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Perivascular epithelioid cell tumors (PEComa) in pregnancy with uterine rupture and ongoing abdominal gestation: A case report

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ABSTRACT

Perivascular epithelioid cell tumors (PEComa) represent a rare family of tumors characterized by distinct histology and immunohistochemistry characteristics. Approximately one-quarter of reported cases are gynecologic in origin and associated pregnancies are rare. We report a case of PEComa in pregnancy with initial undiagnosed presentation at 18 weeks of gestation and subsequent presentation and diagnosis at 30 weeks of gestation. Abdominal pain led to the use of magnetic resonance imaging, which raised concerns about placentation abnormality and abdominal pregnancy. Exploratory laparotomy was notable for a 10 cm by 15 cm posterior uterine defect through which the placenta and amniotic sac containing the fetus were extruded. Placenta-like tissue was noted to be invading through the anterior wall of the uterus, which led to concern regarding placenta percreta. A total abdominal hysterectomy and bilateral salpingectomy were then performed, given the complete loss of normal uterine architecture. Pathology returned with findings of placenta accreta and PEComa. Indolent uterine rupture in the setting of PEComa led to an ongoing viable abdominal pregnancy. Uterine PEComa can masquerade as a placenta and lead to obstetrical complications.

1. Introduction

Perivascular epithelioid cell tumors (PEComa) refer to a recently classified group of tumors notable for their distinct histology and immunohistochemistry characteristics [1]. They are composed of epithelioid cells, which have no normal equivalent cell but do demonstrate properties of smooth muscle and melanocytes. The cells of these tumors tend to cluster around blood vessels, thus the acronym PEComa. Angiomyolipoma, pulmonary clear cell tumors, and lymphangioleiomyomatosis are among the many independently rare tumors described as PEComas because of these distinct histologic features. However, within this family of PEComas, there is variation in genetic basis and in malignant behavior. There is little insight into appropriate surveillance and treatment given this heterogeneity and relatively recent recognition of this class of tumors.

Uterine PEComas have been previously described. In a 2015 systematic review, 65 cases of gynecologic PEComas were described, accounting for approximately one-quarter of reported PEComas in the literature [2].

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2. Case report

A 38-year-old woman (gravida 1, para 0) at 30 weeks and 2 days of gestation presented to the gynecology oncology clinic for consultation regarding possible abnormal placentation. The patient's medical history was notable for a temporal lobectomy at age 18 for intractable seizures since childhood. Subsequent to this procedure, she had no further seizures. During this pregnancy, she had an episode of abdominal pain at 18 weeks of gestation, and a computerized tomography (CT) scan demonstrated hemoperitoneum, thought to be secondary to a ruptured ovarian cyst. This scan also demonstrated benign-appearing sclerotic bone lesions and a 1.8 cm right renal lesion suggestive of an angiomyolipoma. The patient was expectantly managed, but reported ongoing abdominal pain, particularly with fetal movement. Magnetic resonance imaging (MRI) was done in the early third trimester due to these symptoms with concern for abnormal placentation prompting referral to gynecology oncology clinic. The MRI scan (Fig. 1) showed no definite myometrium covering the fetus, findings suspicious for placenta percreta, and an abnormally heterogenous lower uterine segment inferior to the placenta and superior to the cervix. The patient was brought to labor and delivery for maternal and fetal monitoring. The fetal heart rate tracing was reactive without any decelerations. Because of the patient's reported ongoing severe abdominal pain as well as

