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PEComa of the Ovary: A Case Report

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ABSTRACT

Perivascular epithelioid cell tumors (PEComas) are a group of mesenchymal neoplasms with different biological presentations from benign to malignant types. Hence, we report the first description of PEComa presenting with an acute surgical abdomen. Her definite diagnoses were made based on the histomorphology results at Imam Khomeini Hospital, Tehran University of Medical Sciences, in 2020.

Considering different clinical presentations, unknown characteristics of imaging, and rarity of PEComa, preoperative diagnosis of it seems impossible. However, IHC can play an important role in its diagnosis.

Keywords: Malignant, MRI, Ovary, Perivascular epithelioid tumors, Tumor marker, Ultrasound

Introduction

Perivascular epithelioid cell tumors (PEComas) are a group of mesenchymal neoplasms with a different biological presentation from benign to malignant types **(1)**. The PEComas neoplasms include angiomyolipoma (AML), lymphangioma, the falciform ligament teres clear cell myomelanocytic tumor, as well as other tumors with the same features at different organs **(2)**.

Although due to its rarity, the most common anatomic location with PEComa has not been accurately determined, the genitourinary tract, especially the uterus and kidney, were the most frequent site of PEComa in previously reported cases **(3-5)**

On the other hand, the clinical presentation of PEComa is not well recognized; spontaneous hemoperitoneum in pregnancy **(6)**, mild abdominal discomfort **(1)**, intermittent lower abdominal pain, and vaginal bleeding **(2)** are some examples of PEComa clinical features.

Hence, herein we report the first description of PEComa presenting with an acute surgical abdomen. The patient's definite diagnoses were made based on the histomorphology results at Imam Khomeini Hospital, Tehran University of Medical Sciences, in 2020.

Case Report

A previously healthy 32-year-old virgin single woman with a chief complaint about acute severe abdominal pain was referred to Imam Khomeini Hospital's emergency department, Tehran, Iran. Her physical examination and laboratory findings were unremarkable.

Ultrasound examination showed hypovascular and solid tumor 9×9×18 cm in the right adnexal with huge ascites. CT scan of the abdomen revealed severe ascites, hypodense lesion 5×5 cm in liver, solid cystic mass in right ovary, multiple omental and peritoneal tumor maximally 5 cm near liver (Figure 1A, B).

Furthermore, the thorax CT scan detected three small nodules (3mm/3mm/4mm) in the left lung with mild pleural effusion on the right side. Her tumor marker tests showed an elevated serum CA125 concentration

up to 557 U/mL, and the other tumor markers were within normal limits.

Intraoperative findings revealed a huge amount of bloody ascites (six liters). There was a 20×12 cm solid hemorrhagic cystic tumor in the right adnexal tumor with omental involvement. During laparotomy, bilateral salpingo-oophorectomy (due to undifferentiated epithelial histology of frozen result in both ovaries), pelvic lymphadenectomy, infra-colic omentectomy with uterine preservation were performed.

The post-operation pathology report revealed nests of epithelioid cells by cleared-out cytoplasm and uniform nuclei by mild cytologic atypia. Furthermore, CD99, vimentin, NSE, melan A, C-kit, SAX10, human melanin black-45 (HMB-45), S100 (few scattered), synaptophysin (few scattered) were positive in immunohistochemical (IHC) staining analysis.

According to the above histopathological findings, the diagnosis of a PEComa was made. After cytoreductive surgery, chemotherapy with BEP was immediately initiated. After four cycles of chemotherapy, the tumor recurred, and the patient was a candidate for the second cytoreductive surgery. Still, she refused and did not refer to the hospital to complete her therapy. After that, we did not know about the detail of her treatment, and unfortunately, her relatives said that she had died.

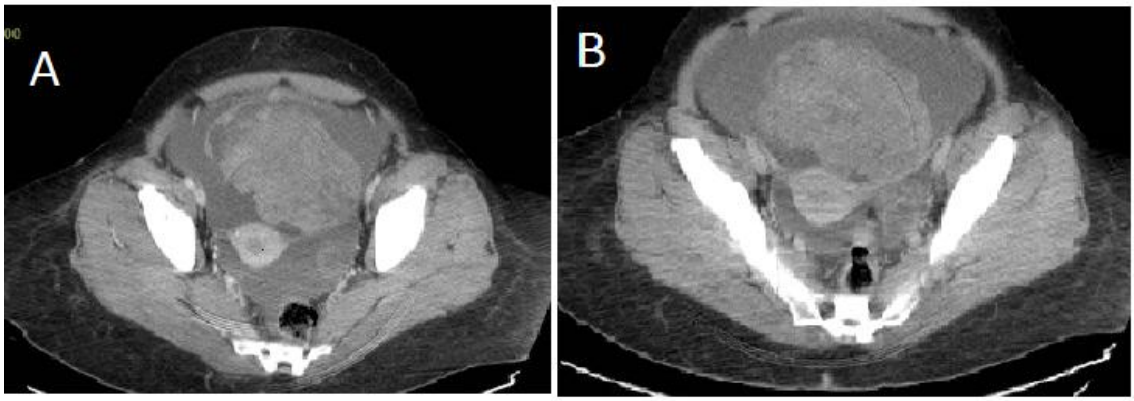

Figure 1. CT-scan of the abdomen in a 32-year-old female with malignant perivascular epithelioid cell tumors of the ovaries.

Discussion

PEComa is a rare tumor with a unique diagnostic challenge that leads to a late diagnosis, similar to our study case (7). However, Knowledge of its specific clinicopathological, imaging, and tumor marker may accelerate the diagnosis of PEComa (8).

It is worth mentioning IHC plays a vital role in its diagnosis. Synchronous melanocytic and smooth muscle marker expression is the hallmark of PEComas. Immunoreactivity for melanocytic markers include HMB-45 and melan A, and myogenic markers consist of smooth muscle actin, desmin, and h-caldesmon **(7, 9)**.

Evidence showed that the peak age for genital tract PEComa incidence is the mid-40s, although the reported patients with PEComa have had different ages from 9 to 79 years **(2)**. As the study case was 32 years (at the high-risk age), PEComa must be placed as a differential diagnosis in her ovary masses.

Although most PEComas behave as benign tumors, a few of these will be malignant. The characteristics of invasive tumors were indicated in former studies; the size of the tumor (greater than 5 cm), histological features, necrosis, and invasion to vessels are some of these predictor factors to malignancy **(10)**.

Because of rare cases reported and lack of clinical trial, the proper therapy for PEComa in the female genital tract is not available. There is only a strong consensus about tumor-free margin cytoreduction as a primary treatment, and chemotherapy and radiotherapy's usefulness is ambiguous. Recently, targeted therapies such as mTOR inhibitor usage seem to have promising results **(2, 11, 12)**.

Conclusion

Considering different clinical presentations, unknown characteristics of imaging, and rarity of PEComa, preoperative diagnosis of it seems impossible. However, IHC can play an important role in its diagnosis.

Acknowledgments

None.

Conflict of Interest

The authors declared no conflict of interests.

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