

## Perivascular Epithelioid Cell Neoplasm (Pecoma) of the Uterus: A Case Report and Review of the Literature

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## 1. Summary

Uterine perivascular epithelioid cell tumors (PEComa) are rare lesions, composed of cells with mesenchymal, epithelioid and smooth muscle differentiation with expression of melanocytic markers [1].

Clinical symptoms and imaging findings are usually non-specific and diagnosis is confirmed by histological analyses. Due to the metastatic potential of PEComas, complete surgical resection followed by close oncological follow-up is crucial in the therapeutic workup [2]. This case report presents a 55-years-old woman with an incidental finding of a uterine PEComa with a diameter of more than 6 cm of unknown malignant potential.

## 2. Abstract

**2.1. Background:** Perivascular epithelioid cell tumors (PEComas) of the uterus encompass a rare family of mesenchymal neoplasms characterized by co-expression of smooth muscle and melanocytic markers in epithelioid or spindle cells. Due to their unclear malignant potential, a close follow-up after surgical resection is mandatory.

**2.2. Case Presentation:** We present the case report of a 55-year-old woman who underwent hysterectomy due to the diagnosis of a uterus myomatosis with growing tendency. Preoperative imaging

by ultrasonography and magnetic resonance imaging scan (MRI) did not show specific findings. A uterine PEComa was diagnosed by morphology and immunohistology.

**2.3. Conclusion:** Uterine PEComas are extremely rare tumors, which, in most cases, are diagnosed incidentally based on specific morphological and immunohistochemical features.

## 3. Introduction

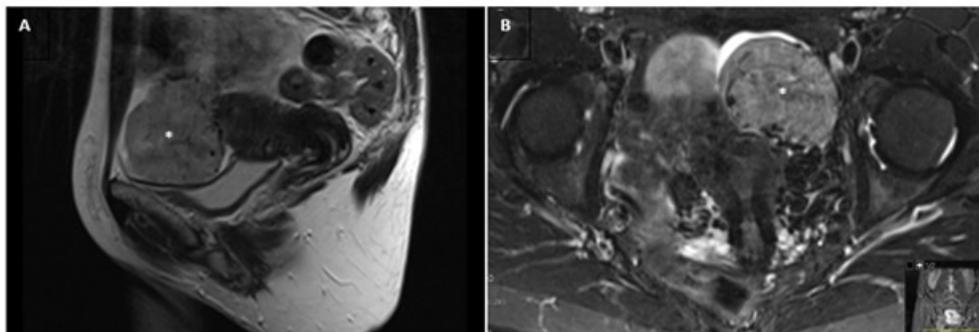
In 1992, Bonetti et al. published the first report of a lesion belonging to the heterogenic family of neoplasms with perivascular epithelioid cell tumors (PEComa) in the lung and kidney [3]. However, PEComas can be found in various locations including the complete urogenital tract and even in bones. Uterine PEComas, first discovered in 1996 by Pea et al., are classified as sarcomas but only account for less than five percent of uterine cancers [4]. According to the histopathologic classification by the World Health Organization (WHO) in 2003, there are four main types of uterine sarcomas: leiomyosarcomas, endometrial stromal sarcomas, adenocarcinomas and other sarcomas of the uterus, such as PEComas [5].

## 4. Case Report

A 55-year-old woman without pre-existing medical conditions was admitted to our hospital due to unspecific lower abdominal pain

and menorrhagia. Treatment with a progestin mono-therapy led to amenorrhea and pain relief. Preoperative ultrasound scan and pelvic magnetic resonance imaging (MRI) showed multilocular uterine fibroids with a growth tendency and a 6 cm subserosal solid and inhomogeneous, partially enhancing mass on the MRI scan, adjacent to the left side of the uterine fundus. The mass appeared

