

Case Report

Paediatric Pure Non-gestational Choriocarcinoma of ovary: A case report

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Abstract:

Choriocarcinoma of the ovary is a rare germ cell tumor. Pure non-gestational choriocarcinoma of ovary is an extremely rare occurrence. Here we present a case of 9 year old girl with pure non-gestational choriocarcinoma of the ovary which is the youngest case in literature. She was treated with multimodality therapy of complete cytoreductive surgery followed by adjuvant chemotherapy. Prognosis in these cases remains to be dismal.

Introduction:

Choriocarcinoma is a rare germ cell tumour of ovary which is more common in younger females and is usually associated with gestation. Non-gestational choriocarcinoma (NGCC) is extremely rare. We encountered a 9 year old girl diagnosed with non-gestational choriocarcinoma. This is the youngest case of NGCC reported in the literature.

Case Report:

A 9 year old girl was brought by her parents with the complaints of on and off pain in abdomen which was vague and dull aching. It was associated with constipation and loss of appetite. Her per abdominal examination revealed a mass of size 10 x 8 centimeter (cm) in the hypogastric region extending into the umbilical region. It was firm in consistency, non tender and with restricted mobility. Her routine hematological and biochemical investigations were normal. Her serum Beta Human Chorionic Gonadotropin (B-hCG) was

2,86,600 mIU/ml, Alfa Feto Protein (AFP) was 5.0 ng/ml and CA 125 was 32.87 U/ml.

Her Contrast Enhanced Computerised Tomography (CECT) abdomen and pelvis showed 12.9 x 11 x 12.3 cm lobulated, mixed density heterogeneously enhancing mass with non-enhancing cystic areas within in the pelvis extending into the lower abdomen. There were specks of calcification noted within the lesion and it was causing extrinsic mass effect over adjacent small and large bowel loops. Both the ovaries were not seen separately from the mass. There was no free fluid in the abdomen and no retroperitoneal lymphadenopathy.

On exploration, she was found to have a large pelvic mass arising from the left ovary. Right ovary had a multiloculated cyst. The mass was infiltrating posterior wall of the uterus. (Figure 1) There was ascitis but retroperitoneal and pelvic nodes were not enlarged. Total abdominal hysterectomy with bilateral salphingo-oophorectomy (TAH with BSO)

and omentectomy with retroperitoneal lymph node sampling was done.

Histopathology report showed a partially encased biphasic tumour which was admixture of cytotrophoblasts and syncytiotrophoblasts. There were areas of haemorrhage and cystic degeneration. The tumor showed capsular invasion and lymphocytic infiltration. (Figure 2) The diagnosis of choriocarcinoma was confirmed. There was infiltration of the omentum and ascitic fluid cytology was positive for malignant cells. Right ovary showed multilocated cysts. Para aortic lymph nodes were free of tumour. This was choriocarcinoma ovary stage IIIB by FIGO staging. On 21st post op day her B-hCG level came down to 121 mIU/ml. She was given adjuvant chemotherapy in the form of bleomycin, epirubicin and cisplatin in four cycles three weeks apart. She responded well to the treatment but rapidly relapsed post chemotherapy when she developed multiple lung metastases. Patient defaulted for further treatment.

Discussion:

Choriocarcinoma of ovary is an extremely rare tumour associated with pregnancy. It can occur once in 3.69×10^8 pregnancies [1]. It can be a result of metastasis from uterus or primarily in ovary secondary to ovarian pregnancy. Pure primary non-gestational ovarian choriocarcinoma is still rare accounting only to less than one per cent of all ovarian tumours [2].

Most of the reported cases are in age group of 20 – 60 years. This tumour is extremely rare in paediatric age group and three cases are reported so far [3, 4, 5]. The youngest case is 9 year old girl similar to present case [3].

Pathologically gestational and non-gestational choriocarcinoma can't be distinguished. Grossly it is a haemorrhagic mass which on microscopy shows clusters of cytotrophoblasts and intermediate

trophoblasts separated by syncytiotrophoblasts. B-hCG is expressed by syncytiotrophoblastic cells on immunohistochemistry.

The only way to differentiate between the two is clinical. A pathologically confirmed choriocarcinoma of ovary without any disease in uterus is a patient who is not engaged in sexual activity or is unable to perform sexual activity is labelled 'pure non-gestational' [6]

Molecular analysis like DNA polymorphism is a reliable method to distinguish between the gestational and non-gestational types. There are multiple case reports where this method is used to find out the origin of the choriocarcinoma [7]. Clinically the patient presents as an abdominal lump. CECT scan is a reliable tool for staging of the ovarian neoplasm. Tumour markers aid in differential diagnosis of ovarian neoplasm and B-hCG is marker specifically raised in choriocarcinoma is used for diagnosis and to monitor treatment response.

Treatment of choriocarcinoma ovary is multimodality. Surgery recommended is TAH with BSO, omentectomy and lymphadenectomy. In young patient with stage I tumour, fertility preserving surgery can be performed. In NGCC as the disease is very rare, hardly any data is available regarding ideal chemotherapy regimen. Multiagent chemotherapy is used as adjuvant treatment [8]. In our cases bleomycin, cisplatin and etoposide were used as chemotherapeutic agents. The prognosis and response to chemotherapy is dismal in non-gestational choriocarcinoma.

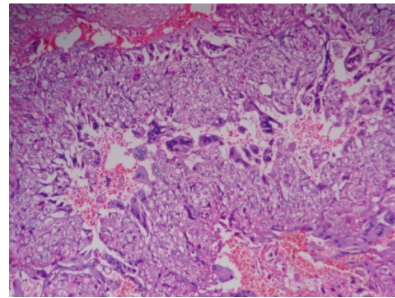
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Figure 1 – Left ovarian mass infiltrating uterus with right ovarian cyst



Figure 2 – Microscopy



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