

A Placental Site Trophoblastic Tumor Complicated with Arteriovenous Malformation: A Case Report

Tomoko Nakamura^{1*}, Akira Iwase¹, Chiharu Ishida², Sachiko Takikawa², Maki Goto² and Fumitaka Kikkawa²

¹Department of Maternal and Perinatal Medicine, Nagoya University Hospital, Japan

²Department of Obstetrics and Gynecology, Nagoya University Hospital, Japan

Abstract

A placental site trophoblastic tumor requires care in management and a prompt diagnosis. Placental site trophoblastic tumor is a rare type of gestational trophoblastic disease and displays non-specific presentations similar to those of uterine arteriovenous malformation and those of non-neoplastic gestational trophoblastic diseases such as placental polypoid tumor. Diagnosis from biopsy is extremely rare, and a delay in diagnosis can result in a poor prognosis for patients with placental site trophoblastic tumor. Although patients with placental site trophoblastic tumor, uterine arteriovenous malformation, and placental polypoid tumor are all commonly in their reproductive years, most patients with uterine arteriovenous malformation and placental polypoid tumor can have their fertility preserved with transcatheter arterial embolism treatment. However, hysterectomy is the primary choice of treatment, which should be promptly performed when placental site trophoblastic tumor is suspected. We report a patient presenting with a hypervascular uterine tumor with added complications of uterine arteriovenous malformation. Transcatheter arterial embolism was performed in attempt to manage arteriovenous malformation and to preserve fertility, but the treatment was incomplete and only temporary. This raised the suspicion of placental site trophoblastic tumor, which was confirmed by hysterectomy. Incomplete and temporary success with transcatheter arterial embolism may suggest placental site trophoblastic tumor.

Keywords: Placental site trophoblastic tumor; Arteriovenous malformation; Transcatheter arterial embolism

Introduction

Placental Site Trophoblastic Tumor (PSTT) is a rare type of gestational trophoblastic tumor arising from implantation-site intermediate trophoblastic cells. Patients with PSTT commonly present with non-specific symptoms such as abnormal vaginal bleeding, amenorrhea, and a low serum level of Human Chorionic Gonadotropin (HCG) [1]. PSTT may occur months to years after any pregnancy regardless of its outcome, making it difficult to diagnose from clinical findings [2]. Imaging studies show a variety of presentations ranging from a solid to cystic tumor, though hyper vascularity is a common feature of PSTT [3-5]. Metastasis is reported in 16-54% of patients at initial diagnosis and usually results in very high morbidity rates [2,6,7]. The disease is relatively chemo resistant; therefore, hysterectomy is the primary choice of treatment [7,8] Uterine Arteriovenous Malformations (AVM) and non-neoplastic gestational trophoblastic diseases such as placental polypoid tumors show similar presentations to those of PSTT. This similarity makes it difficult to differentiate between PSTT and others [9,10]. However, fertility can be preserved in patients with uterine AVM or placental polypoid tumors. Although some of these patients experience massive hemorrhage, the latter can be controlled with Transcatheter Arterial Embolism (TAE) and a subsequent Transcervical Resection (TCR) [10-14]. PSTT, uterine AVM, and placental polypoid tumors all commonly affect patients typically in their reproductive years, therefore suggesting a requirement for extensive care in fertility management. Here, we report of a patient suspected of having a gestational trophoblastic disease complicated with AVM. TAE was unsuccessful, and hysterectomy revealed PSTT. The aim of this article is to report that the abundant uterine blood flow and the gradual re-enlargement of the AVM after TAE may have been the result of the tumorigenic character of PSTT. Careful consideration of such pre-operative findings may lead to a precise management of patients suspected of having PSTT.

