Case Report

Malignant Perivascular Epithelioid Cell Tumor of Ovary: A Rare Case Report

Anuradha Sharma, Reetika Sharma, Jyoti Bala, Monika Sharma[1]

Departments of Pathology and [1]OBG, Dr. RKGMC, Hamirpur, Himachal Pradesh, India

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Abstract

Perivascular epithelioid cell tumors (PEComas) have unique morphology comprised perivascular epitheloid cells and express both melanocytic and smooth muscle cell markers. Gynecological PEComas account for approximately 25% of all PEComas, and in most cases, the primary site of the tumor is the uterine body. Ovarian PEComa is exceptional. Here, we report a case of primary malignant PEComa of ovary in a 38 years female.

Keywords: *Immunohistochemistry, malignant perivascular epithelioid cell tumor, ovary*

Introduction

Perivascular epithelioid cell tumors (PEComas) include the group of mesenchymal neoplasm present in different sites. These neoplasms have unique morphology comprised perivascular epitheloid cells and express both melanocytic and smooth muscle cell markers, human melanoma black-45 (HMB-45), human melanosome-associated antigen-1, MelanA/Mart1, microphthalmia transcription factor, smooth muscle actin (SMA), pan-muscle actin, muscle myosin, calponin, sometimes h-caldesmon, and less commonly, desmin.[1-3]

Gynecological PEComas account for approximately 25% of all PEComas, and in most cases, the primary site of the tumor is the uterine body. Adnexal PEComas are exceptional, with only 5 primary cases and 7 metastatic cases described in the literature.[2] Here, we report a case of primary malignant PEComa of the ovary.

Case Report

A 38-year-old female presented with complaints of pain abdomen and fever. Contrast–enhanced computed tomography showed the right abdominopelvic mass measuring 8.8 cm × 9.2 cm × 10 cm with nonvisualization of the right ovary likely neoplastic. The left adnexa was unremarkable. The liver, spleen, pancreas, lung, bilateral kidneys, urinary bladder, bowel, and mesenteric lymph nodes were free from any lesions.

We received a specimen of the uterus with bilateral fallopian tubes, left ovary, and right adnexal mass in histopathology; the specimen was already cut open and distorted. Uterus with cervix measuring 7.5 cm × 4 cm × 2 cm. Myometrium measuring 1.2 cm. Bilateral fallopian tubes and left side ovary were unemarkable. The right side ovarian mass was measured 10 cm × 9 cm × 10 cm. The cut surface of the mass was gray-white to yellow with brown friable areas with necrotic areas [Figure 1]. Microscopic examination from the mass showed sheets of tumor cells that were polygonal and spindle in shape with well-defined cell borders with marked nuclear pleomorphism, vesicular nuclei, and prominent nucleoli with clear to granular cytoplasm [Figure 2]. Scattered in between large tumor giant cells (uninucleated/multinucleated) seen [Figure 3]. In addition, areas of necrosis, hemorrhage were also seen. Focal sclerosing pattern was also seen. Periphery of the tumor showed normal ovarian parenchyma. The tumor was infiltrating the parametrium. The endometrium, myometrium, cervix, and bilateral fallopian tubes are free from the tumor. On other side ovary, the omental biopsy showed metastatic tumor deposits. On immunohistochemistry, the tumor cells were positive for EMA (1+), Vimentin (3+), Ki 67 (25%–30%),

Address for correspondence: Dr. Reetika Sharma, Department of Pathology, Dr. RKGMC, Hamirpur, Himachal Pradesh, India.
E-mail: dr_ritu85@yahoo.com

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Desmin (2+), SMA (2+), h-Caldesmon (3+), CD10 (1+), CYCLIN D1 (3+), and INI 1 retained in tumor cells. P16 positive in block positive pattern, p53 (strongly positive), HMB-45 (2+), and MELAN A (1+). The tumor cells were negative for CK, SALLA4, S-100, ER, LMW-CK, and TFE3 [Figure 4]. On basis of above morphology and IHC report, the diagnosis of malignant PECOma was given.

Discussion

The World Health Organization defined PEComas as mesenchymal tumors for the first time in 2002. PEComa tumors are considered to be heterogeneous. Mainly three hypotheses are given in their cell of origin, one being neural crest origin, second being myoblastic, and third being derived from perivascular epitheloid cells which are considered to be progenitor of fat and muscle cells. The angiomyolipoma is thought to be derived from this.[3]

Folpe *et al*.[4] classified PEComas as benign, of uncertain malignant potential or malignant on the basis of 6 findings represented by tumor size, infiltrative growth pattern, nuclear grade and cellularity, mitoses/50 high power fields (HPF), necrosis, and vascular invasion. Schoolmeester *et al*.[5] described only 5 worrisome features, i.e., tumor size >5 cm, high-grade atypia, mitoses >1/50 HPF, necrosis, and lymphovascular invasion. PEComas were defined as either benign/uncertain malignant potential or malignant according to whether there were <4 versus ≥4 worrisome features, respectively. In our case, the tumor was 10 cm^3 high-grade nuclear atypia, 5 mitotic figures/50 HPF, and areas of necrosis with absent lymphovascular invasion. Hence, the diagnosis of malignant primary PEComa of the ovary was made.

