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A case report of placental site trophoblastic tumor with maxillofacial metastasis

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Summary

The authors report a case of a placental site trophoblastic tumor (PSTT) in a 38-year-old Chinese woman eight years after a term delivery. The woman presented with abnormal vaginal discharge and underwent hysterectomy. The final diagnosis was confirmed by histological examinations in conjunction with immunohistochemical studies. However five weeks after hysterectomy, serum hCG concentration increased, which indicated recurrence of PSTT, therefore the patient was admitted and cured by four courses of EMA/CO. Five years later, the serum-hCG level increased again and the patient underwent a pulmonary lobectomy. The serum hCG concentration dropped and be kept to normal till a mass located in right maxillofacial was found one year later. After one year and two months , the patient received right maxillofacial hemangioma in her hometown hospital. The pathological report of PSTT is confirmed by the experts. After one cycle of EMA/EP chemotherapy, the patient died of multiple organ failure after nearly 8-year-long fight with PSTT.

Key words: Placental site trophoblastic tumor (PSTT); Diagnosis; Treatment; Fowllow-Up; Prognostic factor

Introduction

The authors report a case of a placental site trophoblastic tumor (PSTT) presented with right maxillofacial metastasis.

Case Report

A 38-year-old woman, gravida 1, para 1 (vaginal production), presented to her gynecologist complaining of vaginal bleeding, who had a term delivery eight years prior. B-ultrasound identified a few suspicious masses limited to the myometrium of the uterus. The patient underwent hysterectomy, and the pathological report was suggestive of PSTT, which infiltrated endometrial muscularis and surface of uterus. The immunohistochemical pattern was ambiguous. The staining showed that human placental lactogen (hPL) was intense positive and hCG partly positive. One month postoperatively, the follow-up demonstrated normal levels of serum hCG and the lungs were clear. However one week later serum hCG concentration increased to 2.4 IU/L, which surpassed the normal value 0.05 IU/L. Therefore the patient was admitted and cured by four courses of EMA/CO (etoposide, methotrexate, actinomycin D, cyclophosphamide, vincristine). Five years later, the serum-hCG level increased to 2.6 IU/L and the lung CT suggested the bubble-like change. One week later the patient underwent a pulmonary lobectomy in another hospital. The pathology slides were reviewed by the expert of pathologists of the present hospital and showed no sign of PSTT. The immunohistochemical stain was not available. The serum hCG concentration dropped and remained normal until a mass located in right maxillofacial was found one year later. After incision drainage and anti-infection treatment, the mass shrunk but did not disappear completely. One year later the tumor grew quickly with persistently elevated levels of β-hCG up to 102.9 IU/L. Meanwhile CT scan of lungs showed pulmonary bulla again, The authors concluded that the case might be the recurrence of PSTT eight years prior and therefore proposed more chemotherapy, but the patient refused the treatment plan. Two months later, the patient received right maxillofacial hemangioma in her hometown hospital. The pathological report confirmed that the tumor consisted of a population of intermediate trophoblast and implied extensive necrosis, hemorrhage, and vascular invasion was observed in right salivary gland tissue cells. The immunohistochemical staining of hPL was partly positive, the staining of hCG, HER-1, and placental alkaline phosphatase (PLAP) were negative. Ki-67 was expressed less than 10% of the tumor cells. Finally, the staining of CKAE1/AE3 was positive. The pathological diagnosis of PSTT was confirmed by the experts. One week postoperatively, the patient was admitted and received EMA/EP (etoposide, cisplatinum/etoposide, methotrexate, actinomycin) chemotherapy in the present hospital because of high β-hCG (60.1I U/L). After one cycle of EMA/EP chemotherapy, CT revealed distant spread to left frontal lobe of brain and abdominal retroperitoneal lymph nodes. The patient died of multiple organ failure after an eight-year-long fight with PSTT.

Discussion

PSTT is a tumor of intermediate trophoblast, and accounts for 1-2% gestational trophoblastic neoplasia [1]. PSTT was first recognised as a separate entity to other GTDs in 1976. This atypical variant of GTD is associated with a 20% mortality rate, and was re-named as PSTT in 1981.The nomenclature was revised because of the apparent late-onset aggressive nature of the disease,

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its unpredictable malignant potential, and its relative resistance to standard trophoblastic disease chemotherapy [2]. Placental-site trophoblastic tumours can occur following a full-term delivery or miscarriage, and less commonly arise following a hydatidiform molar pregnancy [3]. It derives from intermediate trophoblastic cells which infiltrate both myometrium and blood vessels [4]

Serum β-hCG is a less sensitive tumor marker in PSTT, but in the present case, each time before the patient developed a new metastatic neoplasm, there was always an elevated hCG, perhaps not so high. The serum-hCG in PSTT may be mildly elevated or just at normal level, which does not assist in the diagnosis, but is helpful at follow-up because late recurrences are more common in PSTT. In view of this, follow-up with clinical evaluation, as well as serological examination, are necessary for at least five years.

PSTT is not very sensitive to chemotherapy. Surgical resection is the main therapy. The treatment options after operation including observation *vs.* adjuvant chemotherapy were discussed. However, in the present authors' experience, patients can respond to chemotherapy with relapse of PSTT. A recent analysis of a UK PSTT series has shown that the most important and only factor to remain significant on multivariate analysis is the duration between the antecedent pregnancy and the clinical presentation. Thus 98% of women were cured if they presented within four years, whereas all 13 women presenting beyond this time eventually succumbed to their disease, regardless of disease stage or β-hCG levels [5]. The present case's duration between the antecedent pregnancy and the clinical presentation was eight years. Therefore it is suggested that the patient with the same factor should accept surgery with adjuvant chemotherapy, regardless of disease stage which can most likely arrest the late metastasis of PSTT. Adjuvant chemotherapy like EMA-CO or EMA-EP (etoposide, methotrexate, actinomycin, and cisplatin) is a good option.

Notably, histologic morphology is frequently equivocal for PSTTs, with diagnosis depending upon immunohistochemical staining [6]. hPL is highly expressed in the trophoblasts of placental site trophoblastic tumors and exaggerated placental site (EPS), but minimally expressed in epithelioid trophoblastic tumors (ETT) and placental site nodules (PSN). Ki-67 can be used to differentiate EPS from PSTT. Lastly, β-hCG is used to identify choriocarcinoma. In this case, the tumor stained partly positive for hPL and scattered positive for pKi-67 with β-hCG negative These results are more consistent with PSTT.

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