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Five cases of uterine perivascular epithelioid cell tumors (PEComas) and review of literature

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Abstract

Objectives Perivascular epithelioid cell tumor (PEComa) is a rare condition and the recognition of this condition is limited. Here we report five cases of uterine PEComa to add to the limited understanding of this rare condition.

Methods Five cases from Obstetrics and Gynecology Hospital of Fudan University were diagnosed as uterine PEComas. We collected the patients' clinical and pathological data as well as their outcomes.

Results All the five cases were diagnosed post-operationally. Fertility-sparing surgery was done for the first case and had a mass resection only. She delivered a healthy boy through the cesarean section in November 2016 and neither recurrence nor metastasis was found for 71 months. Hysterectomy was done for the other four cases. Adjuvant chemotherapy was also given for case 2 and case 4. Case 2 had combined endometrial cancer, which could be associated with tuberous sclerosis complex (TSC). She was followed up for 22 months and neither recurrence nor metastasis was detected. Neither recurrence nor metastasis was found in case 3 for 33 months. However, the patient in case 4 died of multiple dissemination and multiple organs failures, 10 months after the second surgery. The patient in case 5 had the hysterectomy and left adnexal resection and in this case we had no data about her long-term outcomes.

Conclusion It is still challenging to detect and diagnose uterine PEComa clinically and no consensus or guidelines have been established regarding the treatment of this condition. More case studies are needed to enlighten the underlying mechanism and help optimize the therapies for this condition.

Keywords PEComa · Uterus · Case reports

Introduction

PEComa is a rare condition, characterized by both expressions of melanotic and myogenic markers. It occurs at any site, and most commonly in kidneys, known as angiomyolipoma (AML). It has also been reported in liver, pancreas, rectum abdomen, gynecological tract and so on. Uterine PEComa is reported to be the main type of PEComa, outside of the kidney. Few cases have been reported till now, mainly due to its rarity. Here we report five cases of PEComa of the uterus (summarized in Table 1), from Obstetrics and Gynecology Hospital of Fudan University, to try to add knowledge and clinical evidence to this rare condition.

Case 1. A 23-year-old unmarried woman who presented with left lower abdominal pain was admitted in October 2011 for 1 day. Her menstruation was regular without any history of abnormal vaginal bleeding. A gynecological examination was done and a 10 cm diameter mass was found

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Table 1 Clinical and pathological characteristics of five cases

Characteristics	Case 1	Case 2	Case 3	Case 4	Case 5
Age (year)	23	25	38	27	53
Association of TSC	No	Likely	No	No	No
Synchronous cancer	No	USC	No	No	No
Initial presentation	Left lower abdominal ache	AUB	HMB, dysmenorrhea	Pelvic mass	HMB
Tumor site/size (cm)	10	3	5	6	8
Involvement	No	Yes	Yes	Yes	No
Preoperative diagnosis	Left ovarian cyst torsion	USC, uterine fibroids	Leiomyoma with cystic degeneration	Degenerative leiomyoma	Uterine fibroids
IHC profiles	PR (+), HMB45 (+++)	HMB45 (+), vimentin (++), SMA (+++), caldesmon (+++)	HMB45 (+), vimentin (+++), SMA (+++), desmin (+++), caldesmon (+++), ER (+++), PR (+++), Ki-67 (3% +)	HMB45 (+), vimentin (±), desmin (±)	SMA (+), desmin (++), caldesmon (−), HMB45 (+++), S-100 (−), EMA (−), AE1/AE3 (−), CD99 (+++), CD10 (+), inhibin-a (−), MyoD1 (−), myoglobin (−), Ki-67 (5% +).
Treatment					
Surgery	Laparoscopic mass resection	LAVH + BSO + PLOA	Laparoscopic uterine tumor resection; LAVH	First surgery and second surgery[$]	Laparoscopic myomectomy + LSO; LAVH
Chemotherapy* (cycles)	No	3	No	3	No
Follow-up (months)	71	22	33	10[#]	Lost

