DOI: https://doi.org/10.5114/pjp.2017.67623 Pol J Pathol 2017; 68 (1): 92-95

Quiz

Correct answer to the quiz. Check your diagnosis

Perirenal perivascular epithelioid cell tumor (PEComa) coexisting with other malignancies: a case report

Marian Danilewicz[1], Janusz M. Strzelczyk[2], Małgorzata Wagrowska-Danilewicz[3]

[1]Department of Pathology, Medical University of Lodz, Lodz, Poland
[2]Department of General and Transplant Surgery, Medical University of Lodz, Lodz, Poland
[3]Department of Nephropathology, Medical University of Lodz, Lodz, Poland

Perivascular epithelioid cell tumor (PEComa) is a very rare lesion and is described by the World Health Organization (WHO) as a mesenchymal tumor composed of histologically and immunohistochemically distinctive perivascular epithelioid cells. In this report we describe PEComa with perirenal manifestation, which is exceedingly rare and to our best knowledge up to now worldwide only three cases have been described. Despite the reports that most PEComas are benign, this tumor met criteria for malignancy and coexisted with mucinous gallbladder cancer and nonresectable pancreatic head tumor. We concluded that despite the rarity of perirenal PEComas, in cases with an unusual epithelioid histological pattern the diagnosis of PEComa should also be taken into consideration on the basis of the immunohistochemical study.

Key words: perivascular epithelioid cell tumor, immunohistochemistry.

Introduction

The descriptive name perivascular epithelioid cells (PEC) was introduced by Bonetti *et al*. in 1992, whereas Zamboni *et al*. in 1996 proposed the term perivascular epithelioid cell tumor (PEComa) [1, 2, 3]. PEComa is a very rare lesion and is described by the World Health Organization (WHO) as a mesenchymal tumor composed of histologically and immunohistochemically distinctive perivascular epithelioid cells [4]. On the immunohistochemical basis the cells characteristically express HMB45 but are negative for S100 protein. They have an uncertain origin and the histological pattern is not uniform [3]. The PEComa family of tumors includes angiomyolipomas, clear cell "sugar" tumors of the lung, lymphangiomyomatosis and clear cell myomelanocytic tumors [5]. The PEComa with perirenal manifestation is exceedingly rare and to our best knowledge up to now worldwide only three cases have been described. Notably, none of these tumors coexisted with other malignancies.

Case report

A 66-year-old woman presented to the Department of General and Transplant Surgery with pancreatic head tumor, 66 × 45 mm in size. Furthermore, a perirenal tumor 15 × 10 mm close to the superior pole of the right kidney was found (Fig. 1). She was scheduled for laparotomy, and during the surgery the tumor of the pancreas was found to be nonresectable. It was involving the common hepatic artery with multiple enlarged lymph nodes in the hepatoduodenal ligament. The perirenal tumor was resected, and hemostasis of the pole of the kidney was obtained using Tachosil. Additionally

cholecystectomy was performed as multiple gallstones were present in the gallbladder and the its wall was thick. The postoperative period was uneventful and the patient was discharged on the 5th postoperative day.

Macroscopically the perirenal tumor consisted of a considerably fragile tissue fragment measuring 1.5 × 1.0 × 0.6 cm. Additionally, from the same patient the gallbladder with thick walls was submitted for histopathological examination. Routine hematoxylin-eosin stained sections from formalin-fixed, paraffin-embedded specimens from both materials were examined. Histologically, the perirenal tumor was composed of polygonal clear cells or cells with eosinophilic cytoplasm with moderate epithelioid appearance (Fig. 2). In most cells nuclear polymorphism was prominent. Up to 2-3 mitoses per 10 high-power fields were present. Furthermore, the tumor showed abundant vascularity (Fig. 3). The resection was microscopically incomplete. The specimens taken from the gallbladder showed mucinous cancer. Initially, the perirenal tumor was diagnosed as malignant neoplasm and further immunohistochemical study was performed. Cytokeratin AE1/AE3, cytokeratin 20, cytokeratin 7, CD10 and protein S-100 were negative, whereas we noted positivity of HMB-45 (++) (Fig. 4), α smooth actin (+++), vimentin (++), estrogen receptor α (++) and focally desmin (+). Nuclear immunoexpression of Ki-67 was present in 8% of tumor cells. The small vessels were revealed using CD34 antibody. The histological examination and immunohistochemical study fulfilled the criteria for the diagnosis of malignant PEComa with perirenal manifestation coexisting with mucinous gallbladder cancer and nonresectable pancreatic head tumor.

