EJGO European Journal of Gynaecological Oncology

Perivascular epithelioid cell tumor of vulva: a rare case

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Summary

Background: Perivascular epithelioid cell tumor (PEComa) is very rare mesenchymal tumor with an unpredictable natural history. To the best of the present authors' knowledge, there is only one reported case of PEComa of vulva in the English language medical literature. *Case Report:* A 49-year-old woman presented with PEComa of vulva. The diagnosis was established postoperatively by immunohistochemistry. To date, two months after operation, the patient is alive with no evidence of recurrence or metastasis. *Conclusion:* A variety of imaging methods and pathological methods including a surgical resection with clear margin may be necessary in cases of patients with mesenchymal tumor of vulva suspecting PEComa. Urgent diagnosis and treatment of these patients seemed to be critical.

Key words: Perivascular epithelioid cell tumor; PEComa; Vulva.

Introduction

Perivascular epithelioid cell tumor (PEComa) is a rare mesenchymal neoplasm [1-4]. According to World Health Oraganization (WHO), PEComas are defined as mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells [2, 3]. PEComas of the female gynecological tract account for nearly 25% of all the PEComa cases [1-3]. In most of the PEComa cases, the original site of appearance of the tumor was the uterine corpus, while there is only one reported case of PEComa of vulva in the English language medical literature [1-4]. Its natural history is unpredictable, and there are no established treatment guidelines for this tumor type [1-3]. Here, the authors present a rare case of PEComa of vulva.

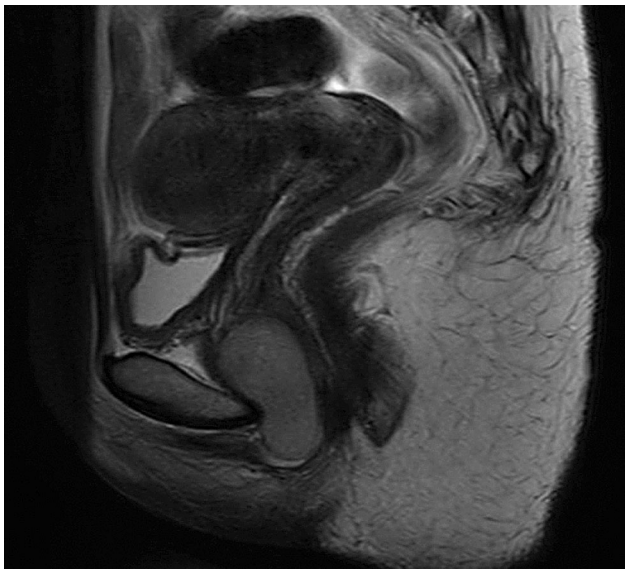

Figure 1. — MRI examination of the pelvis which demonstrates a highly vascular mass of left vulva. There is no evidence of tumor invasion to the surrounding tissue.

Case Report

A 49 year-old woman without a past medical history of interest was referred to this hospital with discomfort of left vulva. No other symptoms were observed. Pelvic examination revealed a solid tumor of left vulva. No other abnormal findings were observed including vagina. Ultrasonography revealed a highly vascular tumor arising from left vulva. MRI examination of the pelvis, which demonstrated a 3×4 cm highly vascular mass of left vulva, suggesting a vulvar tumor (Figure 1). CT examination of the abdomen and chest demonstrated no metastasis. Laboratory investigations showed no remarkable findings including serum LDH level. Serum tumor markers were as follows: CA125: 13 U/mL, CEA: 0.5 ng/mL, CA19-9: 22 U/mL, SCC: 0.8 ng/mL, and AFP: 2.0 ng/mL. The biopsy of tumor of left vulva revealed a spindle cell tumor. The cytological examination of cervix, endometrium, and vagina showed no abnormality. Therefore, mesenchymal tumor of left vulva was initially considered the most likely diagnosis.

The patient underwent surgery. Macroscopically, the tumor (size 3×4 cm) was seen arising from left vulva, and was grey-white in color and solid (Figure 2). The tumor was very vascular similarly with the time of biopsy. The deep portion of the tumor was separated from vagina and urethra, and the tumor was removed with clear margin. Stamp cytology of the tumor showed atypical cells suspecting spindle cell tumor. There were no other remarkable findings. The patient had an uneventful postoperative course.