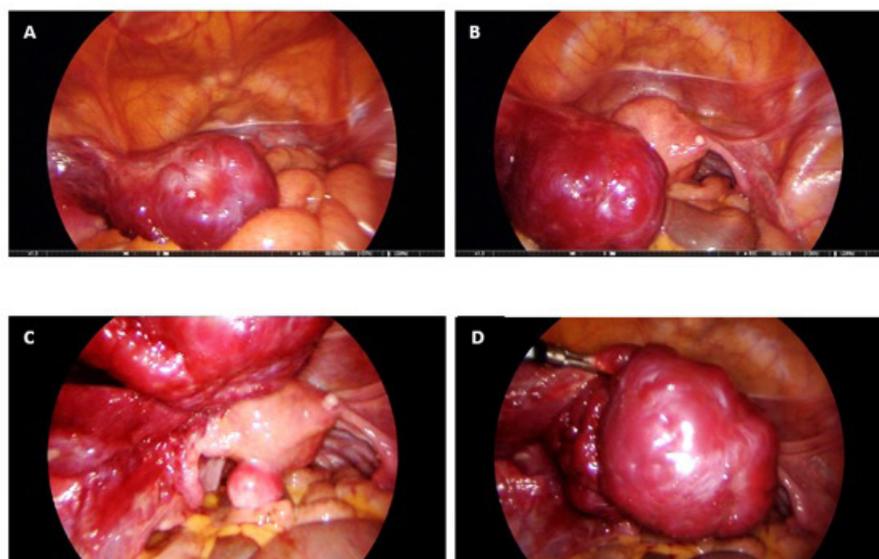
as non-infiltrating, with a well-circumscribed border with signs of hyper vascularization. The endometrial morphology was normal (Figure 1). Both, the fallopian tubes and the ovaries showed no abnormalities in the imaging procedures. We performed a total laparoscopic hysterectomy with bilateral opportunistic adnexectomy. The tumor was locally resected in sano.



**Figure 1:** Preoperative pelvic MRI findings of a suspect subserosal mass in the lower abdomen originating from the uterus  
 A: Sagittal T2-weighted image shows a heterogeneous partially enhancing subserosal mass on fundus of the uterus  
 B: Axial fat-suppressed contrast-enhanced T1 weighted image shows the enhancement of the well-circumscribed subserosal mass with a regular shape

Due to safety considerations and because of the atypical vascularization of the lesion with a wide vessel pedicle reaching caudal from the uterus wall to the left round ligament, the uterus was removed in toto under strict oncologic safety aspects. Because of the

unknown dignity, the excision borders of the tumor were chosen as wide as possible. Subsequently, the uterus and the adnexa were placed in a laparoscopic surgery retrieval pouch and removed in toto through the vagina (Figure 2).



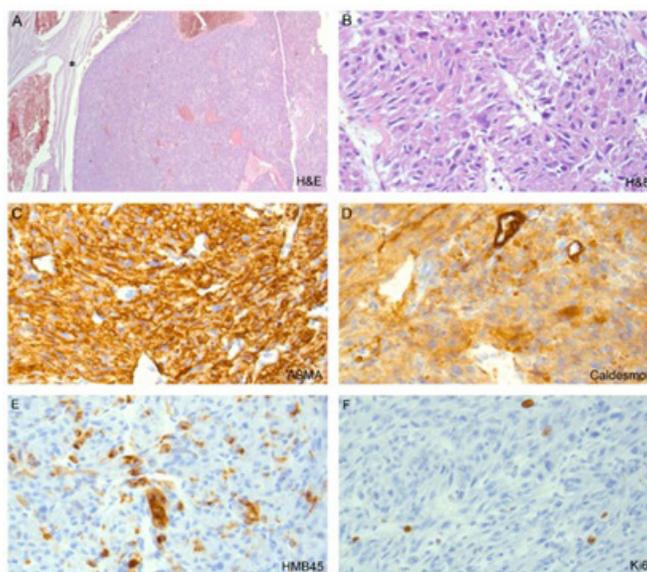
**Figure 2:** Intraoperative view during totally laparoscopic hysterectomy.  
 \*: suspect mass  
**Figure A:** A: Laparoscopic view of the lower abdomen  
**Figure B:** Mobile, broadly pedunculated, vulnerable tumor originating from the left round ligament  
**Figure C:** Posterior view of the tumor. The mass is adjacent to the left round ligament and is not connected to other organs.  
**Figure D:** The tumor is strikingly soft and vulnerable.

