



PEComa-A Rare Uterine Neoplasm: A Case Report

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Abstract

Background: Perivascular Epithelioid Cell Tumors (PEComas) are rare mesenchymal tumors originating from perivascular epithelioid cells. The second common affected organ is uterine. Most of PEComas are benign and patients have good prognosis. At the present time, surgery is the main treatment and adjuvant chemotherapy is used in malignant cases, although the best diagnostic and management method is yet to be discovered considering the rarity of this neoplasm.

Case Presentation: The patient was a 53 year old lady with a history of two vaginal deliveries and no previous surgery. She had severe pelvic pain and underwent MRI with the primary impression of sarcoma. In MRI, she had a 7 cm mass in lower segment of uterus. The patient underwent laparoscopic hysterectomy, bilateral oophorectomy, lymphadenectomy, and omental biopsy in Jam Hospital. Pathologic report of the patient revealed malignant PEComa without lymph node and omentum involvement.

Conclusion: Diagnosis of PEComa before surgery is difficult and its differential diagnoses form uterine leiomyoma or leiomyosarcoma. Final diagnosis can be made after surgical biopsy and immunohistochemistry evaluation. Surgery is still the main treatment and adjuvant therapy is used in high risk patients.

Keywords: Case report, Pathology, PEComa, Perivascular epithelioid cell tumor.

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Introduction

Perivascular Epithelioid Cell Tumors (PEComas) are rare mesenchymal tumors first reported in 1995. The term "PEComa" was used to describe tumors originated from perivascular epithelioid cells in 1996 (1). PEComas are a family of mesenchymal tumors including angiolipomas, clear cell tumors of the lung, and lymphangiomyomas. Most of these rare tumors are benign. Besides peritoneum, the second common organ affected by the tumor is uterus (2). Up to now, about 100 patients with PEComas are reported among whom one third had uterine in-

volvement. As affected cases of this disease were not reported, standard diagnostic and management procedure have not been described yet. In this case report, a patient with uterine PEComa is reported and described.

Case presentation

The patient was a 53 year old lady, who reached menopause 3 years ago, with a history of 2 vaginal deliveries. She had severe pelvic pain and abnormal uterine bleeding and underwent diagnostic curettage before visiting us. The report was nor-

mal with atrophic endometrium. Sonographic evaluation revealed a lobulated hypoechoic mass, 67×46×64 mm in size, from posterior uterine wall towards cervix. Gynaecologic evaluation revealed a mass in left side of uterus and MRI report was suggestive of a uterine sarcoma.

The patient underwent bilateral oophorectomy and hysterectomy. A 5 mm and a 12 mm trocar were used to enter abdominal cavity. A mass behind cervix and frozen pelvis were observed. An enlarged uterine with a bulging cervix, adhesion of ovaries to pelvic floor, and intestinal adhesion to cervix and left adnexa were observed. Uterine with the mass in cervix was separated intactly and removed from vagina (manipulator was not used due to probability of malignancy). The sample was sent to frozen section and in frozen section, using cryotomy and touch smear, a tumor consisting of monomorphic cells with moderate atypia and necrosis sites was observed. Mesenchymal tumor in favor of cervical sarcoma was reported. Left parametrium was thoroughly involved compressing left urethra. Right parametrium had lower involvement. Bilateral double J was inserted. The mass had sarcomatous pattern. Left parametrium was completely removed. An omental biopsy was performed. Then lymph nodes of both obturator and iliac region, left and right sides, were removed completely. An old thrombotic vein in pelvic floor and left side was observed and was not touched due to its creation of thrombosis in her last pregnancy. Samples were sent to pathology laboratory for cytological and immunohistochemistry evaluation.

In pathologic sample, a round soft mass in uterus wall, 7 cm in diameter, was observed and cervical mucosa was normal. Tissue samples were fixed by formalin 10% and colored with hematoxylin-eosin. A tumor consisting of monomorphic cells in regular nests and hypervascular background including delicate microvasculature separating cellular nests was observed. In larger view, multidimensional cells with clear cytoplasm and same nucleus with low to moderate atypia were observed. Cells were around vessels. Cells were colored using immunohistochemistry with HMB45, vimentin, and Melan A makers. P63, S100, cytokeratin, desmin, h-caldesmon were negative and differential diagnosis for the patient was PEC-omas (Figures 1-6).