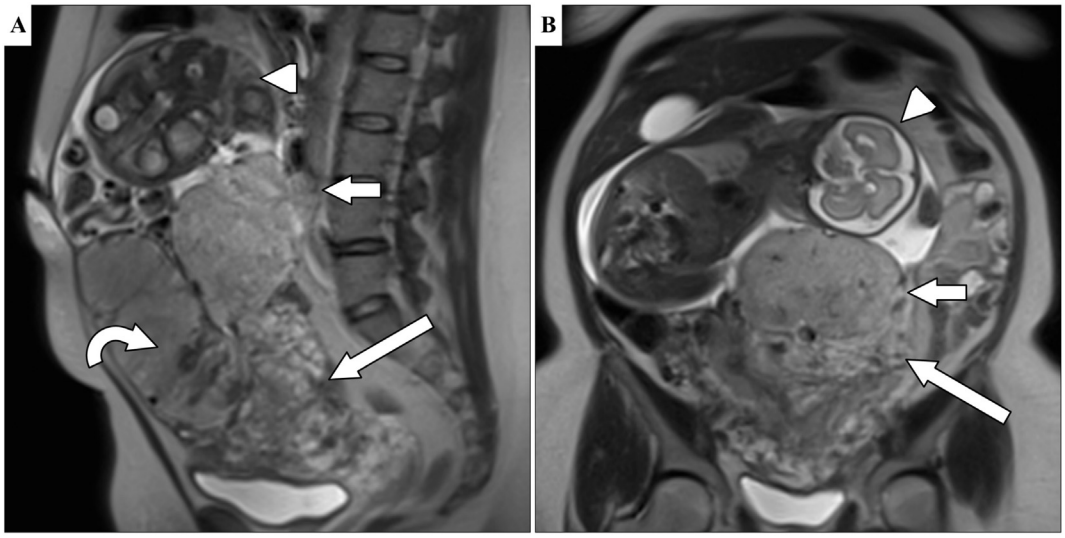

Fig. 1. A) Sagittal T2 and B) Coronal T2 magnetic resonance images demonstrate an extrauterine fetus (arrowheads), heterogeneous placenta with dark T2 bands (curved arrow in A) and bulging along its posterior margin (short arrows) suspicious for placenta accreta spectrum and abnormally heterogeneous myometrium in the lower uterine segment above the cervix (long arrows).

imaging that raised concerns regarding possible abdominal pregnancy, the recommendation was made to proceed with delivery in coordination with maternal fetal medicine, gynecology oncology and anesthesia.

After reviewing the risks, benefits and alternatives to exploratory laparotomy and preterm delivery with anticipated hysterectomy, the patient was taken to the operating room and underwent delivery of an extrauterine fetus via midline vertical incision (Fig. 2). Intraoperative findings were notable for a 10 cm by 15 cm posterior uterine defect through which the placenta and amniotic sac containing the fetus were extruded. No normal myometrial tissue was identified and the uterus appeared hypervascular with gross appearance consistent with a possible anterior placenta percreta. The neonate was delivered following intentional rupture of the intraabdominal amniotic sac with a birthweight of 1.92 kg and Apgars of 2 and 4 at 1 and 5 min.

A total abdominal hysterectomy and bilateral salpingectomy were then performed, given the complete loss of normal uterine architecture. The estimated blood loss was three liters. The patient received four units total of packed red blood cells during her hospitalization. She was discharged home on postoperative day four in a stable condition. The neonate had an uneventful course in the neonatal intensive care unit and was discharged home after 42 days.

Pathology returned with findings of placenta accreta and perivascular epithelioid cell neoplasm (PEComa) (Fig. 3). An 11 cm tumor, overlying the cervix and invading through the anterior uterine wall, was notable for infiltrative cellular lesion of epithelioid cells with moderate to marked nuclear atypia. Immunohistochemistry showed tumor cells positive for HMB45, smooth muscle actin, and desmin. Eighty percent of cells were progesterone receptor positive and 5–10% were estrogen receptor positive. These findings supported the diagnosis of PEComa.

Post-operative chest, abdomen and pelvis imaging demonstrated a 1 cm low-density lesion in the right internal iliac chain, a 2 cm low-density mass in the left cardiac ventricle, a 1 cm enhancing lesion in the left kidney and numerous 1-3 mm pulmonary nodules. The patient will undergo a PET CT with ongoing care by oncology teams. The patient consented to submission of a case report.