The most common sites of PEComa are the kidneys, liver, lungs, and uterus,[6] as well as a few cases that have been reported in bladder, prostate, ovaries, pancreas,[4,7] and soft tissues.[4] Recent studies have also revealed cases where PEComas have developed in cutaneous[8] stomach,[9] and gastrointestinal tracts,[10] as well as

Figure 1: Gross photo showing uterus with cervix with right adnexal mass

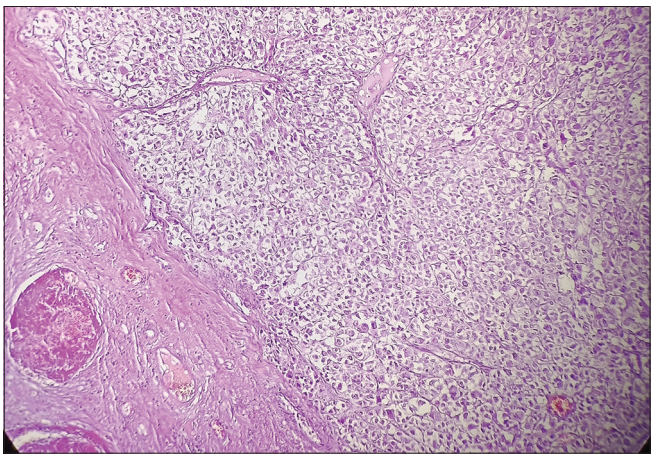

Figure 2: Photomicrograph showing sheets of tumor cells with perivascular arrangement (H and E, ×10)

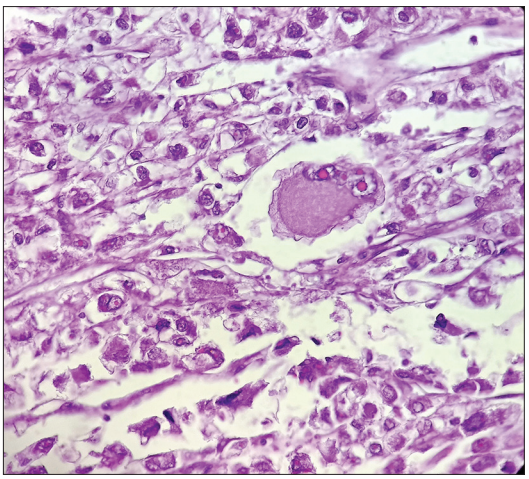

Figure 3: Photomicrograph showing epitheloid cells with clear to granular cytoplasm with prominent nucleoli along with tumor giant cells (H and E, ×10)

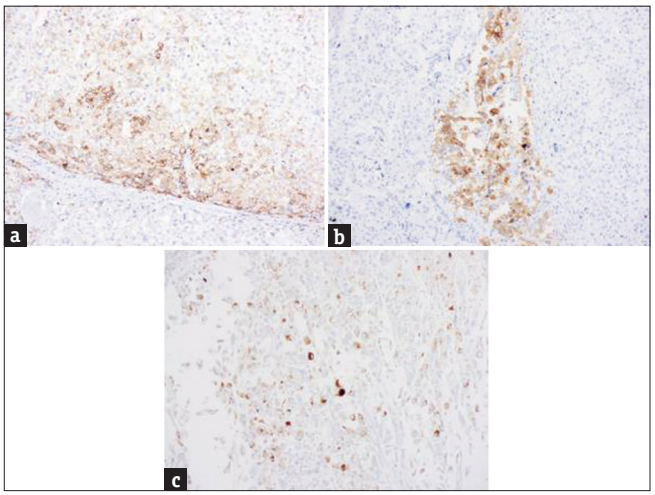

Figure 4: Photomicrograph of IHC of, (a) HMB-45 (2+), (b) Melan A (1+), and (c) Desmin (2+)

