Pecoma of Uterus with Unique Morphological and Immunohistochemical Features

Dr. Surbhi Rajauria1, Dr. Ajamal Singh Bhayal2, Dr. Ramesh C Purohit3

1D.N.B. (Pathology).Assistant Professor, Department of Pathology, Dr. S.N. Medical College, Jodhpur, (Rajasthan).342001
2M. D. (Pathology) Associate Professor, Department of Pathology, RUHS College of Medical Sciences, sector 18 Pratap Nagar Jaipur (Raj) 302033.

Abstract: Perivascular epithelioid cells (PECs) are present in a group of tumor called PEComas including angiomyolipoma (AML), clear cell “sugar” tumor (CCST) of the lung and extrapulmonary sites, lymphangioleiomyomatosis, clear cell myomelanocytictumor of the falciform ligament/ligamentumteres and rare clear cell tumor of other anatomic sites. PEComas have distinct, morphologic and immunohistochemical characteristics, including perivascular epithelioid cells with clear to granular cytoplasm, a round to oval, centrally located nucleus,and an inconspicuous nucleolus. PEComas also express melanocytic and myogenic markers like HMB-45 and smooth muscle actin.We report a case of uterine PEComa.The patient presented with abnormal uterine bleeding and grossly mass in uterus. Histopathologically, the tumor was composed of thick blood vessels and perivascular epithelioid cells. The neoplastic cells were immunoreactive for HMB-45 antigen, Desmin, SMA and CD10-ve;Ki-67 labelling index was <1%. Uterine Pecoma should be considered a tumor of uncertain malignant potential and long term follow up required.

Keywords: Epithelioid cell, Perivascular, PEComa, Uterus

1. Introduction

Perivascular epithelioid cell (PEComa) refers to family of neoplasms showing at least partial morphological or immunohistochemical evidences of putative perivascular epithelioid cells (PEC) differentiation. PEComa may involve the organs, including the liver, kidney ,lung,uterus, small and large bowel, pancreas, vulva, heart and pelvic side wall/fallopian ligament. PEComas have clear to granular lightly eosinophillic cytoplasm and express the biomarkers of melanocytic and smooth muscle. The mean age of patients reportedly 54 years (range 45-75 years old)(Iarc press who soft tissue 2002).This entity is studied less, to quote and has many unanswered questions.

2. Case Report

A 52 years old female presented withmenometrorrhagia. Leiomyomatous uterus was suspected on the basis of clinical and ultrasonographic findings. Total abdominal hysterectomy was performed under laparoscopic guidance.

Pathological findings - Grossly, a solid soft tissue mass was present at the lower uterine segment and part of cervical wall measuring 3x3x2.5cm. Cutsurface was grey white with yellowish areas. Laparoscopically removed multiple solid yellowish white tissue collectively measured 12×5cm.

Microscopically, the sections from the mass and multiple solid tissue revealed tumor composed of round cells having perivascular disposition. Individual cell were having clear to eosinophillicmoderate cytoplasm with round nucleus. Mitotic figure was 2/10hpf. Stroma was hyalinised and showed focal calcification. Tumor margin were well defined. A diagnosis of PEComa/Epithelioid benign leiomyoblastoma was given.

Immunohistochemistry(IHC) was advised. IHC report was positive for HMB-45, Smooth muscle actin, desmin , negative for CD-10; Ki-67 labelling index was 2%.The final diagnosis of Pecoma of uterus was given.

3. Discussion

The term “PEComa”(Perivascular epithelioid cell tumor was introduced in 1996 by Zamboni et al.(2) as synonyms for tumor composed primarily of PECs.In the WHO soft tissue volume,PEComas are defined as “mesenchymaltumors composed histologically and immunohistochemically distinctive perivascular epithelioid cells.”(1)including entities such as angiomyolipoma(AML),lymph angioleiomyomatosis (LAM), clear cell sugar tumor (CCST) of lung.In recent years, more tumors are being reported and categorized as members of the PEComa family, including monotypic epitheloid AML, extrapulmonary CCST and clear cell myomelanocytictumor (CCMT) of the falciform ligament/ligament teres.

The first case of uterus PEComa tumor was reported by Pea et al.(3) PEComas share common morphophenotypic. features: perivascular disposition of epithelioid cells having clear to granular lightly eosinophillic cytoplasm and centrally placed round to oval nucleus with small nucleoli, spindled cells resembling smooth cells being away from the vessel; and immunoreactive for both the melanogenic-related markers and to lesser extent muscle markers. The mean age of uterine PEComas is 54 years (range 40-75 years old).In our case the age of the female patient was 52 years.

Uterine PEComas lack specific clinical and imaging changes, the diagnosis mainly relies on pathological approach and should be differentiated from the following
tumors.(a)Epitheloid smooth muscle tumors (ESMT)-the ESMT cells are round, polygonal and spindled shaped; they are usually HMB-45 negative, without characteristic capillarity network of blood vessels. PEComa is supplied by rich blood vessels and the tumor cells surround the blood vessels which are often HMB-45 positive.(b) Endometrial stromal sarcoma (ESS)- The tumor cells are spindle shaped with less cytoplasm and negative for HMB-45; PEComa cells are large and round or polygonal in shape, with eosinophilliccytoplasm, and are HMB-45 positive. Some PEComa patients also have lymphangiolyeniomyomatosis and tuberous sclerosis, which has not been reported in ESS patients. (c) Uterine leiomyosarcoma (UMS)-the tumor is composed of cells with clear cytoplasm with hobnail morphology arranged as solid nodules and papillary shapes being positive for cytokeratin (CK) and negative for HMB-45. (d) Metastatic renal cell carcinoma (MRC)- tumor cells have polygonal shape with clear cytoplasm with cells arranged in nests, alveolar, ductal or papillary shapes but with no perivascular structure, the tumor is HMB-45 negative and CK and EMA positive. (e) Paraganglioma-it should be differentiated with PEComa when the cytoplasm is clear. Paraganglioma cells are arranged in streaks, glands, or nests, with flat supporting cells lying around, besides they are HMB-45 negative, and neuron specific enolase (NSE), synaptophysin and NF positive, the supporting cells are positive for S-100 positive. (4)

PEComas have been reported as predominantly benign. The 2002 WHO soft tissue and bone book states that PEComas having the following features: infiltrative growth, marked hypercellularity, nuclear enlargement, hyperchromasia, high mitotic activity, atypical mitotic figures, and coagulative necrosis should be regarded as malignant. (1) The real behaviour of these tumors is uncertain as some tumor with “benign” appearance have aggressive behaviour and others with “malignant” appearance have indolent course. Late recurrences of the tumor have been reported, including one with lung metastasizing 7 years after the primary tumor. (5) There was no recurrence in our case. So the uterine PEComas should be considered tumor of uncertain malignant potential and long term follow up is required.

### References


### Author Profile


**Dr. Ajamal Singh Bhayal**, M. D. (Pathology) Associate Professor, Department of Pathology, RUHS College of Medical Sciences. 109 B, Rajendra Nagar, Shyam Path, Sirsi Road, Jaipur (Raj) 302021.


---

**Fig. 1** Gross appearance of tumor

**Fig. 2** H & E 4X

**Fig. 3** H & E 10X

**Fig. 4** H & E 40X

**Fig. 5** SMA Positive

**Fig. 6** HMB45 Positive

**Fig. 7** CD1a Negative

**Fig. 8** Pan CK Negative

---

**Volume 4 Issue 9, September 2015**

[www.ijsr.net](http://www.ijsr.net)