Discussion

To date, the extremely rare perivascular epithelioid cell tumor has been described in various anatomic locations: in lungs [1], pancreas [2], uterus [6, 7], bone [8], retroperitoneum [9], ovary [10], liver [11],

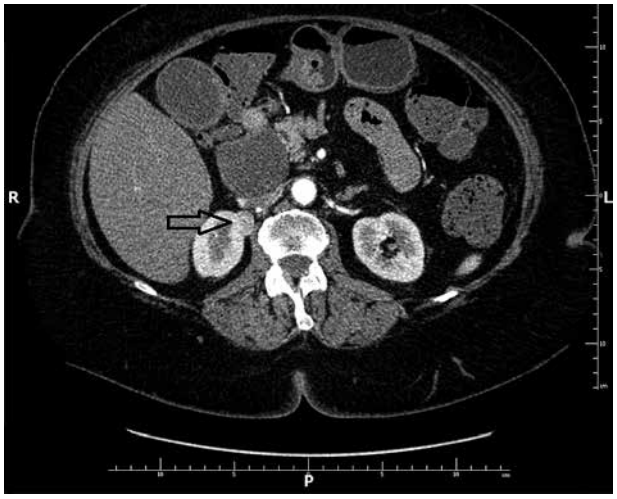

Fig. 1. Abdomen CT revealed perirenal mass close to the superior pole of the right kidney

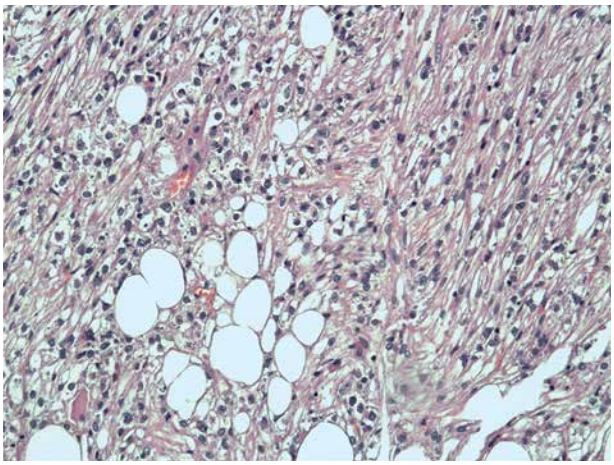

Fig. 2. Tumor composed of polygonal clear cells or cells with eosinophilic cytoplasm with moderate epithelioid appearance. HE, magnification 100×

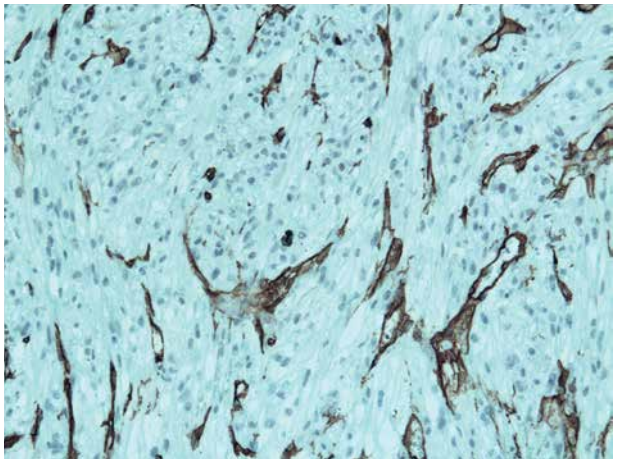

Fig. 3. A large number of the small vessels can be seen between tumor cells. CD34 stain, magnification 100×