TSC tuberous sclerosis complex, *IHC* immunohistochemistry, *USC* uterine serous carcinoma, *AUB* abnormal uterine bleeding, *HMB* heavy menstrual bleeding, *AML* angiomyolipoma, *LAVH* laparoscopic-assisted vaginal hysterectomy, *BSO* bilateral salpingo-oophorectomy, *LSO* left salpingo-oophorectomy, *RSO* right salpingo-oophorectomy, *PLOA* Pelvic lymphadenectomy + omentectomy + appendectomy, *PR* progesterone receptor, *HMB45* human melanoma black 45, *SMA* smooth muscle actin, *ER* estrogen receptor

[&]Abdominal mass resection

[$]Abdominal hysterectomy + RBO + right broad ligament tumor resection + part of the omentum resection + right lumbar muscle mass + abdominal mass resection + right ovarian venous ligation + right pelvic lymphadenectomy

[*]Chemotherapy protocol: ifosfamide + carboplatin + epirubicin

[#]10 months post second surgery and the patient died of distant dissemination and multiple organ failure

in the left adnexa with significant tenderness. Ultrasound revealed a 108 × 101 × 95 mm cyst on the left side of the uterus, with mixed echo and rich blood supply. The diagnosis of a cyst that may have originated from the left adnexa was given. Left ovarian cyst torsion was considered at this point. And, an emergency laparoscopic examination was performed. During the operation, a 10 cm diameter cystic mass was found at the lower part of the anterior wall of the uterus extending to the cervical isthmus, and the vesicouterine peritoneal reflection.

The mass was successfully resected. The cyst had a dark fibrous cystic wall filled with grayish fluid. Bilateral ovaries were grossly unremarkable. Intraoperative pathology reported negative for malignancy. Final postoperative pathology reported the mass as a peripheral epithelioid cell tumor (Fig. 1). This is supported by the strong immunostaining of HMB45 (+++) (Fig. 1) and PR (+). No adjuvant therapy was given post-operation. The patient got pregnant spontaneously in February 2016 and gave birth to a healthy boy through the cesarean section in November 2016. Till September 2017, neither recurrence nor metastasis was found.

Case 2. A 25-year-old married woman was admitted in August 2010 due to abnormal vaginal bleeding that lasted for 2 months. Her previous menstruation was regular and she had no history of abnormal uterine bleeding. Ultrasound indicated an intrauterine mass of the size 35 × 29 × 15 mm with cord-like blood flow and a hypoechoic area 24 × 20 × 18 mm in the uterine cavity. Tumor markers were CA125: 43.8 U/ml, CA199: 90.121 U/ml, CEA: 1.89 ng/ml, AFP:1.55 ng/ml. Hysteroscopic examination and curettage were performed, and pathology reported endometrial serous adenocarcinoma. Further, upper abdominal and pelvic MRI was done and one mass in the uterine cavity

