Case Report

A Pure Nongestational Choriocarcinoma of Ovary: Sharing Experience in Sabah

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Abstract

A pure ovarian choriocarcinoma is a very rare disease which can be either pregnancy related (gestational), may not be related (non-gestational), or commonly correlated with different type of germ cell; teratocarcinoma, dysgerminoma or undifferentiated carcinoma. A pure non-gestational primary ovarian choriocarcinoma is astronomically uncommon and we recorded such condition in 14-year-old teenage girl's ovary. An abdominal operative procedure with the help of a careful histopathology examination revealed choriocarcinoma in absence of other type of germ cell element. Multiple courses of Etoposide/Methotrexate/Actinomycin-D (EMA) regime of chemotherapy were shown to be effective in this case.

Keywords: Choriocarcinoma, germ cell tumor, chemotherapy, non-gestational, ovary

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Introduction

Among the spectrums of gestational trophoblastic disease (GTD) is choriocarcinoma. It is labeled as an uncommon malignant tumour that can be either gestational or non-gestational type. The gestational type actually depends on the contents of at least a paternal chromosome whereas the non-gestational type usually comes about from the ovarian germ cell tumors (GCTs) without paternal genetic influence. We are presenting a case of pure non-gestational primary ovarian carcinoma in 14-year-old teenager with metastasis to the small bowel mesentery, appendix, bladder and pelvic lymph nodes.

Case Report

A 14-year- old, single, non-sexually active teenage girl presented to Gynecology Department after being referred in by a General Practitioner (GP) for complex abdominal mass associated with loss of appetite, compressive symptoms and occasionally low-grade fever for two-month duration.

Clinically, there was a fixed pelvic mass that corresponded to 20-weeks pregnancy with smooth mucosa per-rectally. A pelvic ultrasound indicated a massive complex mass measuring 13.4 cm x 11.2 cm with thick wall comprising cystic and solid elements including papillary projection and minimal free fluid. The uterus was empty and the left kidney was normal. Right kidney appeared hydronephrotic. An abdominal computed tomography (CT) scan indicated a huge heterogeneous mass measuring 17.6 cm x 14 cm with bilateral mild hydronephrosis of the kidney associated with 1 cm para-aortic nodes which concluded carcinoma of ovary with peritoneal and para aortic metastases. Pre-operative serum tumour markers were taken and indicated a marked increase in the level of human chorionic gonadotropins (BHCG) level 169348.30

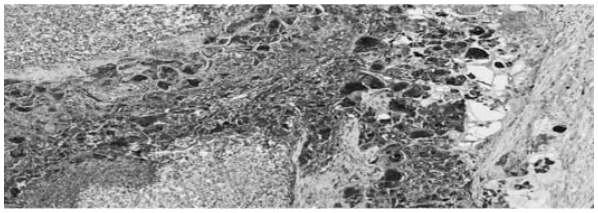

Figure 1: Negative for placental-like alkaline phosphatase (PLAP) and AFP

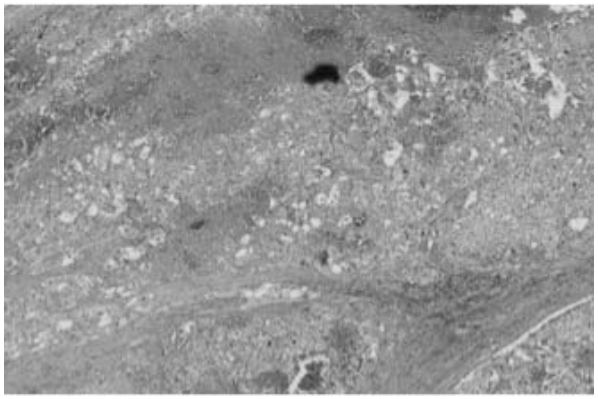

Figure 2: Positive test with B-CHG and CK7

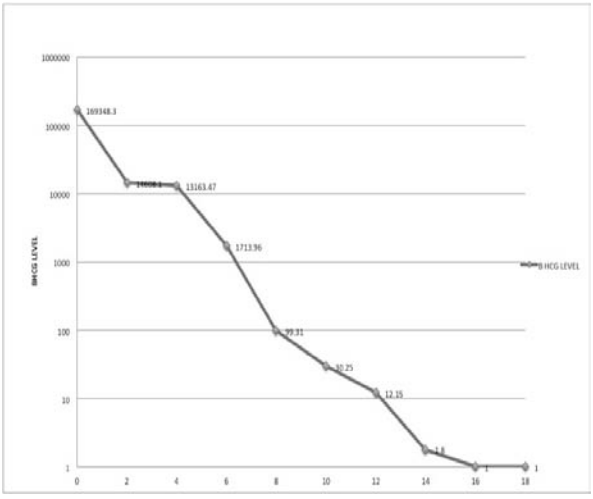

Figure 3: Decrease in HCG level after surgery and EMA regime.