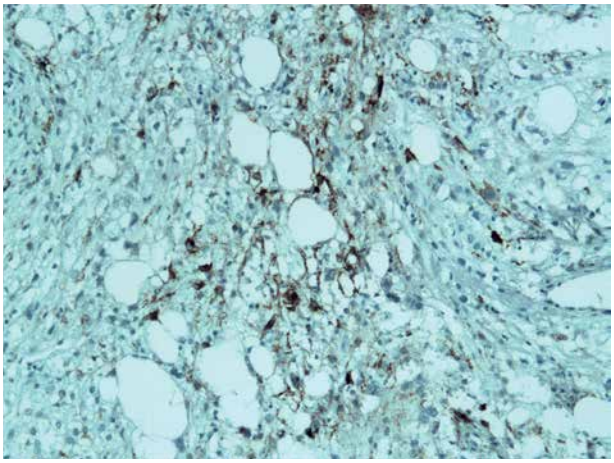

Fig. 4. Focal HMB-45 positivity in the tumor cells. Magnification 100×

cutis [12], transverse colon [4] and in intraorbital location [3]. As was mentioned above, close perirenal location concerns only three cases of PEComas [5, 13, 14]. In the present case a perirenal mass was found during examination of the pancreatic malignancy, but it should be stressed that imaging findings of perirenal PEComas are usually nonspecific, as various other retroperitoneal neoplasms present comparable features [5, 14].

Although the tumor cells have uncertain origin, they manifest distinctive histological and immunohistochemical features. Histological constant findings are represented by a nested architecture composed of epithelioid cells typically surrounded by thin capillary vessels [3]. In our case the histological pattern was in agreement with these findings. Despite the report that most PEComas are benign [4], this tumor met criteria for malignancy presented by Folpe *et al*. [15] as mitotic activity >1/50 HPF, high nuclear grade and cellularity and infiltrative growth. Noteworthy, in the opinion of some authors [5] currently there is no evidence that mitotic activity might serve as a parameter for reliably predicting biological behavior in PEComas.

However, immunohistochemical study plays a crucial role in avoiding misdiagnosis of PEComas. As these tumors are mesenchymal of uncertain lineage, in our study cytokeratin AE1/AE3, cytokeratin 20 and cytokeratin 7 were entirely negative, whereas we noted positivity for vimentin, and focally for desmin. Unusually, the presence of focal cytokeratin expression in malignant PEComa of the fibula was noted [8]. In the present case immunohistochemical staining confirmed the melanocytic and myoid differentiation of the tumor cells. Similarly as in the study of Gunia *et al*. [5], epithelioid cells were focally immunoreactive for HMB-45, whereas Greene *et al*. reported this immunostaining as diffuse [6]. On the other hand, similarly as in the last cited study diffuse positive staining with smooth muscle actin reflected the myoid differentiation. The immunoreactivity for the proliferation marker Ki-67 in these studies was also similar (5% and 8%). In agreement with others [3, 16] the tumor cells in our case were S-100 negative. Contrary to us and to most reports, some authors reported patchy S-100 staining of PEComa cells [6, 11]. In the present study the immunoexpression of CD-10 was negative in the tumor cells, although in some cases of PEComas positivity for CD-10 was noted. In these patients the differential diagnosis with metastasis of renal cell carcinoma may be difficult [12, 17]. Interestingly, in our case we revealed moderate nuclear immunoexpression of estrogen receptor alpha. Similarly, immunoreactivity for estrogen receptor was noted in one case of this tumor with perirenal manifestation as well as in 33% of cases in a series of 41 PEComas of the uterus [5, 7]. On the other hand, another recurrent PEComa of the uterus was for this receptor entirely negative [6].