Case

A 33-year-old gravida 1 Para 1 woman presented with abnormal

vaginal bleeding. Her previous pregnancy resulted in a term-normal vaginal delivery of a baby boy 13 months prior to presentation. Serum HCG and Human Placental Lactogen (HPL) levels were 45.2 IU/L and 0.07 µg/mL, respectively. Ultrasonography showed a 4-cm hypervascular solid tumor with tortuous vessels suggesting gestational trophoblastic disease (Figure 1A). Magnetic resonance imaging showed an ill-defined tumor, replacing the anterior wall of the uterus with a large flow-void lesion (Figure 1B). Computed tomography angiography showed the solid tumor with blood supply mainly from the right uterine artery. A 2.8 × 2.1 cm aneurysm was observed within the tumor, displaying an arteriovenous shunt (Figure 1C). Owing to the high risk of massive hemorrhage, the patient was prepared for angiography followed by TAE. Contrast study showed the right uterine artery to be markedly dilated and tortuous with instant visualization of the veins, consistent with an arteriovenous shunt (Figure 1D). A microcatheter was placed as distally as possible in the right uterine artery where thromboembolism with n-butyl 2-cyanoacrylate/lipiodol (1:3) was attempted. However, because the velocity of the arterial blood flow was too high and because the aneurysm was too large, only partial thromboembolism was achieved. Although the aneurysm was not detectable on the following day of TAE, the aneurysm reappeared on the 11th day post-TAE. TAE also did not have any effect on the dilated intrauterine vessels. While the aneurysm temporarily diminished after TAE, hysteroscopy was performed. The hypervascular lesion was still present, prohibiting mechanical curettage. Suction biopsy revealed

***Corresponding author:** Tomoko Nakamura, Department of Maternal and Perinatal Medicine, Nagoya University Hospital, Japan, Tel: +81-52-744-2261; E-mail: tomonakamura@med.nagoya-u.ac.jp

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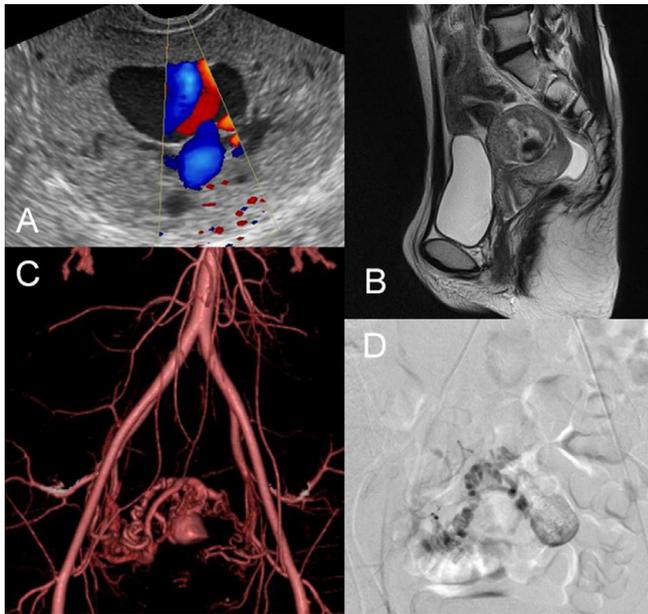


Figure 1: (A) Transvaginal color Doppler ultrasonography. Tortuous vessels and a large aneurysm are observed. (B) MRI. An ill-defined tumor replaced the anterior wall of the uterus. A large flow-void lesion can be seen within the tumor. (C) CT angiography. Blood supply came mainly from the right uterine artery. A 2.8 × 2.1 cm aneurysm was observed within the tumor, displaying an arteriovenous shunt. (D) Contrast imaging. The right uterine artery was markedly dilated and tortuous with instant visualization of the veins, consistent with an arteriovenous shunt.

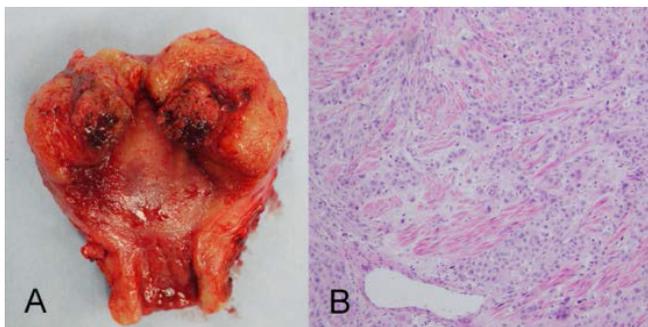


Figure 2: (A) Gross examination of the uterus. An ill-defined and localized friable tumor measuring 3 cm in diameter was noted invading the uterine corpus myometrium. A cystic portion with a thin wall was observed, which was consistent with the aneurysm noted prior to surgery. The uterine serosa was undisturbed. (B) Histopathology. The placental site trophoblastic tumor showed sheets of intermediate trophoblast cells invading the myometrium of the uterus (hematoxylin and eosin, ×100).

only a normal endometrium. Initially, the patient had wished to have her fertility preserved. However, serum HCG levels were persistently elevated, and the aneurysm continued to enlarge after the TAE. PSTT could not be ruled out; therefore, the patient agreed to total laparoscopic hysterectomy. Neither metastasis nor positive lymph nodes were noted on the CT prior to surgery.