The pathological examination showed that the tumor was arranged with A trabecular pattern around the vessels. The tumor cells had clear or slightly granular cytoplasm and small to medium-sized normochromatic nuclei that were oval to spindle-

Revised manuscript accepted for publication March 15, 2017

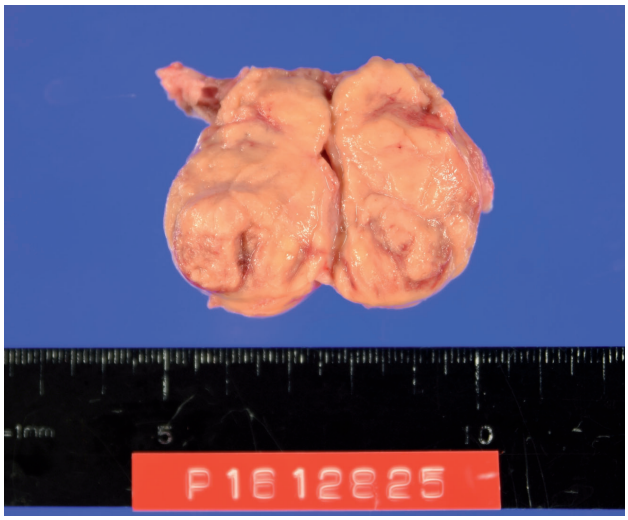

Figure 2. — Macroscopic image of resected specimen. There is a left vulvar tumor (size 3×4 cm) which is grey-white in color and solid.

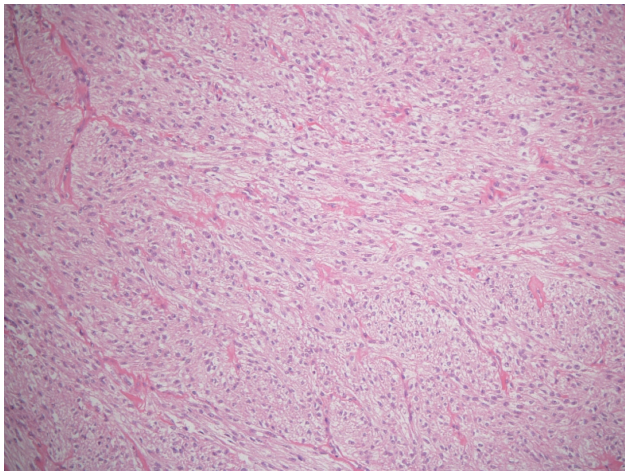

Figure 3A. — Microscopic image of tumor cells (Hematoxylin and Eosin, ×100), which shows clear or slightly granular cytoplasm and small to medium-sized normochromatic nuclei.

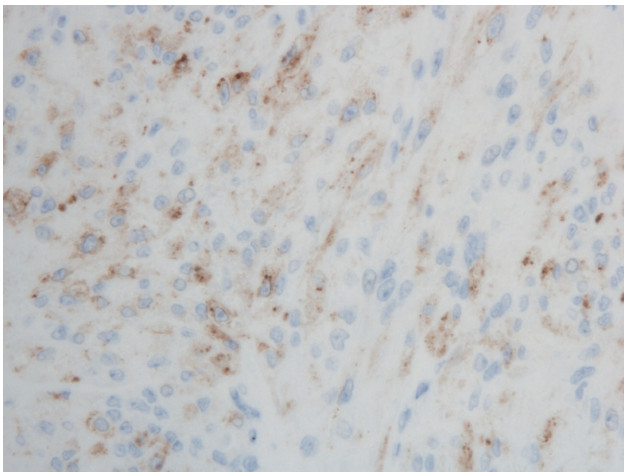

Figure 3B. — Microscopic image of HMB-45-positive tumor cells (×400).

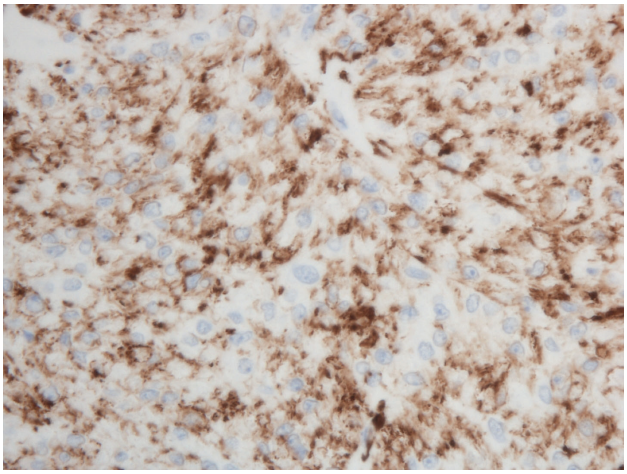

Figure 3C. — Microscopic image of SMA-positive tumor cells (×400).