3. Discussion

Nine percent of gynecologic PEComas are estimated to be tuberous sclerosis complex associated, as was suspected with this patient, given her history of childhood seizures that resolved after lobectomy as well as the radiographic findings of angiomyolipoma and sclerotic bone

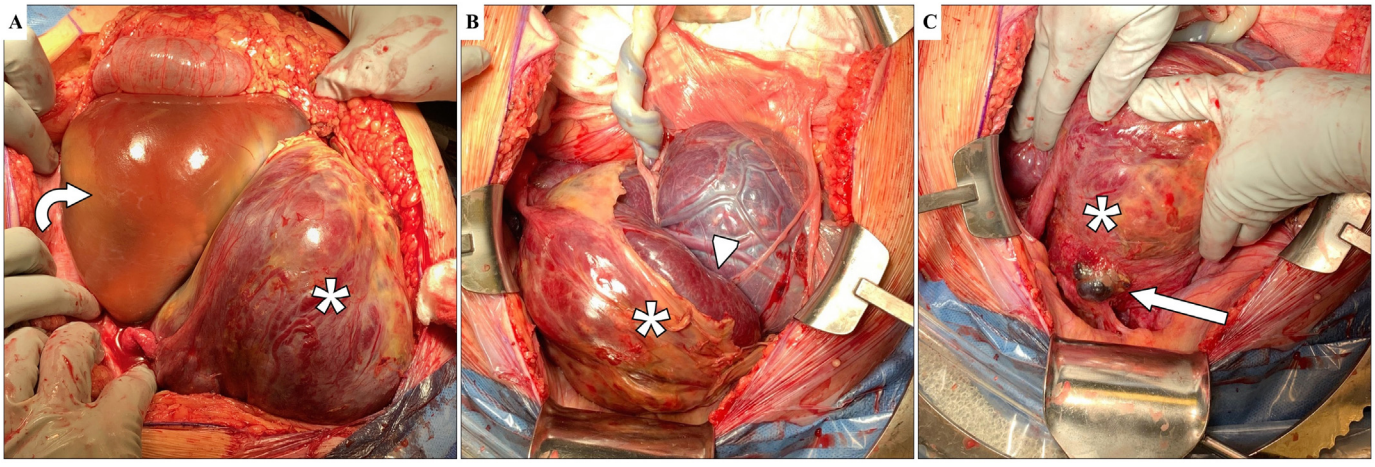

Fig. 2. Intraoperative images with extrauterine fetus (curved arrow in A), posterior uterine defect and extruded placenta (arrowhead in B), and tumor invasion site on anterior uterus (long arrow in C). Asterisk denotes uterus.

