Choriocarcinoma with atypical presentations causing diagnostic dilemma

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Abstract

Even though choriocarcinoma commonly present with abnormal uterine bleeding and pelvic mass, various unusual and atypical presentations of choriocarcinoma have been reported which cause diagnostic dilemmas. Here we present two such cases with unusual presentations of choriocarcinoma- one with pulmonary embolism and another with vulvovaginal mass.

Key words: Metastatic choriocarcinoma; Pulmonary embolism; Vulvovaginal mass.

INTRODUCTION

Choriocarcinoma is the most aggressive form of gestational trophoblastic disease (GTD), belonging to malignant end of GTD spectrum. Choriocarcinoma can occur following molar pregnancy, ectopic pregnancy, abortion, and even normal pregnancy.1,2 It can develop anytime between five weeks and 15 years after gestation or even after menopause.1,2 Even though abnormal vaginal bleeding is known to be the most common gynaecological presentation of choriocarcinoma, clinical presentation is extremely varied and every case may be one of its kind posing diagnostic challenge.1 Here two cases of gestational choriocarcinoma with unusual characteristics are presented which resulted in diagnostic dilemma and hence delay in management.

CASE REPORT

Case 1

A 27 years Para 1+1 lady had presented to Emergency Department with history of shortness of breath, chest pain and cough with occasional haemoptysis for six months. She had history of being treated with antibiotics and anti-tuberculosis drugs but her symptoms had worsened. On examination she had crepitations in bilateral chest and an oxygen saturation of only 80%. Though chest X-ray showed only bilateral infiltrations, Contrast Enhanced Computed Tomography (CECT) chest revealed pulmonary embolus occluding main pulmonary artery along with right and left pulmonary arteries (Figure 1). Emergency pulmonary thromboembolectomy was done and the embolus (Figure 2) was sent for histopathologic examination which diagnosed it to be metastatic choriocarcinoma based upon findings of syncytiotrophoblasts, (large eosinophilic multinucleated cells with large hyperchromatic nuclei) intermixed with cytotrophoblasts (polygonal cells with distinct borders, and single irregular nuclei), along with haemorrhage and necrosis without chorionic villi. Following the receipt of histopathology report serum β-HCG (human chorionic gonadotropin) level was increased to
140,000 mIU/ml. On review of obstetric history she had one normal term vaginal delivery three years back followed by a spontaneous abortion ten months back at two months period of gestation. She was managed with combination chemotherapy (EMACO regimen) and serum β- HCG was serially followed. She had complete remission with five cycles of chemotherapy.

**Case 2**

A 50 years P4 lady with all 4 normal term vaginal deliveries and last childbirth was ten years prior. She gave history of irregular cycles for three years and menopause for one year. She came to Emergency Department with bleeding on and off for three months from a vulval mass which bled on touch. She had been transfused four units of blood in another hospital and referred to our centre as a case of vulval growth with anemia. She was pale with haemoglobin of 8gm/dl. On vulval inspection a 5*5 cm friable fungating bluish growth with infected crusts was seen arising from right labia (Figure 3a). Her urine pregnancy test was positive and serum β- HCG was 15,00,000 mIU/ml. CECT showed enhancing soft tissue density lesion in posterior myometrial wall with similar soft tissue density mass with ulceration in introitus and lower vaginal canal. Heterogenously enhancing nodules were present in bilateral lung fields. Diagnosis of metastatic choriocarcinoma was made and she received six cycles of combination chemotherapy (EMACO) and had complete remission. Her vulval mass disappeared following second cycle of chemotherapy (Figure 3b, 3c).
DISCUSSION

In Southeast Asia, 9.2 in 40,000 pregnant women and 3.3 in 40 patients with molar pregnancy subsequently develop choriocarcinoma. Various unusual presentations of choriocarcinoma have been reported—purple skin lesions, vulvovaginal swelling, intestinal perforation, features of raised intracranial pressure, cardiopulmonary manifestations. Clinicians need to have an index of suspicion of choriocarcinoma in treating women with atypical features as early diagnosis and treatment with or without metastasis have excellent prognosis. Initial high levels of serum β hCG, long duration from previous pregnancy, multiple tumor sites are high risk factors.

Choriocarcinoma presenting with pulmonary embolism as in our first case is rare. Literature review reveals only a few published case reports. Women may present with dyspnoea, chest pain, cough, haemoptysis without abnormal per vaginal bleeding. The diagnosis can be misleading and patient might be treated in the line of pneumonia or tuberculosis. Chest X-rays can show non-specific findings while CT and Magnetic Resonance Imaging may provide evidence of pulmonary embolism. In our case, choriocarcinoma was not diagnosed until histopathological report. However, chemotherapy was initiated as soon as the diagnosis was made and the patient responded well, with serum β- hCG falling to within normal. Choice of single agent versus multiple agent chemotherapy was based on the WHO prognostic score. Both our cases had scores higher than 7 and hence were started on multiple agent chemotherapy regimen-EMACO.

Metastatic choriocarcinoma may present solely as a vulvovaginal growth as in our second case and may pose diagnostic dilemma due to its misleading initial appearance. Bhattacharya, et al write about two cases of vulvovaginal choriocarcinoma, initially misdiagnosed as old infected vulvar haematoma and infected Bartholin’s cyst respectively which underwent surgery leading to massive bleeding. Hence, it is imperative to keep in mind the possibility of choriocarcinoma in cases with suspicious looking vulvovaginal mass. The diagnosis is based on history, clinical presentation along with elevated serum β- HCG titer and biopsy from the local metastatic sites is not mandatory. These masses can be treated completely with chemotherapy and any attempt at surgical removal may be catastrophic. Microsatellite polymorphism analysis is a molecular approach for distinguishing the non-gestational choriocarcinoma from the gestational one but unavailable in our setting.

These patients require lifelong follow-up even after complete remission.

CONCLUSION

A knowledge regarding the variations of different presentations of choriocarcinoma from its classic clinical presentation is, therefore, a must to any practicing clinician so that there is a high degree of suspicion allowing early diagnosis and management.

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REFERENCES