Pathology lab report revealed malignant PEC-oma without involvement of peritoneum, pelvis,

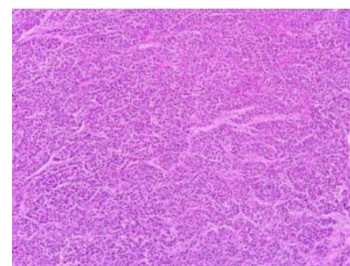


Figure 1. Monomorphic cells in regular nests and a hypervascular background including microvasculature separating cellular nests

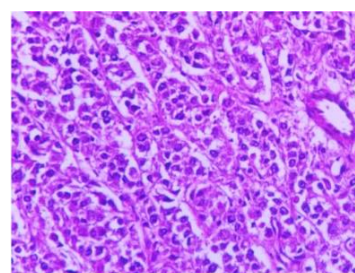


Figure 2. Larger view of multidimensional cells with clear cytoplasm, same-sized nucleus, and low atypia in dense groups



Figure 3. Neoplastic cells around vessels

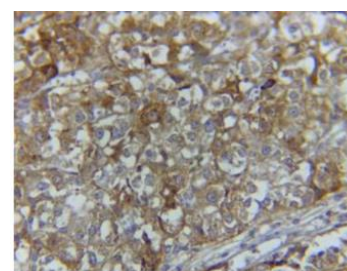


Figure 4. Fibrotic bands with lobular pattern in tumor

parametria, lymph nodes, and omentum. The patient was discharged after two days with a good general health and was referred to the radiotherapist. As there was no pelvic involvement, NEORAPA (antineoplastic agent) and Rapamune were

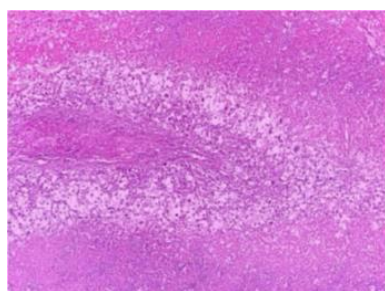


Figure 5. Immunohistochemistry with HBM45 marker

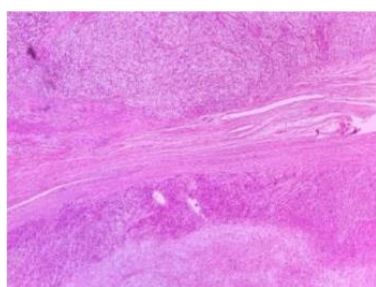


Figure 6. Immunohistochemistry with Melan A marker

prescribed for the patient. The patient is followed regularly.

Discussion

Up to now, about 100 patients with PEComa of uterine are reported. Mean age of patients was about 40 years (4) though it can be observed in all age groups. Hormones may play role in its pathogenesis in women (5, 6). Malignancy potential of PEComa depends on histopharmacology of primary tumor and not its origin. PEComas are more likely to affect retroperitoneum, kindies, and female reproductive system; yet, it occurs in other body parts as well. Cervix is rarely involved (7, 8). Differential diagnosis of PEComas include all types of benign and malignant tumors. PEComas may be mistaken for carcinomas such as clear cell tumor especially if the patient has involvement of cervix or bladder (4, 9). Pathogenesis of PEComa is still unclear and it is hypothesized that they are related to complex tuberous sclerosis (9).

Clinical presentation, physical examination of pelvis, and pelvis ultrasound are not specific and can be mistaken for benign tumors or uterine fibroids or other malignancies. This non-specificity makes difficulties in diagnosis and management of PEComa and delays treatment of some patients (10-12). Furthermore, patients with malignancy

have high survival rates without adjuvant treatment. Chemotherapy regimen for malignant PEComa of reproductive system is not well defined and need more studies. Currently, the role of radiotherapy is not clear enough. Histological analysis of malignant PEComa reveals high mitotic index and multiple necrosis. Necrosis is related to tolerance threshold of chemotherapy and high angiogenesis shows high sensitivity of cells to radiation; therefore, these two statements support the role of radiotherapy for malignant PEComa (4). Systemic chemotherapy regimen without surgery is not an effective treatment. Thus, adjuvant chemotherapy has not an effective role in prognosis of patients with malignant PEComa and surgery is still the main treatment. Most malignant PEComas has good survival without recurrence or metastasis (13). Most patients with PEComa undergo bilateral oophorectomy and total hysterectomy based on their age (14). When a simple surgery may be adequate for both benign and malignant PEComa, adjuvant therapy after surgery should be provided for patients in spite of weak efficacy of radiotherapy and chemotherapy (15). Different studies have indicated increase in survival rate after surgery by using combination of chemotherapy and radiotherapy (16).