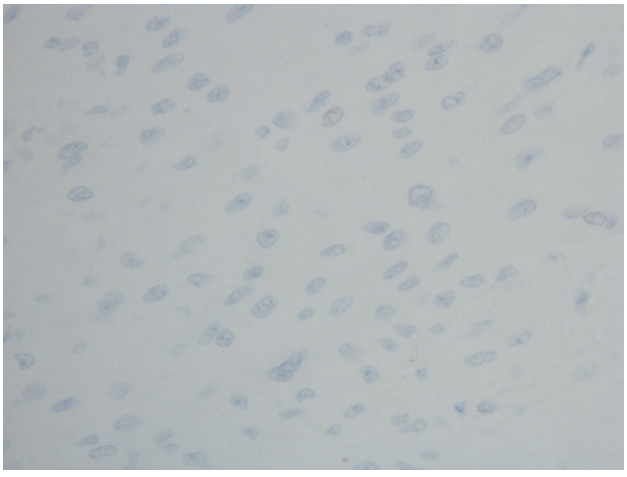

Figure 3D. — Microscopic image of S-100-negative tumor cells (×400).

shaped. The tumor cells showed strong positivity by immunohistochemistry for human melanin black 45 (HMB-45), smooth muscle actin (SMA), cluster of differentiation 10 (CD10), and vimentin, although Melan-A, S-100, microphthalmia transcription factor (MiTF), desmin, AE1-AE3, CD34, c-kit, myogenin, and myogenic differentiation 1 (Myo-D1) was negative (Figure 3). Ki-67 labeling index of 6% of neoplastic cells have been observed. Finally, the pathological examination showed a PEComa. The present authors confirmed the diagnosis of PEComa of left vulva. To date, two months after operation, the patient is alive with no evidence of recurrence or metastasis.

Discussion

PEComa was first described in 1992 [5], and includes a group of tumors with distinctive perivascular epithelioid cells such as angyomiolipomas, lymphangiomyomatosis,

and clear cell sugar tumor of the lung [1-4]. PEComa is a mesenchymal neoplasm composed of histologically perivascular epithelioid cells. PEComa is a very rare tumor reported in almost every site including skin, pancreas, pelvic cavity, uterus, prostate, bladder, digestive tract, heart, trachea, lymph node, breast, bone, soft tissues, and vulva [1-5]. PEComas of the female gynecological tract account for nearly 25% of all the PEComa cases [1-3]. In most of the PEComa cases, the original site of appearance of the tumor was the uterine corpus [1-3]. To the best of the present authors' knowledge, there is only one reported case of PEComa of vulva in the English language medical literature [4].

PEComa is a rare tumor, especially in the vulva. Therefore, it is difficult to consider initially the most likely diagnosis. In the present case, the diagnosis was established postoperatively by immunohistochemistry. In several reports, the diagnosis of PEComa was not made until recurrence [6]. It is possible to make a diagnosis of PEComa by an immunohistochemistry examination, while adequate sampling of deep portion of the tumor is difficult due to high vascularity. Although mesenchymal tumor of vulva was initially considered the most likely diagnosis in the present case, it was impossible to confirm the diagnosis of PEComa by the biopsy of only a small portion of the tumor. Surgical resection may be necessary in cases of patients with mesenchymal tumor of vulva suspecting PEComa.

The natural history of PEComa is unpredictable, and there are no established treatment guidelines for this tumor [1-3]. In most cases, PEComa behaved in a benign fashion and remained confined to the primary sites of origin such as in the present case. In these cases, the resection of the tumor was curative. However, there have been reported cases of local recurrence and distant metastasis [7]. The optimal resection of the tumor with clear margins seems to be necessary for preventing a disease recurrence. The role of adjuvant therapy remains unclear [1-3]. In cases with metastases, the patients have been successfully managed by resection alone [7]. Therefore, patients with PEComa should be carefully monitored for recurrence.

A variety of imaging methods and pathological methods including a surgical resection with clear margin may be necessary in cases of patients with mesenchymal tumor of vulva suspecting PEComa. Urgent diagnosis and treatment of these patients seemed to be critical.

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