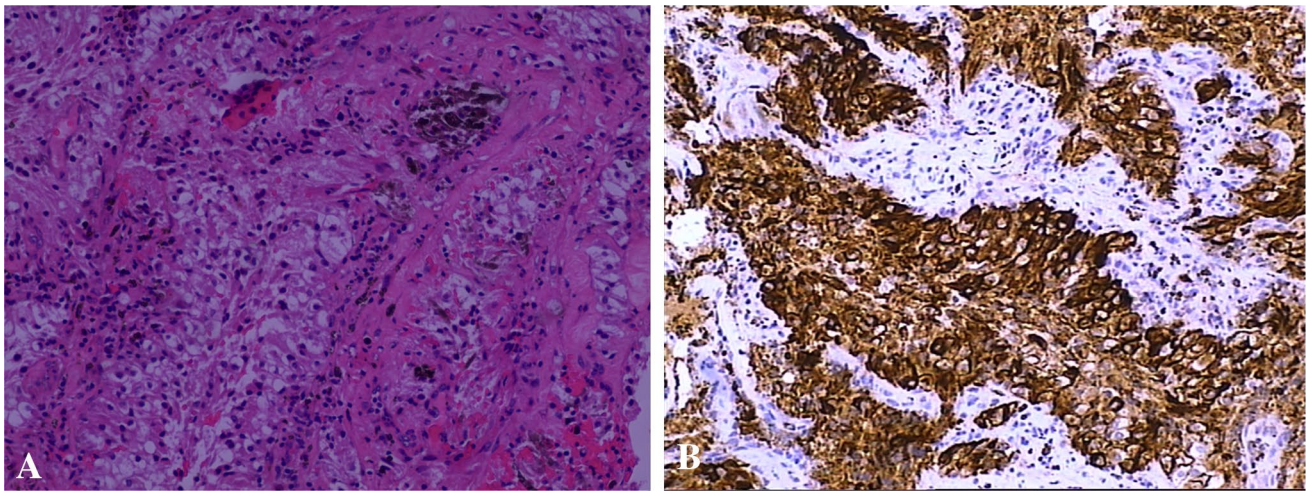

Fig. 1 Case1: **a** the tumor was composed of epithelioid cells with moderate nuclear atypia, no necrosis, no vascular invasion (H&E × 100); **b** the tumor cells were strongly positive for HMB45 staining (IHC × 100). *H&E* haematoxylin eosin, *IHC* immunohistochemistry

was found, which was close to the bottom of the uterus. A diagnosis of endometrial cancer Ib was given. And, another round mass found on the posterior wall of uterus was considered as myometrial fibroid. Besides, multiple tubercles and nodules in bilateral kidneys were found in this patient. The diagnosis of renal multiple vascular smooth muscle lipoma was given. The small nodule found in the lower lobe of the right kidney was considered as a vascular smooth muscle lipoma.

Then, laparoscopic vaginal hysterectomy plus bilateral salpingo-oophorectomy plus pelvic lymphadenectomy plus omentectomy and appendectomy was performed on this patient. During the intraoperative examination, we saw a full uterus with a slight protrusion in the posterior wall, while the bilateral adnexa appeared to be normal. Intraoperative pathologic examination was not performed for this patient.

Final pathology reported perivascular epithelial cell tumor perforating uterine subserosal surface, endometrioid adenocarcinoma grade I invading the uterine superficial myometrium, while the cervix was not involved; right obturator lymph nodes (2/5) and left iliac common lymph nodes (1/6) were positive for metastasis from the perivascular epithelioid cell tumor. Immunohistochemistry staining reported HMB45 (+), Vimentin (++), SMA (+++), and Caldesmon (+++). Three cycles of chemotherapies with ifosfamide, carboplatin and epirubicin were given This patient was followed up for 22 months and no recurrence and metastasis was detected.

Case 3. A 38-year-old married woman was admitted in September 2009 due to heavy menstrual bleeding that lasted for over 1 year accompanied with abdominal pain for 10 days. She used to have regular menstruation with no history of abnormal vaginal bleeding. In the gynecological examination, a 5 cm lump was detected on the posterior uterine wall. Ultrasound showed a hypo-echo mass of the size 52 × 44 × 38 mm on the posterior wall of the uterus and multiple areas with no echo of the size from 3 to 9 mm scattered inside the mass. The mass had abundant dotted blood supply. Ultrasound diagnosis showed a parenchymal mass in the uterus, probably a degenerated uterine fibroid. Tumor markers were also tested with CA125 at 4 U/ml, CA199 at 9.91 U/ml, CEA at 0.2 ng/ml, AFP at 1.73 ng/ml.