orbital,[11] omentum.[12] In gynecological tract, the uterus is most common site for PECOma and ovarian PECOmas are very rare, only five cases in the literature. Most of the literature regarding gynecologic PEComas is composed of case reports and small case series. PEComa can coexist with leiomyoma and fumarate hydratase-deficient atypical leiomyoma.[13] Only 20% of PECOmas are associated with genetic alteration tuberous sclerosis complex (TSC). TSC1 and TSC2 gene products typically act to inhibit mammalian target of rapamycin (mTOR) which is the basis of the current practice guidelines for malignant PEComa.[3] Some PECOmas exhibit TFE3 rearrangement, hence don't respond to mTOR. These tumors strongly exhibit diffuse positive expressions of HMB-45 and TFE3, weakly positive expressions of SMA, and negative expressions of Melan A.[14] In our case, HMB-45 was strongly positive and Melan A was weakly positive. Due to rarity of this tumor and very few case reports, there are no clear guidelines available in the literature. In the available, data the management of PECOmas included surgical resection, chemotherapy, and radiotherapy depending upon the histologic type. The mTOR inhibitors have crucial role in the management wherever the mutation is present.[1] Our case was managed by surgical excision of the uterus with cervix with bilateral fallopian tubes and one side ovary and other side ovarian mass followed by chemotherapy. This case should be reported in the literature due to its rarity. Moreover, since there are very few case reports and case series in the literature, hence, more research is needed to comprehend its pathology and treatment strategies.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Levin G, Capella MP, Meyer R, Brezinov Y, Gotlieb WH. Gynecologic perivascular epithelioid cell tumors (PEComas): A review of recent evidence. Arch Gynecol Obstet 2024;309:2381-6.
2. Gadducci A, Ugolini C, Cosio S, Vistoli F, Kauffmann EF, Boggi U. Primary perivascular epithelioid cell tumor (PEComa) of the ovary: A case report and review of the literature. Anticancer Res 2021;41:4483-8.
3. Bao L, Shi Y, Zhong J, Zhao M, Wu J, Hai L, *et al.* Histopathologic characteristics and immunotypes of perivascular epithelioid cell tumors (PEComa). Int J Clin Exp Pathol 2019;12:4380-9.
4. Folpe AL, Mentzel T, Lehr HA, Fisher C, Balzer BL, Weiss SW. Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: A clinicopathologic study of 26 cases and review of the literature. Am J Surg Pathol 2005;29:1558-75.
5. Schoolmeester JK, Howitt BE, Hirsch MS, Dal Cin P, Quade BJ, Nucci MR. Perivascular epithelioid cell neoplasm (PEComa) of the gynecologic tract: Clinicopathologic and immunohistochemical characterization of 16 cases. Am J Surg Pathol 2014;38:176-88.
6. Kwon BS, Suh DS, Lee NK, Song YJ, Choi KU, Kim KH. Two cases of perivascular epithelioid cell tumor of the uterus: Clinical, radiological and pathological diagnostic challenge. Eur J Med Res 2017;22:7.
7. Zemet R, Mazeh H, Neuman T, Freund HR, Eid A. Asymptomatic pancreatic perivascular epithelial cell tumor (PEComa) in a male patient: Report and literature review. JOP 2011;12:55-8.
8. Stuart LN, Tipton RG, DeWall MR, Parker DC, Stelton CD, Morrison AO, *et al.* Primary cutaneous perivascular epithelioid cell tumor (PEComa): Five new cases and review of the literature. J Cutan Pathol 2017;44:713-21.
9. Shin SA, Choi J, Moon KC, Kim WH. Perivascular epithelioid cell tumor in the stomach. J Pathol Transl Med 2017;51:428-32.
10. Chen Z, Han S, Wu J, Xiong M, Huang Y, Chen J, *et al.* A systematic review: Perivascular epithelioid cell tumor of gastrointestinal tract. Medicine (Baltimore) 2016;95:e3890.
11. Varan A, Bayhan T, Kiratli H, Özoğul E, Kösemehmetoğlu K, Bulut E, *et al.* An orbital perivascular epithelioid cell tumor in a 7-year-old boy: Case report and review of the literature. J AAPOS 2017;21:325-8.e1.
12. Okamoto K, Okada Y, Ohno K, Yagi T, Tsukamoto M, Akahane T, *et al.* A rare case of perivascular epithelioid cell tumor (PEComa) of the greater omentum. World J Surg Oncol 2018;16:113.
13. Liu Y, DeLeon EM, Okour S, Vang R, Hung CF, Wu TC, *et al.* Coexistence of conventional leiomyoma, fumarate hydratase-deficient atypical leiomyoma, and perivascular epithelioid cell tumor in a uterus: A case study. Int J Gynecol Pathol 2021;40:134-40.
14. Kim H, Kim J, Lee SK, Cho EY, Cho SY. TFE3-expressing perivascular epithelioid cell tumor of the breast. J Pathol Transl Med 2019;53:62-5.