

Perivascular epithelioid cell tumor arising in the left parametrium: a case report

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Summary

The perivascular epithelioid cell tumors (PEComas) mimic rare epithelial mesenchymal tumors with epithelioid features and perivascular clear cells that co-express the muscle and melanocytic markers. The PEComas include lymphangioleiomyomatosis, clear cell sugar tumor, angiomyolipoma, and myomelanocytic tumors. We report a case involving 37-year-old woman with a PEComa in the left parametrium. The patient was referred to our hospital with an adnexal tumor detected during medical examination. Therefore, she underwent laparoscopic mass excision surgery. Immunohistochemical staining of the tumor cells indicated positivity for Melan A, HMB45, and desmin. Finally, the tumor was diagnosed as a PEComa. The patient is undergoing regular follow-up, and no recurrence has been noted at 2 years after the surgery.

Key words: Perivascular epithelioid cell tumors; Parametrium; Immunohistochemistry.

Introduction

According to the World Health Organization (WHO), a perivascular epithelioid cell tumor (PEComa) is defined as “mesenchymal tumor consisting of histologically and immunohistochemically distinctive perivascular epithelioid cells” [1]. These cells express melanocytic and smooth muscle markers. Pea et al. first reported these cell perivascular epithelioid cells in both clear cell sugar tumors (CCSTs) and renal angiomyolipomas (AMLs) of the lung in 1991 [2]. In 1992, Bonetti et al. first proposed that a PEComa was a tumor consisting of cells with similar morphological and immune phenotypes, such as clear cell “sugar” tumors, angiomyolipomas, and lymphangioleiomyomatosis [3]. The tumors can develop at any anatomical site, such as the lung, kidney, uterus, liver and bowel. According to Liu et al., gynecologic PEComas are very rare, and we know of only 114 cases that have been reported to date [4]. However, in the previously reported cases, the PEComas did not develop in the parametrium. Immunohistochemically, the PEComas stain positively for the melanocytic markers, such as HMB45, Melan-A, tyrosinase, and MiTF, and muscle markers, such as SMA, pan-muscle myosin, calponin, h-caldesmon; however, desmin positivity is encountered less often [5]. Due to its rarity, the optimal management strategy for the PEComas has not been established to date. Here, we describe a case of a PEComa arising in the left parametrium. To the best of our knowledge, this is the only reported case of a PEComa in the parametrium.

Case report

A 37-year-old, gravida 1, para 1 woman was referred to our gynecologic outpatient department from a local clinic due to an ovarian mass incidentally detected on medical examination. She had a history of breast cancer operation but no other significant medical history. The patient’s body mass index was 21.1 kg/m². The chest radiograph showed unremarkable. The serum CA-19-9 (27.14 U/m), CA125 (11.6 ug/mL), AFP (1.7 ng/mL), and CEA (0.6 ng/mL) levels were not elevated. Pelvic computed tomography (CT) scan revealed a 3.0 × 3.0-cm sized homogeneous enhanced mass in the left parametrium with central low attenuation (Figure 1). Clinically, the patient was diagnosed with a neurogenic tumor; therefore, laparoscopic mass excision operation was performed. Before the operation, the tumor was thought to be of ovarian or broad ligament origin. On operation a 3.0-cm sized hemorrhagic and fragile mass was detected in the left parametrium. It was difficult to remove the tumor intact. The tumor tissue was broken as it was removed with forceps. The resection margin was not clear. There were no metastatic lesions or ascites identified in the pelvic cavity. Macroscopically, the excision specimen was generally fragile and consisted of a 3.2 × 1.7 × 0.6-cm sized lesion with a grayish-brown surface (Figure 2). Microscopic examination revealed that the tumor cells were arranged in nests, and blood vessels with thin walls were observed between them. The cells were relatively uniform and had clear or eosinophilic granular cytoplasm (Figure 3A). The nuclei of the tumor cells were small, round, and unformed. Pleomorphism was not observed. The nucleolus was clearly visible in some cells but not so prominent. Macronucleoli, which are usually present in melanomas,



Figure 1. — Pelvic computed tomography scan showing a 3.0 × 3.0-cm sized homogenous enhanced mass in the left parametrium.

were not observed (Figure 3B). Neither mitosis nor necrosis was observed. The immunochemical studies showed that the tumor cells were positive for HMB45, CD68, and Melan-A and focally positive for desmin. The morphology and immunohistochemistry results were indicative of pigmented PEComa. The patient was discharged 3 days after the operation without any surgical complications. She has not received adjuvant treatment and is currently being followed without recurrence two years after the surgery.