Laparoscopic uterine mass resection was performed. During the operation, a grayish 5 cm mass was found on the posterior wall. Part of the mass was in a pattern of honeycomb translucent. Bilateral adnexa showed no abnormality. Intraoperative pathology showed the mass as a mesenchymal tumor, probably an epithelioid leiomyoma. Final pathology reported the mass as a perivascular epithelial cell tumor. Immunohistochemistry staining reported HMB45 (+), Vimentin (+++), SMA (+++), Desmin (+++), Caldesmon (+++), ER (+++), PR (+++), Ki-67 (3% +). Then, laparoscopic vaginal hysterectomy was performed. And, the postoperative pathology revealed that the uterine myometrium close to the residual cavity had residual vascular epithelioid tumor cells and invasion of the surrounding myometrium. The remaining endometrium was in secretory phase. No further therapy was given. The patient was followed up for 33 months without recurrence and metastasis.

Case 4. A 27-year-old unmarried nulliparous young lady who presented with a pelvic mass that was discovered 2 years before was admitted in February 2006. She had no history of abnormal uterine bleeding. Gynecological examination showed a mass of the size 5 cm on the right side of the uterus. Consistently, the ultrasound showed a 58 × 62 × 38 mm mass on the right side of the uterus with abundant blood supply with PI: 0.58 and RI: 0.44. At this point, diagnosis of fibroids with degeneration on the right

broad ligament was considered. Serum tumor markers were still in normal range with CA125 at 9 U/ml, AFP at 2.53 ng/ml. The diagnosis of uterine broad ligament fibroids with degeneration was made, and abdominal uterine tumor resection was performed. Intraoperative examination showed a 6 cm diameter cystic mass on the right side of the broad ligament with no clear boundary, which was closely attached to the uterus. The cyst was filled with a pale-yellowish fluid with locally dark brown dots. Bilateral adnexa were grossly unremarkable. Intraoperative pathology reported benign smooth muscle and fibrous connective tissue, with inflammatory cells and hemorrhagic necrosis, and there was no evidence of malignancy. However, the final postoperative pathological diagnosis was (mass in the right broad ligament) perivascular epithelial cell tumor. Immunohistochemistry showed HMB45 (diffuse +), Vimentin (±), and Desmin (±).

Four months after operation, ultrasound detected a new mass near the right side of the uterus of the size $71 \times 50 \times 41$ mm with abundant blood flow. The former pathology was confirmed as perivascular epithelial cell tumor. Considering tumor recurrence, a second operation was performed. The removal of the whole uterus, the right adnexa, the right broad ligament, part of the omentum, the mass behind the right lumbar muscle and abdominal mass was done. Besides, both the right ovarian artery and vein were highly ligated and the right pelvic lymph nodes were resected. It was confirmed that the tumor recurred right in the place where the primary tumor was removed. Second pathology was consistent with the primary as perivascular epithelial cell tumor, infiltrating the right side of uterus and right adnexa, with no metastasis in the right pelvic lymph nodes. Furthermore, three cycles of chemotherapies were performed, with ifosfamide, carboplatin and epirubicin which is the same as case 2. However, the patient died of multiple metastases (liver, posterior side of the back of bladder, right psoas major and the abdominal wall) and multiple organ failure 10 months after the second operation.

Case 5. A 53-year-old married woman was admitted in December 2011 due to heavy menstrual bleeding for more than a year. She had uterine fibroids for 5 years. Ultrasound examination indicated multiple uterine fibroids and one ovarian cyst in the left adnexa. In the gynecologic examination, an 8 cm hard mass can be palpated on the left posterior side of the uterus with no tenderness. A palpable 6 cm diameter cyst was found in the left adnexa with no tenderness. Laparoscopic myomectomy with left adnexa was performed. During the operation, we saw that the uterus was of the size $100 \times 80 \times 70$ mm^3, and an 8 cm diameter intramural fibroid was found at the fundus of the uterus. Another subserosal fibroid at the size of 1 cm was seen on the anterior wall of the uterus. The left fallopian tube was thickened, twisted with edema which formed a $50 \times 40 \times 40$ mm^3 cyst, adhered to the left pelvic wall. The right adnexa was grossly unremarkable. Intraoperative pathologic examination reported (uterine mass) epithelioid cell nests with atypical nuclei were distributed diffusely in collagen interstitium, left ovarian cyst and endometrioid cyst. After notification of the situation, the patients' family asked for hysterectomy, then laparoscopic vaginal hysterectomy was performed. Postoperative pathology reported uterine local adenomyosis, endometrium in secretory phase, chronic cervicitis (uterine mass), peripheral epithelioid cell tumor (PEComa), left ovarian endometrioid cyst, and left hydrosalpinx with chronic inflammation. Immunohistochemistry of the uterine mass reported SMA (+), desmin (++), caldesmon (−), HMB45 (+++), S-100 (−), EMA (−), AE1/AE3 (−), CD99 (+++), CD10 (+), inhibin-a (−), MyoD1 (−), and myoglobin (−), Ki-67 (5% +).

