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Placental site trophoblastic tumor: A rare case with an unusual presentation

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Abstract:

We present a rare case of a 23-year-old female with intraperitoneal hemorrhage from uterine rupture as an uncommon presentation of placental site trophoblastic tumor (PSTT) after spontaneous abortion. A high index of suspicion with this clinical presentation and the use of appropriate diagnostic tools to arrive at a diagnosis can go a long way in decreasing the adverse outcome of this disease. The histopathological findings and immunohistochemical staining were helpful armamentaria for the confirmation of PSTT. The patient was successfully managed with primary hysterectomy and postoperative chemotherapy.

Keywords:

Gestational trophoblastic disease, placental site trophoblastic tumor, spontaneous abortion

Introduction

Gestational trophoblastic disease (GTD) is a spectrum of diseases of abnormal trophoblastic proliferation. Gestational trophoblastic neoplasia (GTN) represents the malignant end of GTD spectrum.[1] GTN includes Invasive mole (IM), choriocarcinoma (CC), placental site trophoblastic tumor (PSTT), and epithelioid trophoblastic tumor (ETT). These tumors have a high propensity for invasion and metastasis. The national prevalence rate of CCs and other GTNs has remained almost constant at 0.55 and 0.52/1000 pregnancies, respectively, from 1990 to 1994 and 1997–2001.[2]

PSTT refers to a rare type of GTN originating from the placental implantation site and characterized by intermediate cytotrophoblasts on histology. It is the rarest subtype of gestational trophoblastic neoplasm.[3,4]

PSTT may be easily missed. Consequently, not being wary of its incidence in high-risk cases can be fatal. High index of suspicion for the possibility of GTN is the most important factor in arriving at a diagnosis of such.

Case Report

This is a case of a 23-year-old G4P2 (2022), admitted in our institution due to an acute abdomen.

The patient's first two pregnancies were unremarkable and delivered term. This was followed a year later by an incomplete abortion treated with completion curettage.

Two months prior to admission, the patient was diagnosed with another incomplete abortion and underwent completion curettage. Histopathology showed decidual tissues. However, after discharge, the patient had increasing vaginal bleeding using 4–5 fully soaked pads/day.

One month prior to admission, persistence of bleeding prompted another consult at

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the emergency room. Ultrasound revealed a globular slightly enlarged uterus with a uterine cavity completely filled up with soft tissue, mixed echoes with cystic spaces consistent with a highly decidualized endometrium without embryonic pole with increased vascularity on color flow mapping. Both ovaries were within normal size. The sonographic considerations were retained products of conception, hydropic placenta, and hydatidiform mole. B-human chorionic gonadotropin (B-hCG) level was 3,000 mIU/ml. The patient was managed with completion curettage. Whitish meaty curettings with areas of hemorrhage and no vesicular tissue were seen. She was discharged improved. Histopathological findings were suggestive of an exaggerated placental site (EPS) reaction. Immunostaining for Ki-67 was suggested.

Three weeks prior to admission, the patient had persistence of vaginal bleeding. A repeat transvaginal ultrasound showed an enlarged anteverted uterus occupied by a well-circumscribed mass measuring 5.87 cm × 5.07 cm × 4.58 cm, vascular, with cystic spaces. The endometrium was not delineated.

The patient was scheduled for hysteroscopy which showed a firm irregularly shaped, whitish, nonnecrotic, nonvascular mass occupying 60% of the endometrial cavity. A portion of the mass was taken for biopsy. Histopathology result showed a gestational trophoblastic tumor, favoring a PSTT. CC cannot be totally ruled out. Immunohistochemical staining using Ki-67 was suggested. The patient was supposed to undergo complete work-up prior to chemotherapy but was lost to follow-up.

Two weeks after the hysteroscopy, the patient was noted to have a sudden onset of right lower abdominal pain, hence admission. Her vital signs were within normal, but on abdominal examination, there was generalized tenderness and guarding noted. No masses palpated. Internal examination revealed a closed cervix with cervical motion tenderness, slightly enlarged uterus, and bilateral adnexal tenderness.

The admitting diagnosis was a uterine rupture secondary to GTN.

Upon opening the abdomen, 2 l total of hemoperitoneum was evacuated. On further exploration, the uterus was noted to be enlarged to 14-week age of gestation (AOG) size with a 1-cm thinned out point of rupture at the right postero-fundal wall with moderate bleeding and no extruding tissue [Figure 1]. Both fallopian tubes and ovaries were grossly normal. The rest of abdominopelvic organs were grossly normal. Total abdominal hysterectomy with bilateral salpingectomy was done.

