific and

include abdominal pain, vaginal bleeding, pelvic masses, and high beta HCG levels. Some cases may exhibit endocrine abnormalities and precocious puberty. Some may experience irregular cycles, as seen in our patient. These lesions are often haemorrhagic and can lead to catastrophic bleeding, our patient experienced a blood loss of 2 Liters at time of presentation, requiring aggressive fluid resuscitation.

It is very important to differentiate gestational from non-gestational choriocarcinoma, as not only the prognosis but also the treatment differs. The diagnostic criteria of non-gestational choriocarcinoma were first described by Saito and colleagues as follows: absence of disease in the uterine cavity, pathologic confirmation of the disease, exclusion of molar pregnancy, and exclusion of coexistence of intrauterine pregnancy (10). However, these standards may fail to differentiate NGCO arising from ectopic pregnancy. DNA analysis and presence of β2-microglobulin (BMG) antibody can help in differentiating NGOC from GOC. The presence of paternal DNA favours the gestational type, but DNA analysis is expensive, making it difficult to offer to everyone. Some researchers have focused on identifying other strategies to distinguish between these two types of trophoblastic tumours. Hayashi et al, observed that the tumour cell β2-microglobulin (BMG) antibody was histochemical positive (11). Another way to differentiate is by the presence of other germ-cell tumours. Given the mechanism of tissue origin, NGOC is often accompanied by other germ-cell tumours, such as immature teratoma, endodermal sinus tumour, and dysgerminoma (8). Our patient did not have any other germ-cell component. The points in favour of pure NGOC in our patient were the age of presentation, very high beta HCG level, poor prognosis and poor response to chemotherapy.

Generally, the treatment involves surgery along with chemotherapy. NGCO that develops as a component of mixed-type germ cell tumours, seems to show a favourable prognosis following surgery and BEP regimen (12). When it is NGCO of pure type, the tumour behaves more aggressively (13) with a poorer prognosis, as seen in our patient. Our patient was positive for PD-L1 marker. PD-L1 is a sensitive but nonspecific marker for trophoblast and related tumours. The frequent strong PD-L1 expression suggests that immune checkpoint blockade could be a promising approach in treating trophoblastic tumours, merits further investigation (14). Finding new pathological and immunological ways to differentiate NGOC from GOC can help is early diagnosis and discovering novel treatment modalities, which can improve the prognosis.

CONCLUSION

We present a case of pure NGOC in an 11-year-old girl, which is a rare tumour with a poor prognosis. Early diagnosis and differentiation from gestational variety can help improve prognosis. Still research is needed to find novel treatment modalities to improve survival rates.

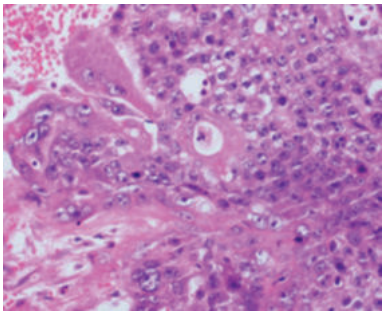

Figure-1: 40x Atypical Cytotrophoblasts and Syncitiotrophoblasts with Increased Mitotic Activity.

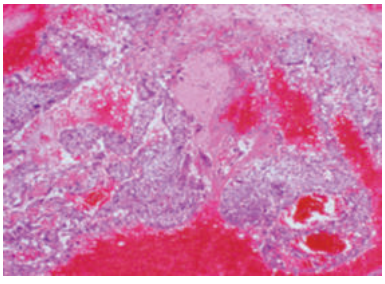

Figure-2: 10x Solid Sheets of Atypical Syncitiotrophoblasts, Cytotrophoblasts Associated with Necrosis and Hemorrhage

Abbreviations: NGOC - Non-Gestational Ovarian Choriocarcinoma, GOC – Gestational Ovarian Choriocarcinoma, HCG – Human Chorionic Gonadotropin, LDH – lactate dehydrogenase , BMG – beta 2 microglobulin, BEP – Bleomycin Etoposide Paclitaxel, PD-L1 – Programmed Death Ligand 1

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