Gross examination of the uterus revealed an ill-defined and localized friable tumor measuring 3 cm in diameter and invading the uterine corpus myometrium (Figure 2A). A cystic portion with a thin wall was observed and was consistent with the aneurysm noted prior to surgery. The uterine serosa was undisturbed. Pathological examination

showed large polygonal cells resembling implantation-site intermediate trophoblastic cells with nuclear atypia and eosinophilic cytoplasm. These cells deeply infiltrated the myometrium in an interdigitating pattern. Neither hemorrhage nor necrosis was observed, and the mitotic activity was unremarkable (Figure 2B) Immunohistochemistry demonstrated diffuse staining with hPL while scattered positivity with human chorionic gonadotropin and placental alkaline phosphatase was present. Ki-67 index was approximately 4%. At a follow-up examination 6 months after surgery, the patient was in good health with no evidence of recurrence or metastasis.

Discussion

Our patient with PSTT presented with uterine AVM, which had formed a large aneurysm in the uterine cavity. Cautious management of the AVM and careful differentiation of PSTT from placental polypoid mass was imperative. Patients with uterine AVM and/or placental polypoid masses can preserve their fertility. However, patients with PSTT cannot. An AVM is an abnormal connection between an artery and a vein, which can result in profuse bleeding. Placental polypoid masses are non-neoplastic gestational trophoblastic diseases that show similar clinical presentations to PSTT. Some patients with placental polypoid masses experience massive vaginal bleeding. However, most cases of massive vaginal bleeding resulting from AVM, and placental polypoid masses can be managed successfully with TAE [10,11,13,14].

The AVM in our patient formed a large aneurysm with dilated feeding and draining vessels. There has been only one reported case of uterine AVM associated with PSTT in the literature so far [5]. Our case is the first in which TAE was performed as an attempt to preserve the patient's fertility. The blood flow velocity of the AVM in our patient was too high, resulting in an unsuccessful TAE, which only temporarily diminished the size of the aneurysm that gradually re-enlarged. A small number of AVMs are reported to result from gestational trophoblastic diseases [12,13,15,16]. In such cases, the trophoblastic invasion of the spiral arteries is thought to destruct the uterine vasculature, resulting in an AVM [3,15]. It is unclear if PSTT has any relation with the destruction of the uterine vasculature, to have been strong enough to render TAE unsuccessful. However, as a facility with experience in TAE [11,14], we conclude that this incomplete and temporary effect of TAE as well as persistent mildly elevated HCG levels raised the possibility of PSTT.

PSTT is difficult to clinically diagnose because it is the rarest type of gestational trophoblastic disease and presents with non-specific symptoms and ultrasound findings. Delay in diagnosis can be fatal owing to its chemoresistant nature and high mortality in advanced cases [7,8] a PSTT series showing that the most important significant factor on multivariate analyses was the time from the previous pregnancy to diagnosis [17]. When clinical diagnosis was made within 48 months of the previous pregnancy, 98% of the women achieved cure, whereas all women presenting otherwise eventually succumbed to their disease regardless of disease stage or HCG levels. The survival rate of patients 48 months after diagnosis was 91-92% for those with stage I disease and 0% for those with stage II-IV diseases [7]. Diagnosis by biopsy is rare, and hysterectomy is the first choice of treatment when PSTT is suspected. PSTT commonly affects women of reproductive age, raising a diagnostic dilemma. Because delayed diagnosis of PSTT can result in poor prognosis, clinicians should consider PSTT when presented with any findings varying from the usual treatment course such as that during TAE. Hysterectomy should be performed promptly in such cases.

Here we report of a case of PSTT, which was complicated with AVM. There has been only one such case reported in the literature [5]. The AVM in our patient had such a high blood flow velocity that complete embolism by TAE was unsuccessful. Patients with uterine AVMs or placental polypoidal masses present with similar symptoms. However, these patients are usually treated with TAE and TCR, allowing them to have their fertility preserved. In our case, gradual re-enlargement of the AVM after TAE and persistent HCG levels raised the likelihood of the patient having PSTT. The abundant blood flow may have been the result of the tumorigenic character of PSTT. Although histology confirmed PSTT in our case, careful pre-operative evaluation is imperative before choosing hysterectomy for patients when PSTT is suspected.

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