Gross pathological analysis demonstrated 6.5 x 4.5 x 4.0 cm tumor with smooth borders and surrounded by a shiny surface. The lesion presented itself as an encapsulated rubbery mass with a lobulated, irregular cutting surface in tan-pink and brown due to foci of hemorrhage.

Microscopical analysis of the solid lesion showed polygonal plump spindle cell to epithelioid cells and moderate nuclear size variations, as well as abundant blood vessels of varying caliber. The

tumor mass had neither necrosis nor infiltrative growth. The tumor cells exhibited oval nuclei with prominent nucleoli and abundant granular slightly eosinophilic cytoplasm (Figure 3 (A)+(B)).

Immunohistochemical analyses were consistently positive for smooth muscle actin and diffusely positive for caldesmon (Figure 3 (C)+(D)). The tumor cells showed patchy expression of HMB45 (approximately 30% of tumor cells) (Figure 3 (E)). The proliferation activity of tumor cells (Ki67) was less than 5%, and only a low mitotic rate was detected (Figure 3 (F)).



**Figure 3:** Histopathology of the PEComa.

A: Low magnification (40x) of tumor tissue section with the tumor capsule (\*) on the left (H&E staining) B: High magnification (400x) of the tumor showing high vessel density with radially arranged spindle cells with granular eosinophilic cytoplasm (H&E staining). C-F: Immunohistochemical staining of the PEComa shows coexpression of both the smooth muscle markers C: alpha smooth-muscle actin (ASMA), as well as of D: caldesmon and E: the melanocytic marker HMB45. F: Low Ki67-(MIB1) proliferation rate. \*: tumor capsule

## 5. Discussion

According to the WHO, PEComas are defined as a rare subtype of tumors of mesenchymal origin, composed of epithelioid cells with eosinophilic and partly granulated cytoplasm, which are characterized by immunohistochemical co-expression of melanocytic (e.g. HMB45, melan A and S100) and smooth muscle markers (e.g. smooth muscle actin, desmin and caldesmon) [5, 6]. PEComas occur most frequently in the abdominopelvic cavity, the retroperitoneum and the uterus [7, 8]. To date, approximately 150 cases of PEComas of different localizations were described, but only 90 cases originated from the gynecological tract. The clinicopathological characteristics of uterine PEComas are summarized in (Table 1). Two different molecular subtypes of PEComas have been identified. Most PEComas are characterized by a loss of function of the TSC1/TSC2 complex and in majority a loss off heterozygosity (LOH) in the TSC2 gene, subsequently leading to increased mTORC1 activation and deregulated cell growth signal-

ing [9]. Additionally, a distinct small second subset of PEComas were identified which showed no association to tuberous sclerosis, but presented rearrangements of the TFE3(Xp11) gene locus [10]. The TFE3-translocated PEComas display an epithelioid phenotype and attenuated or missing expression of myogenic markers [11]. Mesenchymal neoplasms composed of perivascular epithelioid cells present a heterogeneous group of tumors including Clear-Cell «Sugar» Tumor (CCST) of the lung and extrapulmonary sites, AngioMyoLipoma (AML), LymphAngioleioMyomatosis (LAM), as well as clear-cell epithelioid and spindle cell tumors of other anatomical sites [12]. Ten percent of PEComas are associated with the rare multisystem disorder LAM, which in most cases affects the lungs [13]. LAM can occur either in association with the tuberous sclerosis complex (TSC-LAM) or, in sporadic cases, without tuberous sclerosis (sporadic LAM) [14]. Less than ten percent of uterine PEComas are associated with tuberous sclerosis.