(normal <1.2) and cancer antigen 125 (CA125) was 173.6 (normal 0-35 iu/l). Other markers such as carcinoembryonic antigen (CEA) were 2.43 and Alpha feto-protein (AFP) was 0.6.

She underwent a left salphingoophorectomy with infragastricomentectomy, debulking and appendicectomy. At laparotomy, a huge left ovarian tumour was seen and appeared to be densely adhered to a small bowel, sigmoid colon, pelvic peritoneal, bladder and also appendix. It was entirely removed. Optimal debulking was accomplished without any macroscopic indication of extra-ovarian malady. Pelvic lymph nodes were inspected which revealed the enlargement of the left external iliac lymph nodes (LN), omental LN and small bowel mesentery LN. The para aortic LN was not enlarged. The uterus, right ovary and tubes were normal. The Histology Pathological Examination (HPE) result showed an evidence of positive test with B-CHG and CK7 and negative for placental-like alkaline phosphatase (PLAP) and AFP consistent with pure ovarian choriocarcinoma with no association with other possible germ cell tumour. The lymphovascular invasion was observed with metastasis evidence to appendix, small bowel mesentery and left external iliac LN. In view of her extensive disease and the histological findings, she was planned for EMA (Etoposide/Methotrexate/Actinomycin-D) for first cycle and EMA plus CO (Vicristine/ Cyclophosphomide) chemotherapy regimes for second cycle.

After four weeks following these two cycles of chemotherapy, the level of HCG reduced significantly from 14608.10, post-operatively to 1718 for the first cycle and further plummeted to 99.31 following the second cycle. However, her liver enzymes were observed to be deranged after the additional of CO regimes in second chemotherapy cycle. Therefore, she was continued with only EMA regime in view of lesser side effect and responded well by evidence of reduction of HCG level during follow-up. She had been under a follow-up treatment for almost 18 months at present and had undergone up to a total of seven cycles of EMA chemotherapy. She is currently on a remission state with the latest CT scan had shown no evidence of recurrence or metastasis and latest HCG was <1.2. She was scheduled for 3-monthly outpatient reviews with HCG level monitoring.

Discussion

A pure non-gestational ovarian carcinoma (NGOC) is extremely rare as reported by Hay & Steward in 1969, Panayaton et al. in 1971 and A Oladipo et al. in 2007 (1,2,3). This condition is usually diagnosed in pre-pubertal female. In reproductive age group who is non-sexually active, it may be suspected clinically but the definitive diagnosis required a specific DNA polymorphism test to histologically determine the paternal alleles in the tumor (4,5,6,7). However, a conclusive literature review including PubMed and Medline demonstrates as few as five cases reported with NGOC diagnosed utilizing this test while others 43 cases reported only based on sexual history given the non-availability of the test.

It is important to determine this two main sub group of ovarian choriocarcinoma, as the gestational ovarian

choriocarcinoma (GOC) type is a more common condition with better chemotherapy option, good prognostic and less aggressive disease. The latter was reported about 25% and is associated with normal pregnancy and abortion and about 50% following molar pregnancy. This is because HCG increases 1000-2000 folds chances to manifest this condition. The GOC commonly metastasizes to lungs and brain. It is effectively treated with Methotrexate (MTX) based chemotherapy regime such as EMA, EMA/CO and EP /EMA. On the other hand, the NGOC is extremely rare condition. It was reported by Goswami et al. (8) that the mean age of incidence is 13.6 +/- 6.9 years old and usually presented with pelvic mass associated with nausea, vomiting and amenorrhea. There is no specific management guideline for chemotherapy option to date and usually associated with poor prognosis due to very aggressive disease. Pentheroudakis G et al. in 2004 (9) reported that they usually required a highly mylosupressive chemotherapy regime in order to control the progression of the disease and yield a better prognosis.

In this case, she was diagnosed as pure - NGOC based on the background of sexual history alone. There was no DNA polymorphism test was carried out due to non-availability of the test. We reported that this 16-year old teenage girl was successfully treated with conservative cytoreductive surgery and EMA chemotherapy regime alone that results in excellent responses. This case was considered unusual as previously reported that NGOC was usually aggressive and not responding well to MTX based chemotherapy alone. Therefore, we concluded that the mainstay treatment for this condition is by case-to-case basis depending on medical personnel experiences and patient condition during the first presentation. The type of surgical excision, thorough histological evaluation of all elements present and patient general condition also contributed to determine the choice of combination chemotherapy. Our case shows that conservative cyto-reduction type of surgery in tandem with post-operative EMA chemotherapy may be an efficacious therapeutic treatment for pure ovarian non-gestational choriocarcinoma in young girl. Taking into account that pure ovarian choriocarcinoma is aggressive with a high risk of metastasis and recurrence; we recommend a thorough follow-up treatment with serum HCG and imaging examinations as key to the success in managing this condition.

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