Case Report

Epithelioid trophoblastic tumor: a case report of a rare trophoblastic neoplasm

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ABSTRACT

Epithelioid Trophoblastic Tumor (ETT) is a rare neoplasm of the chorionic type intermediate trophoblastic cells. It is a neoplasm of reproductive age women and usually follows a gestational event. ETT can occur at both intra uterine and extra uterine sites and can be confused with other entities such as squamous cell carcinoma, placental site nodule, placental site trophoblastic tumor etc. Hence, proper diagnosis of this tumor is necessary to avoid unnecessary, excessive treatment as surgical treatment is considered sufficient for ETT. We present a case of ETT in a 36 year old female, who came with symptoms of pain abdomen, white discharge per vaginum and a cervical mass.

Keywords: Epithelioid trophoblastic tumor, Immunohistochemistry, Intermediate trophoblastic cells

INTRODUCTION

The term Gestational Trophoblastic Disease (GTD’s) includes a spectrum of lesions composed of partial and complete hydatiform mole, placental site nodule, invasive mole, choriocarcinoma, Placental Site Trophoblastic Tumor (PSTT) and epithelioid trophoblastic tumor.¹ ² ETT was first described by Shih and Kurman in 1998 as a rare form of trophoblastic disease.³ We are presenting a case of 36 year old female who was diagnosed as having ETT on a cervical biopsy.

CASE REPORT

A 36 year old female presented with complaints of pain abdomen and profuse foul smelling discharge per vaginum since 6 months and amenorrhea of 10 months and her obstetric history was P2L2D1; her last child birth was 9 years back. All were Full term normal vaginal deliveries. First child died 4 days after birth. No other relevant personal and family history was noted.

There was a palpable mass of 14 weeks size per abdomen which was non tender, transversely mobile. Per speculum examination revealed cervical hypertrophy with an erosion on the upper lip and a whitish mass of 4x5cm near external os (Figure 1). Biopsy was taken from the cervix mass and submitted for histopathological examination.

We received a follow up hysterectomy with bilateral salpingo-oophorectomy. Uterus was measuring 16x12x8 cm. Cervix was bulky with an ulcerated nodule in the posterior lip. On cut section, the lesion was seen involving the cervix, extending upwards replacing the endometrium and invading myometrium. Myometrium was irregularly thickened with a pale gray coloured circumscribed, trabeculated firm areas (Figure 3).
Microscopy of sections from Uterus and cervix revealed uniform population of mononucleate cells resembling intermediate trophoblastic cells forming nests and cords surrounded by eosinophilic hyaline like material and areas of extensive perivascular necrosis producing a geographical pattern was noted within the tumor. Rare mitotic figure noted (Figure 4, 5, 6). Immunohistochemically the tumor was positive for CK7, EMA, p63, Inhibin and negative for HMB 45. Ki67 index was low (Figure 7, 8, 9, 10).

Figure 1: Showing a whitish mass near external os on per speculum examination.

Figure 2: MRI pelvis showing contrast enhancing mass in the endometrium.

Figure 3: Gross specimen showing ulcerative nodule on the cervix extending upwards replacing the endometrium and invading the myometrium.

Figure 4: Microscopy showing uniform population of intermediate trophoblastic cells in cords x100-H&E.

Figure 5: Showing extensive geographical perivascular necrosis x100, H&E.

Figure 6: Showing tumor invasion into myometrium x100, H&E.

Figure 7: CK positivity.
DISCUSSION

ETT was recognized as a separate entity from PSTT in the year 1998 by Shih and Kurman. The rarity of the lesion is such that, less than 100 cases of ETT have been reported till date worldwide. Histologically similar lesions were described in the lung, in patients undergoing chemotherapy for choriocarcinoma in the year 1994, and subsequent intrauterine lesions with this histology were also reported. But until 1994 these extra and intrauterine lesions were only described in patients with previous history of chemotherapy for GTD’S suggesting that these tumors were a result of either chemotherapy prolonging the course of GTD and causing these atypical growth pattern or that chemotherapy directly induced tumor cell alterations. But it was in the year 1998 that Shih and Kurman published a review of 14 cases of ETT in patients with no antecedent history of chemotherapy for GTD’s.

Uterus is the most common site of presentation (40%) whereas most common extra uterine site is lung (19%). Mean age of presentation of ETT is around 36 years (16-48 years) with very rare post-menopausal presentation. ETT mostly follows a gestational event including full term deliveries (67%), spontaneous abortions (16%) and hydatiform moles (16%) and the interval between the previous gestation and reporting of tumor is around 1 to 18 years which was consistent in our case i.e. 9 years after FTNVD.

Most cases of ETT present with abnormal vaginal bleeding and with elevated beta hCG levels, but in our patient serum beta hCG levels were low which is a rare possibility according to Palmer et al.

The histopathology of ETT is also distinct as it is a circumscribed lesion composed of uniform mononuclear trophoblastic cells arranged in nests and cords with surrounding eosinophilic material and necrotic debris. The extensive necrosis surrounding the tumor gives a characteristic geographic pattern which was very evident in our case. The typical arrangements of small blood vessels within the tumor nests were also prominent in our case.

On IHC staining the tumor is typically positive for CK, EMA, inhibin and p63 in most of the cases. Other markers which are focally expressed include beta hCG, HPL, MEL-CAM, PLAP. The Ki67 proliferation index in ETT generally is 10-25%. Similar immunohistochemical profile is noted in our case with CK7, EMA, p63, inhibin and Ki67 positivity and negative staining for HMB 45.

Appropriate diagnosis of ETT is very essential as treatment of ETT is mainly surgical and the ETT is relatively chemo resistant when compared to other GTD’s. Hence establishment of accurate diagnosis is utmost important.

CONCLUSIONS

ETT is a recently delineated type of Gestational trophoblastic tumor with a distinct histology and IHC but as it is a rare neoplasm, it is often mistaken for other entities such as PSTT and squamous cell carcinoma cervix which delays the treatment and prolongs patients suffering. Our case presented with bleeding PV and cervical mass. But with low beta-hCG levels which is a rare presentation of ETT. But with high suspicion and characteristic histological pattern we were able to
diagnose ETT even on cervix biopsy, followed by further confirmation on hysterecomy specimen with IHC.

Thus, thorough knowledge of this entity while dealing with uterine and cervical masses helps in early diagnosis and improved patient care.

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