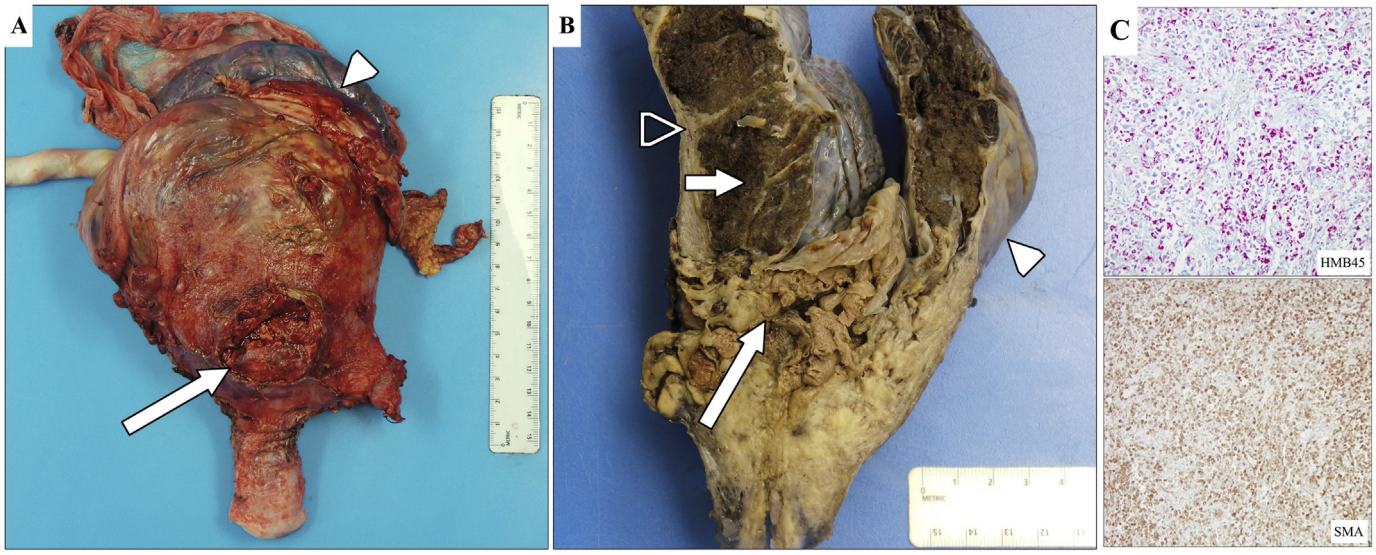

Fig. 3. Gross and microscopic images of perivascular epithelioid cell neoplasm – PEComa. A) Anterior view with extruded placenta (arrowhead) and tumor invasion site (long arrow). B) Sagittal section of uterus with placenta (short arrow), tumor (long arrow), and anterior/posterior uterine walls (arrowheads). C) Tumor cells stain positive for Human Melanoma Black-45 (HMB-45, red chromogen) and Smooth Muscle Actin (SMA, brown chromogen). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

lesions. Varied clinical presentations of these tumors are reported, but they typically are incidentally noted or discovered in the setting of chronic abdominal pain or vaginal bleeding [2]. PEComas stain positive for HMB-45, cathepsin-K and at least one muscle marker [3]. These tumors may be benign, of uncertain malignant potential or malignant. Malignant features are size >5 cm, infiltrative growth pattern, high nuclear grade cellularity, mitotic rate > 1/50 high-powered fields, necrosis, and vascular invasion [4].

There are few reported cases of uterine PEComa in pregnancy. One describes a woman who presented with spontaneous hemoperitoneum requiring exploratory laparotomy and resection of a ruptured PEComa of the round ligament [5]. Another identified a uterine PEComa incidentally at the time of cesarean section [6]. Another case report describes a PEComa masquerading as postpartum retained placenta [7]. Uterine PEComas may infiltrate into the myometrium and can be isointense to myometrium on both T1-weighted and T2-weighted images, as reported in our case. As pelvic MRI exams during pregnancy are often performed without gadolinium, antenatal diagnosis remains challenging [8,9]. Many PEComas have estrogen and progesterone receptors present that may contribute to growth in the context of hormonal changes during pregnancy. Our clinical suspicion in this case was that a previously small and asymptomatic PEComa rapidly expanded in response to pregnancy. This may have led to subacute increase in intrauterine pressure which culminated in a posterior uterine wall rupture. The placenta accreta anchored the placenta to the myometrium, maintaining fetal support. The gradual nature of this evolving pathophysiology allowed the amniotic sac to remain intact, minimizing maternal symptoms.

Contributors

Elnur Babayev contributed to clinical care of the patient, drafting sections of the manuscript, editing, and approving the final submitted version.

Kathryn E. Fay contributed to clinical care of the patient, drafting sections of the manuscript, editing, and approving the final submitted version.

Jeanne M. Horowitz contributed to clinical care of the patient and drafting sections of the manuscript.

Jeffery A. Goldstein contributed to clinical care of the patient and drafting sections of the manuscript.

Amy L. Alexander contributed to clinical care of the patient and drafting sections of the manuscript.

Anna E. Strohl contributed to clinical care of the patient and drafting sections of the manuscript.

Emily S. Miller contributed to clinical care of the patient and drafting sections of the manuscript.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient Consent

Obtained.

Provenance and Peer Review

This case report was peer reviewed.

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