

Metastatic Epithelioid Trophoblastic Tumor: A Rare Case Report

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Received: 16 January 2011 / Accepted: 5 July 2012 / Published online: 4 October 2012
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Introduction

Epithelioid Trophoblastic Tumor (ETT) is a rare tumor recently added to the list of Gestational Trophoblastic Neoplasia (GTN) [1]. Less than 100 cases have been reported till now. Although a locally indolent tumor, metastases are reported in 25 % and deaths in 10 % of cases [2]. We report a case of metastatic ETT.

Case Details

A 26-year-old parous woman presented with profuse vaginal bleeding. Obstetric history was P1L1A4, first pregnancy was terminated for molar pregnancy, second pregnancy was a term delivery by LSCS after 3 years, third and fourth pregnancies were unconfirmed abortions, fifth

pregnancy was incomplete abortion at 8 week's gestation, underwent D&E elsewhere, following which she had persistent vaginal bleeding. One week after evacuation, she was prescribed COC pills for 6 months to control bleeding.

On examination, she was hemodynamically stable, profuse vaginal bleeding present, uterus enlarged to 12 weeks. Urine hCG was positive, and serum BhCG was 68,000 mIU/ml. Ultrasound pelvis showed enlarged uterus (11.1 × 9.7 × 9.1 cm) with a mixed echoic area in the endometrium measuring 7.4 × 6.3 cm suggestive of molar pregnancy (Fig. 1). Other abdominal organs were normal. With the diagnosis of GTD, suction evacuation and curettage were attempted, but no molar tissue was obtained. To relieve the complaint of heavy and persistent bleeding, TAH was done (ovaries retained). On further investigation, Chest X-ray showed cannon ball metastases (Fig. 2). CT scan brain showed metastases in right parietal cortex.

Pathological examination showed uterus with cervix measuring 9 × 8 × 7 cm. On cut section, a nodular gray white growth of 6 × 6 cm seen in the endometrial cavity (Fig. 3). Cut section of cervix was unremarkable. Microscopic examination showed a tumor composed of monomorphic cells with round uniform nuclei and eosinophilic to clear cytoplasm with well-defined cell borders arranged in nests and cords (Figs. 4, 5). These cells are surrounded by eosinophilic material resembling keratin. Rich vascularity is observed with areas of necrosis. Invasion up to inner half of myometrium was seen. The diagnosis was ETT and pathologically FIGO stage IV disease.

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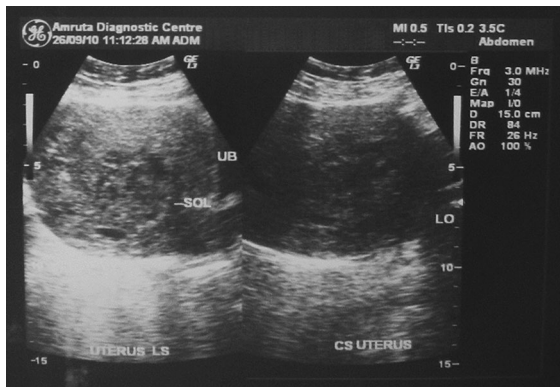


Fig. 1 USG of uterus showing a mixed echoic lesion in the endometrial cavity

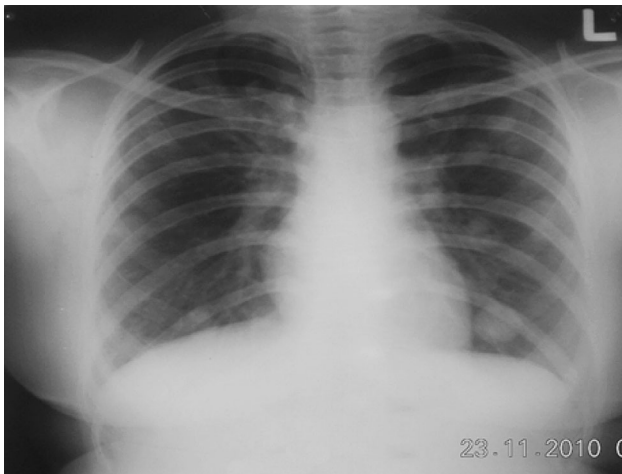


Fig. 2 Chest X-ray showing cannon ball metastases

Fig. 4 Low-power microscopic view of the tumor showing monomorphic tumor cells arranged in nests