Discussion

We report a PEComa that arose from the left parametrium without metastasis. This term was introduced in 1992 and 2002 by the WHO. Epithelioid smooth muscle tumors, endometrial stromal sarcomas, and leiomyosarcomas should be considered in the differential diagnosis of uterine PEComas. The diagnosis may be difficult; therefore, careful analysis is needed. HMB45 is a commonly used immunochemical marker for the detection of a melanoma that reacts against an antigen present in the melanocytic tumors. PEComas usually stain negative or partially positive for S100. S100 protein positivity is observed in most clear cell sarcomas; therefore, the S100

marker is necessary for the differential diagnosis of PEComas and clear cell sarcomas. Melan A is a melanocytic marker, also known as MART-1. The expression of melanocytic markers, such as Melan-A and HMB45, and negative staining for cytokeratins are strongly suggestive of PEComas. There are many limitations in determining the malignant potential of tumors based on the histological findings. Therefore, Folpe et al. proposed a criteria for classifying the malignant potential of PEComas based on the various clinical features [6]. These criteria set the size of the mass, degree of invasion, nuclear grade, cellularity, mitotic index, necrosis, and vascular invasion as high-risk factors. A benign tumor was defined as a tumor < 5 cm in size without high risk factors. Uncertain malignant potential tumor was defined as tumor size > 5 cm with no other high-risk features or PEComas with nuclear pleomorphism or multinucleated giant cells only. A malignant tumor was defined as a tumor with two or more high-risk features. In our case, the size of the tumor was 3.2 cm and no high-risk factors were observed; therefore, the tumor in our patient can be classified as a benign tumor according to Folpe's classification.

Primary treatment consists of surgical excision of the tumor. Systemic chemotherapy and recently emerging m-



Figure 2. — The gross appearance of the resected tumor. The tumor has a black pigmented surface and measures 3 cm in diameter.

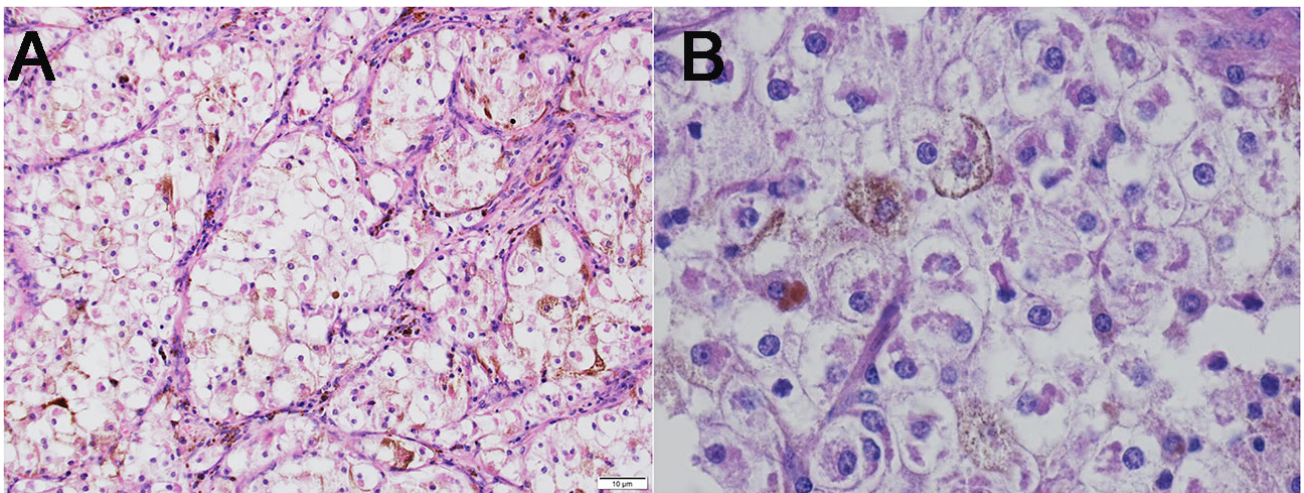


Figure 3. — A. Tumor cells are arranged in nests and have clear or eosinophilic granular cytoplasm. B. Tumor cell nuclei are small, round, and uniform. Pleomorphism is not observed.

TOR inhibitors are used for treating metastatic recurrent PEComas. Bleeker et al stated that systemic chemotherapy and radiation had shown little benefit in case of malignant PEComas. However, PEComas were included in the soft tissue sarcoma category, and doxorubicin and ifosfamide have been reported to be active against soft tissue sarcomas [7, 8]. Owing to the rarity of the disease, the optimal treatment strategy for this tumor has not been established.

PEComa can recur and metastasize; therefore, physicians should carefully monitor these patients.

In summary, a uterine PEComa is an epithelioid mesenchymal tumor that mimics leiomyosarcomas. They are rare epithelioid mesenchymal tumors with perivascular clear cells with epithelioid features that co-express melanocytic and muscle markers. The malignant potential of PEComas is uncertain.

Ethics approval and consent to participate

All subjects gave their informed consent for inclusion before they participated in the study.

Acknowledgments

Thanks to all the peer reviewers and editors for their opinions and suggestions.

Conflicts of interest

The authors declare to have no conflict of interest.

Submitted: January 06, 2020

Accepted: June 17, 2020

Published: August 15, 2020

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