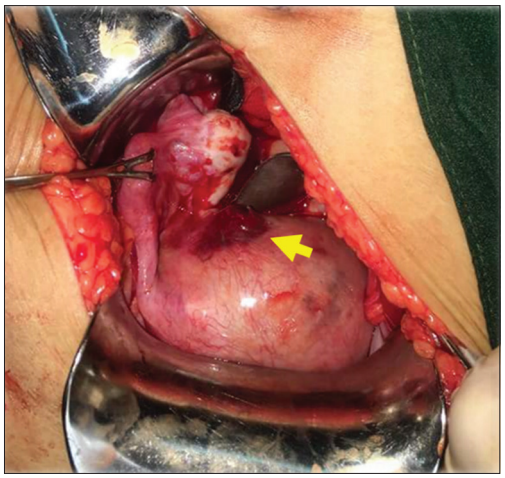

Figure 1: The uterus is enlarged to 14 weeks size with a 1-cm thinned out point of rupture (yellow arrow) at the right postero-fundal wall. 1 cm away from the utero-ovarian ligament, with moderate bleeding without extruding tissue. Bilateral fallopian tubes and ovaries are grossly normal

A pathological report revealed a uterus measuring 9.5 cm × 9.5 cm × 7.5 cm with smooth serosa. At the right upper posterior part of the uterus is a point of rupture [Figure 2]. On cut section, the uterine cavity is occupied by a firm, well-circumscribed mass measuring 6.5 cm × 5.0 cm × 5.0 cm with hemorrhagic and necrotic areas attached to the fundal wall with full-thickness myometrial involvement. The rest of the endometrium is smooth and thin (0.3 cm) [Figure 3]. At ×40 magnification, section of the serosal side shows hemorrhage at the point of rupture [Figure 4]. At ×10 magnification, sheets of polygonal and spindle-shaped trophoblastic cells are noted [Figure 5].

The histopathological diagnosis was gestational trophoblastic neoplasm favoring a PSTT, with uterine wall invasion and serosal involvement. Immunohistochemical staining using Ki-67 showed positive for more than 90% and P63 was negative. Human placental lactogen (HPL) and HCG immunohistochemical staining showed positive results.

Postsurgery, the B-hCG level was 5000 mIU/ml. The patient underwent three cycles of single-agent chemotherapy with methotrexate. The patient developed mucositis after the second cycle of chemotherapy which spontaneously resolved. The B-hCG level subsequently decreased as follows: 229 mIU/ml, 24.6 mIU/ml, and 12.6 mIU/ml.

Discussion

Kurman was the first to describe PSTT in 1976 as syncytial endometritis and eventually named it Sertoli cell pseudotumor. In 1981, Scully and Young described its morphological details and thought it to be a potentially malignant neoplasm developing from

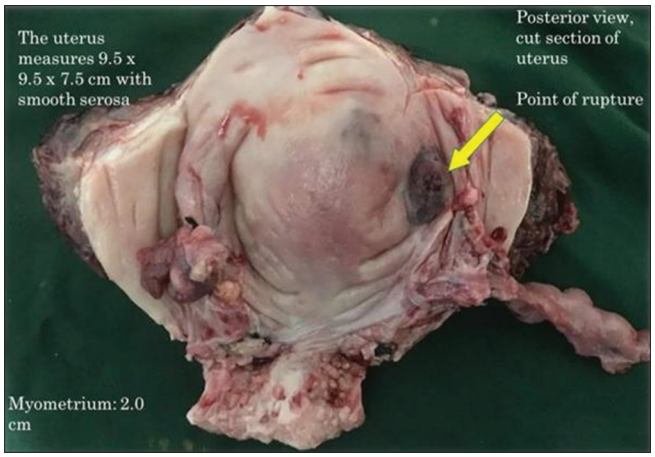

Figure 2: Posterior view of the cut section of the uterus. Gross examination revealed a uterus measuring 9.5 cm × 9.5 cm × 7.5 cm with smooth serosa and a point of rupture at the posterior wall of the uterus (yellow arrow)

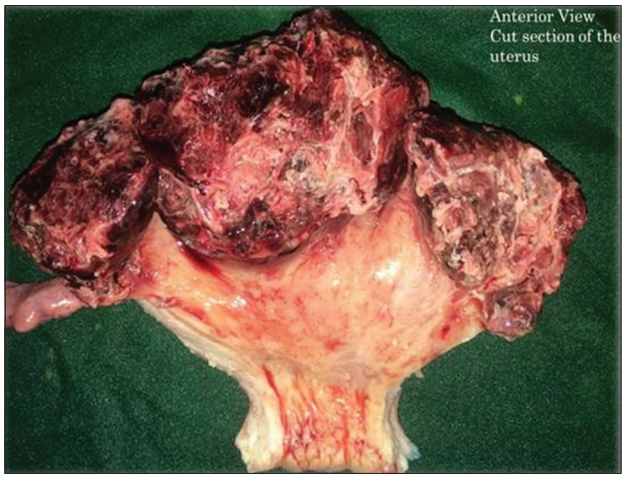

Figure 3: The uterine cavity I occupied by a firm, well-circumscribed mass measuring 6.5 cm × 5.0 cm × 5.0 cm with hemorrhagic and necrotic areas on cut section attached to the fundal wall with full-thickness involvement of the myometrium. The endometrium is smooth with no gross lesions (0.3 cm)