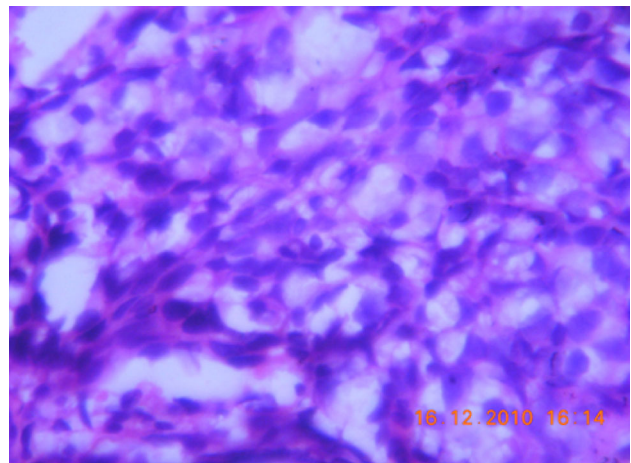


Fig. 5 High-power view of the tumor showing monomorphic tumor cells with clear cytoplasm and round nuclei



Fig. 3 Cut section of the uterus showing gray white nodular growth in the cavity

Post-operative period was uneventful. As the patient had stage IV GTN with WHO risk score of 12, adjuvant chemotherapy of EMA-CO regime was started with serum BhCG monitoring. She needed 12 cycles of chemotherapy and is in remission till date.

Discussion

ETT is a rare form of GTN seen in women of reproductive age (15–45 years) [3] developing from chorionic type of intermediate trophoblast. Vaginal bleeding is the most common presentation. In two-thirds of cases, preceding pregnancy is a term gestation. Prior pregnancy event may be remote, and the average interval between prior pregnancy and the diagnosis ranges between 1 and 18 years

(average 6.2 years). Serum BhCG is usually elevated in the lower range (average 2,500 m IU/ml). Site of the lesion in 50 % cases is in the lower uterine segment and cervix, 30 % in the uterine corpus, and rarely in lungs, small bowel, and spine [4].

Microscopy shows the characteristic trophoblastic cells arranged in monomorphic pattern of nests and cords with eosinophilic to clear cytoplasm set in a classically hyaline matrix [5]. Immunohistochemistry reveals diffuse expressions of pancytokeratin, cytokeratin 18, epithelial membrane antigen, and p63 [6].

Because of the rarity, the biologic behavior is not fully established. Unlike choriocarcinoma, ETT is chemoresistant. Control and cure of localized disease is achieved with hysterectomy [5].

In our case, history, examination, and investigations suggested GTD at the first instance. We attempted suction evacuation and curettage, but no molar tissue was obtained. Hence, emergency hysterectomy was done for heavy and persistent bleeding, as we had no facilities for angiographic embolization. Further investigations revealed GTN with lung and brain metastases. ETT was diagnosed based on the pathological examination of uterus. Choriocarcinoma was excluded by the absence of dimorphic cells with pleomorphic, hyperchromatic nuclei, and extensive hemorrhages. PSTT was excluded by the absence of the microscopic size, circumscribed borders, less cellularity with low mitotic activity, and the absence of necrosis.

As she had high risk disease, adjuvant chemotherapy of EMA-CO regime was started. At the time of hysterectomy, chemotherapy was not given as there was no sufficient evidence of neoplasia. She has received 12 cycles of chemotherapy. Initially, serum BhCG became undetectable

after three cycles, and three additional cycles were given. During follow up, serum BhCG increased to 98 mIU/ml, which became negative after three cycles of chemotherapy and further three cycles were administered to complete the course. Follow-up chest X-ray and CT Brain were found to be normal. She is in remission for the last 9 months and is still under follow up.

Case report shows that there was a significant delay of about 10 months before diagnosing the disease because of which the patient has ended up with metastases. This case highlights the need for high index of suspicion for GTN following any pregnancy event with abnormal vaginal bleeding and the need to follow up with serum B hCG.

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