Discussion

Till now, limited cases of uterine PEComas have been reported. No specific signs or symptoms can be identified for this rare condition and imaging presentations are not typical either. The diagnoses were made based on the pathological reports combined with immunohistochemistry staining for the five reported cases. Obviously, we had limited experience in diagnosing and treating this condition. Till now, only around 80 cases were reported in English literature [3]. Our experience in treating these five cases might help in recognizing and managing this condition.

For the cases reported currently, the patients' age ranged from 9 to 79 years, with the median age at 47.5 years [3]. Patients reported in this paper were of 23 years, 25 years, 38 years, 27 years and 53 years old, respectively. Most patients presented with abnormal uterine bleeding or lower abdominal pain. Ultrasound usually can detect palpable masses. The nonspecific signs and symptoms and imaging presentation make it difficult to discriminate PEComas from other uterine tumors. For our five cases, preoperative ultrasound reported two cases with leiomyoma, two cases with leiomyoma with degeneration and one case with the adnexal cyst. MRI examination was also performed in one case, but the mass was still leiomyoma. It is still challenging to preoperatively discriminate PEComas from other uterine masses.

Genetically, about 6% of uterine PEComas are associated with tuberous sclerosis complex (TSC) [1, 3]. Here, our second case also presented with suspicious renal angiomyolipoma (AML) and liver angiomyolipoma, but no further tests were done due to patient's refusal. The mass sizes ranged from 3 to 10 cm combined with multiple nodules, cystic degeneration, bleeding, and even necrosis. Mutations of TSC1 and TSC2 and translocations of TFE3 (transcription factor E3) were reported to be related with PEComas [1, 7]. TFE3 is one member of the microphthalmia transcription

factor (MiTF) gene family located in the short arm of chromosome X. TP53 pathway was also thought to be correlated with aggressive biology and unpredictable clinical behavior of pure epithelioid PEComas [1]. However, more evidence is still needed for these genetic abnormalities in PEComas.

No specific serum makers are found to be associated with PEComas, as all of the cases were diagnosed after surgery. Actually, it is not that easy to find a specific marker to differentiate PEComas from other uterine benign tumors. It is really necessary for us to identify such a marker to help us make a relatively precise diagnosis.

Almost all cases published [1–3, 5], including our five cases here, were diagnosed after surgery with pathology and immunohistochemistry. For the five cases, four patients had intraoperative pathology but all failed to make the diagnosis as PEComas. It might be hard to diagnose uterine PEComas only with fast intraoperative pathology reports, as we still lack experience in diagnosing this rare condition. Maybe accumulated clinical experience in this rare cancer can help pathologists recognize this condition faster in the future. Immunohistochemistry staining also plays an important role in diagnosing this condition. All five cases were positive for HMB45 staining. However, for literature review some cases reported were negative for HMB45 expression [3]. Besides HMB45, other two melanocytic markers, melan-A and MiTF, are also usually positive for PEComas, as well as smooth muscle markers, including actin, desmin, and caldesmon [10]. Among the markers mentioned above, HMB45 is used the most. To date, combined pathologic examination and immunohistochemical markers help us to make the diagnosis of PEComas.

