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Pecoma of Uterus with Unique Morphological and Immunohistochemical Features

Dr. Surbhi Rajauria[1], Dr. Ajamal Singh Bhayal[2], Dr. Ramesh C Purohit[3]

[1]D.N.B. (Pathology).Assistant Professor, Department of Pathology, Dr. S.N. Medical College, Jodhpur, (Rajasthan).342001

[2]M. D. (Pathology) Associate Professor, Department of Pathology, RUHS College of Medical Sciences, sector 18 Pratap Nagar Jaipur (Raj) 302033.

[3]M.D. (Path. & Bact.), F.I.C.P.,I.F.C.A.P.; R.P. Diagnostic Centre, Opp. Sardar Children School, Hakim Bagh, Jodhpur, (Raj.).342001

Abstract: ***Perivascular epitheloid 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 epitheloid cells with clear to granular cytoplasm, a round to oval, centrally located nucleus,and an inconsipicuous 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 epitheloid 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: Epitheloid cell, Perivascular, PEComa, Uterus

1. Introduction

Perivascular epitheloid cell (PEComa) refers to family of neoplasms showing at least partial morphological or immunohistochemical evidences of putative perivascularepitheloid cells (PEC) differentiation. PEComa may involve the organs, including the liver, kidney ,lung,uterus, , small and large bowel, pancreas, ,vulva ,heart andpelvic side wallfalciform ligament. PEComas have clear to granular lightly eosinophillic cytoplasm and express the biomarkers of melanocytic and smooth muscle. The mean age of patients reportedly 54years(ranging from 45-75years old)(iarc press who soft tissue2002).This entity is studied less, to quote and has many unanswered questions.

2. Case Report

A 52 years old female presented withmenometrorhagia. Leiomyomatous uterus was suspected on the basis of clinical and ultrasonographicfindings.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/Epitheloid 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 epitheloid cell tumor was introduced in 1996 by Zamboni et al.(2) as synonymns for tumor composed primarily of PECs.In the WHO soft tissue volume,PEComas are defined as "mesenchymaltumors composed histologically and immunohistochemically distinctive perivascular epitheloid cells."(1)including entites 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 myomelonocytictumor (CCMT) of the falciform ligament/ligament teres.

The first case of uterus PEC tumor was reported by Pea et al.(3) PEComas share common morphophenotypic.
features: perivascular disposition of epitheloid 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 (range40-75 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-45positive.Some PEComa patients also have lymphangioleiomyomatosis and tuberous sclerosis,which has not been reported in ESS patients.(c)Uterine clear cell carcinoma-the tumor is composed of cells with clear cytoplasm with hobnail morphology arranged as solid ductal or papillary shapes being positive for cytokeratin(CK) and negative for HMB-45.(d)Metastatic renal cell carcinoma-tumor cells have polygonal shape with clear cytoplasm with cells arranged in nest, alveolar, ductal or papillary shapes but with no perivessel structure, thetumor 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, nuclearenlargement, 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 metasizing 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.


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

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Author Profile

Dr. Surbhi Rajauria, c/o Lt.Col. Prashant Rajauria. Medical Officer, M.H. Wellington ,Nilgiri District. Tamilnadu.643231.

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.

Dr. Ramesh C Purohit, M.D. (Path. & Bact.), F.I.C.P.,I.F.C.A.P. ; R.P. Diagnostic Centre, Opp. Sardar Children School, Hakim Bagh, Jodhpur, (Raj.).342001