

## Oncology

## Huge retroperitoneal epithelioid angiomyolipoma: A case report

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## ARTICLE INFO

## Article history:

Received 9 September 2017

Received in revised form

8 November 2017

Accepted 21 November 2017

Available online 6 December 2017

## 1. Introduction

Angiomyolipoma (AML) is a type of tumor in the perivascular epithelioid cell neoplasm (PEComa) family. Epithelioid angiomyolipoma (EAML), having malignant potential, is considered a rare variant of angiomyolipoma. The most common site of EAMLs is kidney, and extra-renal EAMLs are very uncommon, with liver being the most common site.<sup>1</sup> Other locations including retroperitoneum, uterus, liver, lung, breast, cardiac septum, pancreas, prostate, and gastrointestinal tract have also been described.<sup>2</sup> To our acknowledgment, only 6 cases of retroperitoneal EAML was reported in the English literature.<sup>3</sup> Presented here is a 46-year-old female with a giant (20 cm × 15 cm × 15cm) retroperitoneal epithelioid angiomyolipoma.

## 2. Case presentation

A 46-year-old Taiwanese female patient visited Chang Gung Memorial Hospital general surgery out-patient department due to a palpable mass over left lower quadrant of her abdomen. The mass was palpated by herself 2 months ago. Her past medical history was unremarkable and she denied any associating symptoms including abdominal pain or recent weight loss. There were no signs or symptoms compared with tuberous sclerosis. As for family history, her aunt and uncle suffered from colon cancer and both her parents have hypertension. During laboratory tests, blood routine shows

microcytic anemia (Hb: 9.9 g/dL, MCV 79 fL). Other lab data including renal function, liver function, electrolytes were all within normal range. CT images of the abdomen shows a huge perirenal cystic tumor below left kidney, 20 cm × 15 cm × 15cm in size. The tumor shows heterogeneous enhancement after contrast injection. It is encapsulated with soft tissue with smooth surface, lack grossly visible fat and displacing the left kidney upwards and compressing abdominal aorta (Fig. 1). Biopsy was done showing some large polygonal cells with eosinophilic cytoplasm and nuclear pleomorphism, favors angiomyolipoma (see Fig. 2).

Owing to the tumor size and high risk of haemorrhage, exploratory laparotomy excision of the tumor with renal sparing was carried out without incident by general surgery surgeon. The cystic mass was decompressed for 1300 ml of blood and was dissected carefully from the left kidney. Pathology revealed tumor composed largely of polygonal epithelioid cells with abundant eosinophilic cytoplasm, marked nuclear pleomorphism and hyperchromasia, and occasional bizarre tumor giant cells. Mitotic figures are seen 0–2/10 HPF. Mature fat cells, thick-walled vessels, and spindle smooth muscle cells are also seen in a minor portion. No identifiable renal tissue is seen. Hence, the tumor is most likely arising from the renal capsule. Further Immunohistochemical study shows the tumor is diffusely and strongly positive for Human Melanoma Black-45 (HMB45), smooth muscle actin, and vimentin, negative for cytokeratin (AE1/AE3) and epithelial membrane antigen (EMA). The diagnosis of epithelioid angiomyolipoma was confirmed.

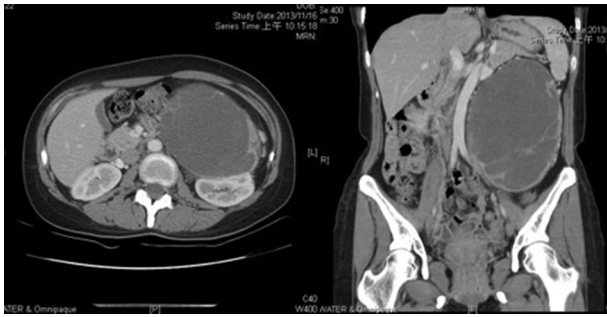
The patient recovered uneventfully and was discharged 5 days later. Follow-up CT urography and creatinine (0.62 mg/dL) after three years later revealed no evidence of recurrence or metastasis until this writing.