**Table 1:** Uterine PEComas – clinicopathological characteristics

|   |   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
|---|---|---|---|------------------|-----------------------------|---|--------------------------------|----------|-------------------|------|-----|---------|-----|------------------------------|
| <b>Definition</b><br>[4, 5]                 | Perivascular epithelioid cell tumors (PEComas) encompass a family of mesenchymal neoplasms consisting of perivascular cells with epithelioid, partially granulated cytoplasm with melanocytic markers, that can occur in any part of the human body   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Epidemiology</b><br>[2, 11]              | Rare mesenchymal neoplasms<br>- women: men = 7:1 in all localisations<br>- mostly premenopausal women with a peak incidence within the fourth decade of life > approximately 90 cases of gynecological PEComas described to date<br>- gynecological PEComas accounted for 25% of PEComa cases   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Pathogenesis</b><br>[9, 13, 14]          | - 10% association with tuberous sclerosis complex (TSC) or lymphangioliomyomatosis (LAM)<br>- uterus as the source of LAM cells<br>- uterus as a target for metastasis rather than the primary source<br>- biallelic TSC2 mutations indicates alternative tumorigenic pathways  |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Pathology</b><br>[16, 17, 30, 31]        | <b>Macroscopy</b>   | <b>Microscopy:</b>  | <b>Immunohistochemistry</b>   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
|   | - majority arise in the uterine corpus the cervix is less frequently involved   | - epithelioid and spindle cells typically arranged around blood vessels which grow in sheets or nests<br>- epithelioid cells are typically localized periluminal, while peripheral cells are more spindle-shaped<br>- cells with small round to oval nuclei without prominent nuclei with clear to eosinophilic cytoplasm<br>- uterine PEComas show variable amounts of stromal hyalinization | - immunoreactive for both smooth muscle and melanocytic markers<br><br>- <b>immunophenotype:</b><br><table border="1"> <tr> <td>HMB45</td> <td>100%</td> </tr> <tr> <td>smooth muscle actin</td> <td>73%</td> </tr> <tr> <td>vimentin</td> <td>56%</td> </tr> <tr> <td>CD10</td> <td>25%</td> </tr> <tr> <td>Melan-a</td> <td>24%</td> </tr> <tr> <td>h-caldesmon cathepsin k TFE3</td> <td>&lt;10%</td> </tr> </table> | HMB45            | 100%                        | smooth muscle actin                         | 73%                            | vimentin | 56%               | CD10 | 25% | Melan-a | 24% | h-caldesmon cathepsin k TFE3 |
| HMB45                                       | 100%  |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| smooth muscle actin                         | 73%   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| vimentin                                    | 56%   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| CD10  | 25%   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| Melan-a                                     | 24%   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| h-caldesmon cathepsin k TFE3                | <10%  |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Differential diagnosis</b>               | - epithelioid smooth muscle tumors of the uterus<br>- epithelioid stroma sarcomas<br>- paragangliomas   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Diagnostic procedures</b>                | - clinical presentation is non-specific and varies depending on the localization<br>- Ultrasound, Computed tomography scans (CT) and magnetic resonance imaging (MRI) are mostly used as complementary imaging  |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Classification</b> [22]                  | Histological malignancy criteria (proposed by Folpe et al.):<br><br><table border="1"> <tr> <td>Tumor size ≥ 5cm</td> </tr> <tr> <td>Infiltrative growth pattern</td> </tr> <tr> <td>Mitotic rate &gt; 1/50 high power fields (HPF)</td> </tr> <tr> <td>High nuclear grade cellularity</td> </tr> <tr> <td>Necrosis</td> </tr> <tr> <td>Vascular invasion</td> </tr> </table><br>Three categories:<br><i>benign</i> : no criteria for malignancy<br><i>uncertain malignant potential</i> : one criterium for malignancy<br><i>malignant</i> : ≥ 2 criteria for malignancy |   |   | Tumor size ≥ 5cm | Infiltrative growth pattern | Mitotic rate > 1/50 high power fields (HPF) | High nuclear grade cellularity | Necrosis | Vascular invasion |      |     |         |     |                              |
| Tumor size ≥ 5cm                            |   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| Infiltrative growth pattern                 |   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| Mitotic rate > 1/50 high power fields (HPF) |   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| High nuclear grade cellularity              |   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| Necrosis                                    |   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| Vascular invasion                           |   |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Staging</b><br>[20, 21]                  | in analogy with uterine (leiomyo-) sarcomas with a thoracic and an abdominal CT scan  |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Therapy</b><br>[26, 28, 29]              | - no unanimous consensus<br>- surgical excision as mainstay of primary treatment<br>- in PEComas with aggressive behavior or in metastatic disease:<br><input type="checkbox"/> Possible adjuvant or palliative chemotherapy regimens: Adriamycin, high-dose Ifosfamide, Trabectedin, Docetaxel/Gemcitabine, dacarbazine-based regimens<br><input type="checkbox"/> radiation therapy<br><input type="checkbox"/> targeted immunotherapies with mTOR inhibitors (e.g. Sirolimus, Temsirolimus or Everolimus)  |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |
| <b>Prognosis</b><br>[32, 33]                | Relapse: 20%<br>Lethality: 10%  |   |   |                  |                             |   |                                |          |                   |      |     |         |     |                              |