Folpe et al. divided PEComas into three categories: benign, uncertain malignant potential (UMP) or malignant types based on their morphology and pathologic characteristics [4]. Schoolmeester et al. [8] made some revision to this classification and placed benign type and UMP into one category. Based on four characteristics: gross size $\geq$ 5 cm, high-grade nuclear features, necrosis, vascular invasion, or a mitotic rate greater than or one per 50 HPF, tumors with less than four features were classified as benign/UMP, or else, they are regarded as malignant. Afterwards, Niamh Conlon et al. [3] tried to make some revisions to the Folpe criteria. These criteria all greatly help us to diagnose and manage uterine PEComas. Meanwhile, more cases are still needed to help improve these criteria in the future.

Nowadays, surgery is still the treatment of choice for PEComas. Two cases of these five patients were diagnosed as leiomyoma with degeneration preoperatively; the secondary surgery was performed after primary laparoscopic mass resection. Usually, patients with uterine PEComas received a hysterectomy with or without bilateral salpingo-oophorectomy [1]. Based on our experience, patients' age, fertility requirements and their own will should be taken into consideration when we make the surgical treatment plan. If the uterine PEComas tend to be benign pathologically, mass resection is good enough and long-term follow-up is needed, as in our case 1. Eiko Yamamoto [11] reported a 24-year-old nulligravida woman who had recurrent PEComa with low malignant potential in the uterine cervix, for whom mass excision was done for twice after primary uterine mass removal and no recurrence was found for 1 year after the last surgery. For PEComa malignant potential, hysterectomy should be done. Whether to preserve the bilateral adnexa or ovaries depends on the overall evaluation of the patient. For young patients with local uterine lesion, ovaries can be preserved; otherwise, bilateral salpingo-oophorectomy is more suitable. For patients with malignant uterine PEComa, hysterectomy plus bilateral salpingo-oophorectomy is recommended. Theoretically, uterine PEComas are mesenchymal tumors, and mainly metastasize vascularly, which greatly diminishes the value of lymphadenectomy for this cancer. But interestingly lymph metastasis happened in case 2 here. Till now, there is no consensus on whether lymph nodes should be resected or biopsied. Obviously, more cases and more experience are needed for the guiding the decision making of this condition.

Besides surgery, chemotherapy and radiotherapy were applied in some reported cases [1] while currently there are no guidelines for chemotherapy in uterine PEComas. For here, patients in case 2 and case 4 received the same chemotherapy treatment plan using ifosfamide, carboplatin and epirubicin, but had different treatment outcomes. Limited chemotherapy protocols concerning uterine PEComas were published. Based on Jean IS' report [6], chemotherapies based on ifosfamide were usually applied in PEComa patients, which result in a good response. We also applied similar protocols based in ifosfamide for case 2 and case 4. Still, more researches in this gray area are in urgent need. Besides chemotherapies, some researchers also tried targeted therapies, like mTOR inhibitors, to treat uterine PEComas, but results varied. Gao et al. [5, 9] combined a VEGFR inhibitor, sorafenib, with a mTOR inhibitor sirolimus to treat a patient with rapidly progressive uterine PEComa and got a good response. Long-term follow-up of uterine PEComas is also of great importance, since our knowledge of this condition is still limited.

Conclusion

To conclude, we reported five cases of uterine PEComa with various clinical pathological characteristics. All the five cases were diagnosed after surgery, based on pathology examination and immunohistochemistry. Clinically, we should not neglect this cancer though it is very rare. For malignant PEComas, chemotherapies and target therapies

may be applied. More experience in diagnosing and treating this cancer is demanded.

Author contributions WWS: manuscript writing/editing. XJC: supportive supervision. XZL: provided the source information and idea, manuscript editing, supervision. YS: language editing. QZ: pathology review. BYY, LYX, BL, CCN, QYLV, YLC, BYX, MZB, YHX: follow-up and searching medical records.

Compliance with ethical standards

Conflict of interest All authors declare no conflict of interest.

Ethical approval This report complies with the tenets of the Declaration of Helsinki, and the medical ethics committee of the Obstetrics and Gynecology Hospital of Fudan University approved this study. Informed consent was obtained from all the five patients in this report.

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