Conclusion

Uterine PEComa is a rare disease and the main diagnosis is made after pathologic assessment of patient's sample following surgery and immunohistochemistry. Prognosis of the disease is depending on whether the tumor is benign or malignant. The first treatment for patients is surgery and in malignant cases, adjuvant therapy is needed for reducing the risk of recurrence and metastasis.

Conflict of Interest

The authors declare that they have no conflict of interest.

References

1. Pea M, Bonetti F, Zamboni G, Martignoni G, Riva M, et al. Melanocyte-marker-HMB-45 is regularly expressed in angiomyolipoma of the kidney. *Pathology*. 1991;23(3):185-8.
2. Pea M, Bonetti F, Zamboni G, Martignoni G, Fiore-Donati L, Doglioni C. Clear cell tumor and angiomyolipoma. *Am J Surg Pathol*. 1991;15(2):199-202.
3. Machado I, Cruz J, Laverna J, Rayon JM, Poveda A, Llombart-Bosch A. Malignant PEComa with me-

- tastatic disease at diagnosis and resistance to several chemotherapy regimens and targeted therapy (mTOR inhibitor). *Int J Surg Pathol*. 2017;25(6):543-9.
4. Musella A, De Felice F, Kyriacou AK, Barletta F, Di Matteo FM, Marchetti C, et al. Perivascular epithelioid cell neoplasm (PEComa) of the uterus: a systematic review. *Int J Surg* 2015;19:1-5.
 5. Baird DD, Dunson DB, Hill MC, Cousins D, Schectman JM. High cumulative incidence of uterine leiomyoma in black and white women: ultrasound evidence. *Am J Obstet Gynecol*. 2003;188(1):100-7.
 6. Sadigh S, Shah P, Weber K, Sebro R, Zhang PJ. Primary malignant perivascular epithelioid cell neoplasm (PEComa) of the bone mimicking granular cell tumor in core biopsy: a case report and literature review. *Oncol Lett*. 2018;15(3):2946-52.
 7. Conlon N, Soslow RA, Murali R. Perivascular epithelioid tumours (PEComas) of the gynaecological tract. *J Clin Pathol*. 2015;68(6):418-26.
 8. Sanfilippo R, Jones RL, Blay JY, Le Cesne A, Provenzano S, Antoniou G, et al. Role of chemotherapy, VEGFR inhibitors, and mTOR inhibitors in advanced perivascular epithelioid cell tumors (PEComas). *Clin Cancer Res*. 2019;25(17):5295-300.
 9. Pan CC, Chung MY, Ng KF, Liu CY, Wang JS, Chai CY, et al. Constant allelic alteration on chromosome 16p (TSC2 gene) in perivascular epithelioid cell tumour (PEComa): genetic evidence for the relationship of PEComa with angiomyolipoma. *J Pathol*. 2008;214(3):387-93.
 10. Prasad SR, Sahani DV, Mino-Kenudson M, Narra VR, Humphrey PA, Menias CO, et al. Neoplasms of the perivascular epithelioid cell involving the abdomen and the pelvis: cross-sectional imaging findings. *J Comput Assist Tomogr*. 2007;31(5):688-96.
 11. Giannella L, Carpini GD, Montik N, Verdecchia V, Puccio F, Di Giuseppe J, et al. Ultrasound features of a uterine perivascular epithelioid cell tumor (PEComa): case report and literature review. *Diagnostics (Basel)*. 2020;10(8):553.
 12. Pattamapaspong N, Khunamornpong S, Phongnarisorn C, Pojchamarnwiputh S. Malignant perivascular epithelioid cell tumor of the round ligament mimics leiomyoma on computed tomography. *Singapore Med J*. 2009;50(7):e239-42.
 13. Fletcher C, Bridge JA, Hogendoorn P, Mertens F. WHO classification of tumors of soft tissue and bone. 4th ed. Lyon: IARC Press; 2013. 467 p.
 14. Fadare O. Perivascular epithelioid cell tumor (PEComa) of the uterus: an outcome-based clinicopathologic analysis of 41 reported cases. *Adv Anat Pathol*. 2008;15(2):63-75.
 15. Jeon IS, Lee SM. Multimodal treatment using surgery, radiotherapy, and chemotherapy in a patient with a perivascular epithelioid cell tumor of the uterus. *J Pediatr Hematol Oncol*. 2005;27(12):681-4.
 16. Vang R, Kempson RL. Perivascular epithelioid cell tumor (PEComa) of the uterus: a subset of HMB-45-positive epithelioid mesenchymal neoplasms with an uncertain relationship to pure smooth muscle tumors. *Am J Surg Pathol*. 2002;26(1):1-13.