In conclusion, despite the rarity of perirenal PEComas, in cases with an unusual epithelioid histological pattern the diagnosis of PEComa should also be taken into consideration on the basis of immunohistochemical study.

This work was supported by Medical University of Lodz grant no. 503/1-038-01/

The authors declare no conflict of interest.

References

1. Bonetti F, Pea M, Martignoni G, et al. PEC and sugar. Am J Surg Pathol 1992; 16: 307-308.
2. Zamboni G, Pea M, Martignoni G, Zancanaro C et al. Clear cell "sugar" tumor of the pancreas. A novel member of the family of lesions characterized by the presence of perivascular epithelioid cells. Am J Surg Pathol 1996; 20: 722-730.
3. Lubo I, Fermín I, Massarelli O, et al. Perivascular epithelioid cell tumour with intraorbital location: report of a case and review of the literature. Case Rep Pathol 2016; 2016: 1936421.
4. Baek JH, Chung MG, Jung DH et al. Perivascular epithelioid cell tumor (PEComa) in the transverse colon of an adolescent: a case report. Tumori 2007; 93: 106-108.
5. Gunia S, Awwadeh L, May M, et al. Perivascular epithelioid cell tumor (PEComa) with perirenal manifestation. Int J Urol 2005; 12: 489-492.
6. Greene LA, Mount SL, Schned AR, et al. Recurrent perivascular epithelioid tumor of the uterus (PEComa): an immunohistochemical study and review of the literature. Gynecol Oncol 2003; 90: 677-681.
7. Fadare O. Perivascular epithelioid cell tumor (PEComa) of the uterus: an outcome-based clinicopathologic analysis of 41 reported cases. Adv Anat Pathol 2008; 15: 63-75.
8. Lian DW, Chuah KL, Cheng MH, et al. Malignant perivascular epithelioid cell tumour of the fibula: a report and a short review of bone perivascular epithelioid cell tumour. J Clin Pathol 2008; 61: 1127-1129.
9. Hornick JL, Fletcher CD. Sclerosing PEComa: clinicopathologic analysis of a distinctive variant with a predilection for the retroperitoneum. Am J Surg Pathol 2008; 32: 493-501.
10. Rampisela D, Grossmann P, Donner LR. Rhabdoid myomelanocytic tumor (PEComa) of the ovary: a clinically benign case followed for 7 years. Int J Surg Pathol 2016; 24: 431-435.
11. Strzelczyk JM, Durczynski A, Szymanski D, et al. Primary perivascular epithelioid cell tumor (PEComa) of the liver: report of a case. Surg Today 2009; 39: 916-921.
12. Fernandez-Flores A, Nguyen CM, Cassarino DS. Cutaneous PEComas Express CD10: Implications for the classification of PEComas and the differential diagnosis with metastatic renal cell carcinoma. Am J Dermatopathol 2016; 38: 645-652.
13. Nakanishi S, Miyazato M, Yonemori K, et al. Perirenal malignant perivascular epithelioid cell tumor originating from right retroperitoneum: a case report. Hinyokika Kiyo 2014; 60: 627-630.
14. Gkizas CV, Tsili AC, Katsios C, et al. Perirenal PEComa: Computed Tomography Findings and Differential Diagnosis. J Clin Imaging Sci 2015; 5: 69.
15. Folpe AL, Mentzel T, Lehr HA, et al. Perivascular epithelioid cell neoplasms of soft tissue and gynecologic origin: a clinicopathologic study of 26 cases and review of the literature. Am J Surg Pathol 2005; 29: 1558-1575.

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: 1-13.
17. Koutlas IG, Pambuccian SE, Jessurun J, et al. Perivascular epithelioid cell tumor of the oral mucosa. Arch Pathol Lab Med 2005; 129: 690-693.

Address for correspondence

Prof. Marian Danilewicz
Department of Pathology
Medical University of Lodz
Pomorska 251
92-213 Lodz, Poland
Tel./fax +48 42 633 90 13
e-mail: marian.danilewicz@umed.lodz.pl