## 3. Discussion

Angiomyolipoma (AML) was clinically considered to be a benign lesion due to the lack of recurrence and metastasis. Nevertheless, epithelioid AML (EAML), a rare variant of AML composed of epithelioid cells and scarce adipose tissue, has been reported to pursue a more aggressive clinical course.<sup>4</sup> Epithelioid AML of retroperitoneum is even rarer in literature. Among patients with retroperitoneal EAMLs, the average age was 46 years (ranging from 22 to 80 years) with a male: female ratio of 1–5.3. Two cases of

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**Fig. 1.** CT image of the abdomen with contrast: Heterogeneous enhancement of the tumor after contrast injection, encapsulated with soft tissue with smooth surface and displacing the left kidney upwards and compressing abdominal aorta.

metastasized retroperitoneal EAML have been reported: the first one being a 29-year-old man with tuberous sclerosis complex and retroperitoneal EAML metastasized to liver and mediastinum, the second one being an 80-year-old woman with EAML metastasized to bone and liver.<sup>5</sup>

Owing to the huge tumor size and cystic content of our case, rupturing of the EAML, compression of great vessels are also potential threats that need to be taken into consideration. Thus, preoperative evaluation including imaging, and biopsy is of great significance. The differential diagnosis of a mass with scarce fat in retroperitoneal space may include renal cell carcinoma (RCC) with retroperitoneal spread, malignant fibrous histiocytoma, lymphangioma, paraganglioma, and neurofibroma. Immunoreactivity to an HMB-45 stain positive is a useful tool to differentiate from

other retroperitoneal tumors except RCC and lymphangioma. Moreover, the negative for AE1/AE3 and EMA can rule out renal cell carcinoma and lymphangioma. The confirmed diagnosis of EAML can make patient free from over treatment like partial or radical nephrectomy.

The management of EAML is surgical resection due to malignant potential. Other paper revealed that all patients had remained disease-free and asymptomatic at last follow-up and no recurrence had been documented after a renal sparing nephrectomy or embolization. Folpe, A.L., et al. have proposed an empirical-based criteria for evaluating malignancy of PEComa. The authors divide PEComas into three categories (Benign, uncertain malignant potential, malignant) with characteristics including the tumor size, mitotic rate, high or non-high nuclear grade, cellularity, presence of infiltration, necrosis or vascular invasion. The present case is categorized as malignant (diameter > 5cm, mitotic activity > 1/50 HPF). Hence, long term follow-up is necessary.<sup>5</sup>

#### 4. Conclusion

In summary, EAML of perirenal origin is an unusual entity, which is a potentially malignant tumor that has imaging appearance similar to other retroperitoneal masses. It should be considered in the differential diagnosis for a retroperitoneal mass by biopsy, especially in view of a possible renal-sparing treatment.

#### Informed consent

Permission for publication and informed consent from the patients has been documented in the paper.

#### Conflict of interest statement

None.

#### Acknowledgment

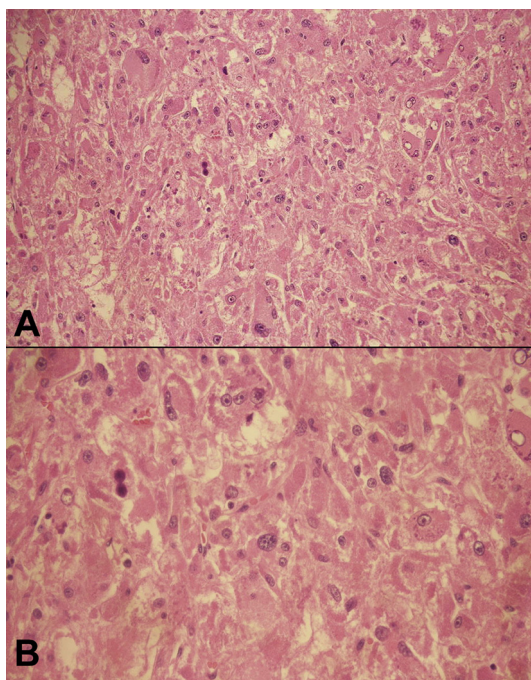
Any institution has not financed this case report.

#### Appendix A. Supplementary data

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.eucr.2017.11.016>.

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**Fig. 2.** Marked variation in cellular size and shape, with nuclear hyperchromasia and giant cell formation. Abundant cytoplasm is eosinophilic and granular. Occasionally, tumor cells with two or more nuclei can be found. (A) Hematoxylin-phloxine-saffron stain, original magnification  $\times 200$ . (B) Hematoxylin-phloxine-saffron stain, original magnification  $\times 400$ .