PEComa: Perivascular epithelioid cell tumor; LAM: Lymphangioliomyomatosis; TSC: tuberous sclerosis complex; HPF: high power fields

The spectrum of possible differential diagnoses for uterine PEComas include endometrial stromal sarcomas, epithelioid smooth muscle tumors and paragangliomas. PEComas can display several histomorphological characteristics and morphological diversities. Sclerosing PEComas for example show stromal hyalinization. Further microscopic features can consist of the detection of multinucleated cells, spider cells and stromal microcysts [15]. Typically, the cells grow in sheets or nests and display impressive vascular invasion [6]. The mitotic rate is usually between 10 and 50 HPFs, and atypical mitoses are frequently observed [16]. Positive staining for HMB45 and h-caldesmon are typical immunohistochemical findings in PEComas. The lack of staining for HHF35, desmin, alpha-smooth muscle actin, CD10, as well as negative staining for synaptophysin, chromogranin A and S-100 protein can favor the diagnosis of PEComa [17].

PEComas may originate from any area of the gynecological tract, but in most cases they were diagnosed in the uterine corpus of premenopausal women. The diagnosis of a PEComa is challenging, due to the lack of specific clinical symptoms or distinct radiological findings, and in most cases, asymptomatic small tumors were discovered accidentally. The most common but non-specific symptoms are atypical uterine bleeding or lower abdominal pain, but clinical manifestations may vary depending on the location and dimensions of the tumor [18].

Commonly used preoperative diagnostic methods are ultrasound and MRI or computed tomography scans (CTs), although imaging features of PEComas are nonspecific. Tan et al. published a large imaging series of 32 cases of malignant PEComas, and described significant enhancement in CT and MRI scans [19]. According to previously cited literature, the preoperative imaging procedures performed in our case suggested the diagnosis of multiple uterine leiomyomas with increased vascularization but without specific signs of malignancy. Additionally, gross pathologic examination showed no suspicious signs for malignancy. In summary, PEComas are diagnosed in most cases by a combination of histopathological features and immunohistochemical findings after surgical excision.

The treatment workup is based on the therapy algorithm for uterine (leiomyo-) sarcomas. No further therapy is indicated after complete surgical resection and if distant metastases were ruled out. Due to the rarity of PEComas, an accurate assessment of prognosis is difficult. Fortunately, approximately 90% of patients had a favorable prognosis. Screening procedures for distant metastases should include both a thoracic and abdominal CT scan [20, 21].

Folpe and Mentzel proposed a classification of PEComas based on morphological high-risk features. A size of  $\geq 5$  cm, infiltrative growth pattern, a mitotic rate of  $\geq 1$ /HPF, high nuclear grade and cellularity, as well as necrosis and vascular invasion were defined as potential malignancy criteria with a significant association with