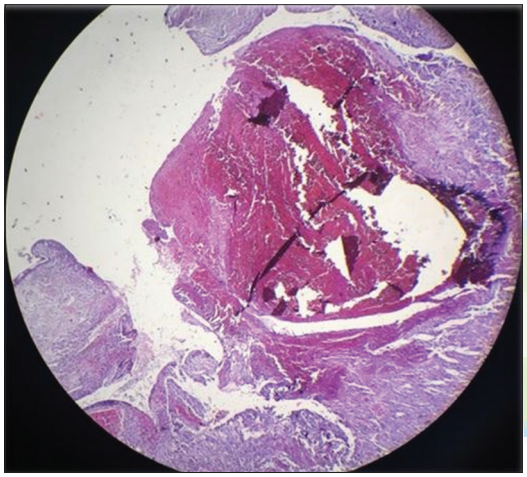

Figure 4: At ×40 magnification, section of the serosal side shows hemorrhage at the point of rupture

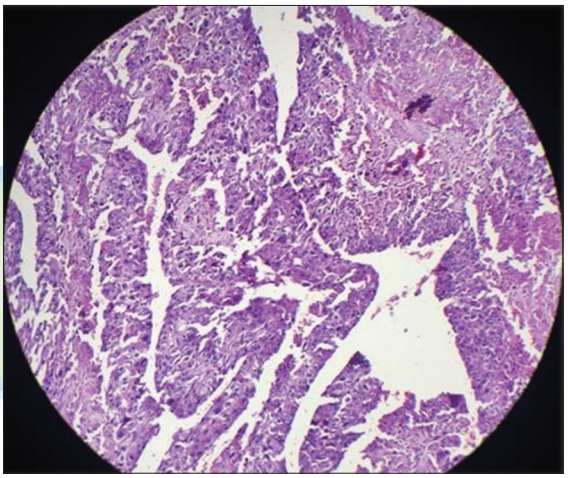

Figure 5: At ×100 magnification, sheets of polygonal and spindle-shaped trophoblastic cells are noted

intermediate trophoblasts and called it PSTT. Since this description, approximately 300 more cases have been cited in the literature.

Through a literature review, Baergen *et al*. mentioned that 57% of PSTT cases had prior term pregnancy, 17% had abortion, and 26% had prior molar pregnancies.[5] As seen in the index case, patients with PSTT usually present with irregular or profuse vaginal bleeding after a period of amenorrhea and an enlarged uterus.[3]

Although the symptomatic presentation of PSTT is similar to other types of GTN, its rarity and tendency to manifest with low-to-moderate levels of B-hCG can make diagnosing PSTT usually difficult. In our case, the diagnosis was not confirmed until after histologic evaluation with immunohistochemical staining specific for PSTT was done.

Even morphological and histopathological examination results can be confusing in cases of PSTT.

PSTT infiltrates deep into the myometrium and penetrates the uterine wall but characteristically lacks extensive hemorrhage despite common vascular involvement. It is evident in this case when the patient initially underwent hysteroscopy and a nonhemorrhagic mass was noted leading to the decision to do a biopsy. Biopsy is not recommended in cases of GTN as it can precipitate life-threatening bleeding. However, the appearance of the mass and low B-hCG level contributed to the diagnostic confusion in this case.

Microscopic evaluation in PSTT bears a resemblance to trophoblastic infiltration but often looks like masses or sheets of cells with large, atypical nuclei (Shih, 2007). PSTT contained intermediate trophoblasts and lacks cytotrophoblasts and chorionic villi unlike in CC and IM. However, the diagnosis usually depends upon the immunohistochemical staining as the histologic morphology is often confusing for PSTT.[6]

A two-step model using p63, hPL, and Ki-67 stains is applied to differentiate confirmed trophoblastic lesions.[7] p63 is a p53-like tumor suppressor. P63 differentiates PSTT and EPS from other trophoblastic lesions. A positive p63 staining is found in chorionic-type trophoblastic disease. A negative p63, as in this particular patient, favors PSTT and EPS.

A hormone produced by the placenta, hPL, is abundant in the PSTT trophoblasts as well as those in EPS. In ETT and placental site nodules it is less expressed. A positive HPL immunostain, as in our patient, leads to a possibility of PSTT and EPS.

To set PSTT apart from EPS, Ki-67 was used. Eight to twenty percent of cells in PSTT and ETT will be positive for Ki-67. The intermediate trophoblasts in EPS will be negative for Ki-67. In this case, a positive stain for Ki-67 supports a diagnosis of PSTT than EPS.

Finally, a high level of B-hCG identifies CC and IM. However, a positive hPL rules out the diagnosis.

PSTT must be diagnosed correctly because hysterectomy is the appropriate treatment as opposed to other GTN where surgery is only adjunctive. In young women, preservation of ovaries is recommended.[3] While most of the GTNs are susceptible to chemotherapy, PSTTs are relatively chemoresistant.

Conclusion

PSTT is a rare form of gestational trophoblastic neoplasm. The rarity of this disease makes diagnosis and management more difficult. Clinicians must have a high index of suspicion, aided by other diagnostic techniques like immunohistochemistry, in confirming this disease. Excision of the tumor or hysterectomy with chemotherapy is still the primary treatment for PSTT.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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