An Aggressive Case of Malignant Renal Pecoma Non-Responsive to Motor Inhibition: A Case Report

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Abstract
Perivascular epithelioid cell tumors (PEComas) are made up of cells with distinctive morphologic, immunohistochemical, ultrastructural and genetic features. They may arise in various locations and are usually benign. Malignant PEComas are rare and associated with an aggressive clinical course and metastatic spread. We present a case of a 56 years old healthy patient who presented with right flank pain and a large mass located at the medial aspect of the right kidney. The mass ruptured during laparoscopic removal. The tumor was eventually labeled as an epithelioid malignant neoplasm most suggestive of malignant PEComa. The patient demonstrated during follow up both local and systemic metastasis that are unresponsive to this point to mTOR inhibition with various agents.

Keywords: Everolimus; mTOR; Pecoma; Renal; Votrient

Introduction
Perivascular epithelioid cell tumors (PEComas) are made up of cells with distinctive morphologic, immunohistochemical, ultrastructural and genetic features. They may arise in various locations and are usually benign. Malignant PEComas are rare and associated with an aggressive clinical course and metastatic spread.

We present such a case with bone, skin and retroperitoneal metastasis, non-responsive to surgical and adjuvant mTOR inhibition.

Case Report
A 56-year-old, generally healthy caucasian male with no significant past medical history started complaining of continuous right flank pain starting April 2013. The patient had no urinary complaints of any sort. Physical examination was normal and blood work and urine analysis were within normal limits. An abdominal ultrasound revealed a hypoechoic mass adjacent to the right kidney causing hydronephrosis of an intermediate degree, and a widened proximal right ureter. The mass was irregular and 7*8 cm in size with many anechoic centers and displayed no blood flow. The urinary bladder was of normal thickness with no stones or residual urine.

CT urography performed demonstrated the mass as a well circumscribed 11 cm mass located at the medial aspect of the left pole of the right kidney. The mass had nodular thickenings that show enhancement. The mass is pressing on the right collecting system, the duodenum and the head of the pancreas. The urinary bladder was normal in appearance and there was no lymphadenopathy. The patient underwent open resection of the mass. A flank incision was performed between the 11th and the 12th rib including removal of the 12th rib. The mass was located at the area of the right pedicle causing rotation the kidney upwards while pressing the right ureter. During the separation of the mass it ruptured releasing fluid and clots. The tumor involved the right upper ureter requiring resection of the ureter followed by closure over a ureteral stent. The patient was discharged 4 days later with no complications and the ureteral stent was withdrawn 6 weeks later.

Microscopic examination revealed a cystic tumor measuring 8 cm in diameter, composed of large atypical cells with eosinophilic and clear cytoplasm, organized in diffuse pattern, trabecular, ribbon like and nesting pattern. Scattered mitotic figures were present (3/50 HPF). Foci of tumoral necrosis was also present. Ki-67 proliferation index was about 5% to 6%. Capsular invasion was identified but no vascular invasion was seen. Focally the tumor was present less than 0.1 cm from the surgical margin.

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Received August 20, 2016; Accepted October 12, 2016; Published October 20, 2016


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metastasis foci stable in size and location for only 6 months. Since then the patient has changed several regimens of treatment including Palbociclib and Aredia, Adriamycin and currently Gemzar without durable effect.

**Discussion**

Epithelioid Angiomyolipoma (Perivascular epithelioid cell tumor [PEComa]) of the kidney is a mesenchymal lesion very closely related to the classic angiomyolipoma. A couple of hundred cases are published, with mean age group of approximately 40 years and male to female ratio of 1:1 [1,2]. The perivascular epithelioid cell, or PEC, appears in a certain group of tumors and has its typical morphological, immunohistochemical and genetical characteristics [3,4]. It has been speculated that the PEComa can modulate its morphology and immunophenotype and appear in a spectrum of forms [4,5], making the diagnosis, as in our case, somewhat challenging. PEComas of the kidney include several entities among them the renal epithelioid angiomyolipoma (or renal PEComa) recognized by the WHO in the 2004 classification of kidney tumors [6]. Peri-renal PEComas as in our case are extremely rare making the differential diagnosis even more difficult [7]. Prior to the recognition of this variant many cases were wrongfully labeled as renal cell carcinoma. This entity is recognized by a unique Immunohistochemical staining for melanocytic markers like HMB45 and smooth muscle markers and not for epithelial markers. Most PEComas are benign and surgical resection with an adequate margin remains the standard treatment of PEComa. But, looking at the literature there have been several reports of distant metastases after surgical resection of renal PEComas with poor prognosis [4,8]. Even though there was spillage during the initial open surgery, one can't ignore the poor prognostic characteristics of the tumor of size and necrosis [1] and the rapid clinical progression. Although it has been proven that PEComas respond to mTOR inhibition [9,10], it seems that our patient shows no such response and we continue to look for a solution on his behalf.

**Acknowledgements**

There is no funding to report for this submission.

**Conflicts of Interest**

There are no conflicts of interest to disclose.

**References**