recurrences [22, 23]. For clinical and therapeutic purposes, PEComas are classified in analogy to the FIGO- or TNM-classification system [2, 24]. Distant metastases are present in approximately 7% of patients at the time of diagnosis. In most cases, they are localized in the liver, the lungs, the bones or the brain. Recurrences occur in one fifth of patients despite the majority of low-grade lesions [2]. Thus, a close oncologic follow-up with surveillance imaging is highly recommended. Adjuvant therapy is not indicated after complete local excision of PEComas. In sporadic case reports heterogeneous results were achieved with various adjuvant chemotherapy treatment regimens like dacarbazine, ifosfamide, doxorubicine or vincristine were tested [25]. In case of relapse or metastasis, neoadjuvant or adjuvant chemotherapy, radiation therapy and immunotherapy regimens were described [26, 27]. In PEComas expressing hormone receptors, antiestrogenic treatment is still a matter of controversy. Nevertheless, a significant clinical response has been previously described after treatment with mTOR inhibitors (e.g. sirolimus, temsirolimus or everolimus) as an impairment of TSC2 gene function has been associated with the mTOR signaling pathway in extragenital angiomyolipomas of the kidneys [28, 29].

## 6. Conclusion

Perivascular epithelioid cell tumors (PEComa) are rare tumors of mesenchymal origin and can affect various anatomic regions. Most of these lesions consist of epithelioid cells with eosinophilic granulated cytoplasm and present a co-expression of smooth muscle and melanocytic markers. Uterine PEComas are part of uterine sarcomas, mostly diagnosed incidentally. Optimal treatment is controversially discussed, although surgical resection remains the gold standard. Systemic treatment is warranted in high risk or metastatic disease. Our patient demonstrates, that a rare tumor diagnosis demanded an interdisciplinary approach.

## References

1. Oliva E, Zaloudek CJ, Soslow RA. Mesenchymal Tumors of the Uterus. In: Blaustein's Pathology of the Female Genital Tract. Springer International Publishing. 2019. 535-647.
2. Horn LC, Schmidt D, Fathke C, Ulrich U. New FIGO staging for uterine sarcomas [Internet]. Vol. 30, Pathologie. Springer Verlag. 2009; 30: 302-3.
3. Bonetti F, Pea M, Martignoni G, Zamboni G. PEC and Sugar. American Journal of Surgical Pathology. Am J Surg Pathol. 1992; 16: 307-8.
4. Martignoni G, Pea M, Reghellin D, Zamboni G, Bonetti F. Perivascular epithelioid cell tumor (PEComa) in the genitourinary tract. Adv Anat Pathol. 2007; 14: 36-41.
5. TAVASSOLI FA. Pathology and Genetics of Tumours of the Breast and Female Genital Organs. World Health Organ Classif Tumours. 2003.

6. 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.
7. Gentile M, Zinna M, Zanella C, Costanza A, Dalfior D, Sina S, et al. Uterine PEComa with aggressive behavior: A review with an additional case of spontaneous vaginal expulsion. *Pathology Research and Practice*. Elsevier GmbH. 2020; 216: 152991.
8. Bennett JA, Oliva E. Perivascular epithelioid cell tumors (PEComa) of the gynecologic tract. *Genes Chromosomes and Cancer*. Blackwell Publishing Inc.; 2021; 60: 168-79.
9. Fang CL, Lin YH, Chen WY. Microscopic endometrial perivascular epithelioid cell nodules: a case report with the earliest presentation of a uterine perivascular epithelioid cell tumor. *Diagn Pathol*. 2012; 7(1): 117.
10. Argani P, Aulmann S, Illei PB, Netto GJ, Ro J, Cho HY, et al. A distinctive subset of PEComas harbors TFE3 gene fusions. *Am J Surg Pathol*. 2010; 34(10): 1395-406.
11. Utpatel K, Calvisi DF, Köhler G, Kühnel T, Niesel A, Verloh N, et al. Complexity of PEComas: Diagnostic approach, molecular background, clinical management. *Pathologie*. 2020; 41: 9-19.
12. Hornick JL, Fletcher CDM. PEComa: What do we know so far? *Histopathology*. *Histopathology*; 2006; 48: 75-82.
13. Grzegorek I, Drozd K, Podhorska-Okolow M, Szuba A, Dziegiel P. LAM cells biology and lymphangiomyomatosis. *Folia Histochem Cytobiol*. 2013; 51(1): 1-10.
14. Hayashi T, Kumasaka T, Mitani K, Terao Y, Watanabe M, Oide T, et al. Prevalence of uterine and adnexal involvement in pulmonary lymphangiomyomatosis: A clinicopathologic study of 10 patients. *Am J Surg Pathol*. 2011; 35(12): 1776-85.
15. Thway K, Fisher C. PEComa: Morphology and genetics of a complex tumor family. *Annals of Diagnostic Pathology*. 2015; 19: 359-68.
16. Bennett JA, Braga AC, Pinto A, Van De Vijver K, Cornejo K, Pesci A, et al. Uterine PEComas: A Morphologic, Immunohistochemical, and Molecular Analysis of 32 Tumors. *Am J Surg Pathol*. 2018; 42(10): 1370-83.
17. Fukunaga M. Perivascular epithelioid cell tumor of the uterus: Report of four cases. *Int J Gynecol Pathol*. 2005; 24(4): 341-6.
18. Prasad SR, Sahani D V., 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: 688-96.
19. Tan Y, Zhang H, Xiao EH. Perivascular epithelioid cell tumour: Dynamic CT, MRI and clinicopathological characteristics - Analysis of 32 cases and review of the literature. *Clin Radiol*. 2013; 68(6): 555-61.
20. Ramaiah S, Ganesan R, Mangham DC, McNally O, Klys HS, Hirschowitz L. Malignant variant of sclerosing perivascular epithelioid cell tumor arising in the Adnexa. *Int J Gynecol Pathol*. 2009; 28(6): 589-93.
21. Dimmler A, Seitz G, Hohenberger W, Kirchner T, Faller G. Late pulmonary metastasis in uterine PEComa. *J Clin Pathol*. 2003; 56(8): 627-8.
22. Folpe AL, Mentzel T, Lehr HA, Fisher C, Balzer BL, Weiss SW. Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: A clinicopathologic study of 26 cases and review of the literature. *American Journal of Surgical Pathology*. 2005; 29: 1558-75.
23. Jafari A, Fischer HP, Von Websky M, Hong GS, Kalff JC, Manekeller S. Primary perivascular epithelioid cell tumour (PEComa) of the liver: Case report and review of the literature. *Z Gastroenterol*. 2013; 51: 1096-100.
24. Prat J. FIGO staging for uterine sarcomas. *International Journal of Gynecology and Obstetrics*. 2009; 104: 177-8.
25. 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.
26. Machado I, Cruz J, Lavernia J, Rayon JM, Poveda A, Llobart-Bosch A. Malignant PEComa with Metastatic Disease at Diagnosis and Resistance to Several Chemotherapy Regimens and Targeted Therapy (m-TOR Inhibitor). *Int J Surg Pathol*. 2017; 25(6): 543-9.
27. Jeon IS, Sung ML. 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.
28. Bissler JJ, Kingswood JC, Radzikowska E, Zonnenberg BA, Frost M, Belousova E, et al. Everolimus for angiomyolipoma associated with tuberous sclerosis complex or sporadic lymphangiomyomatosis (EXIST-2): A multicentre, randomised, double-blind, placebo-controlled trial. *Lancet*. 2013; 381(9869): 817-24.
29. Kenerson H, Folpe AL, Takayama TK, Yeung RS. Activation of the mTOR pathway in sporadic angiomyolipomas and other perivascular epithelioid cell neoplasms. *Hum Pathol*. 2007; 38(9): 1361-71.
30. D'Angelo E, Prat J. Diagnostic use of immunohistochemistry in uterine mesenchymal tumors. *Semin Diagn Pathol*. 2014; 31(3): 216-22.
31. Fadare O. Uterine PEComa: Appraisal of a controversial and increasingly reported mesenchymal neoplasm. *Int Semin Surg Oncol*. 2008; 5: 7.
32. Konsultationsfassung S3-Leitlinie Adulte Weichgewebesarkome.
33. Horn LC, Teichmann G, Einkenkel J. Das PECom des uterus - Ein seltener mesenchymaler tumor: Fallbericht und literaturübersicht. *Geburtshilfe Frauenheilkd*. 2011; 71(2): 140-3.