Case Report

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Multiloculated Cystic Type Renal Epithelioid Angiomyolipoma Mimicking Renal Cell Carcinoma: A Case Report

신세포암으로 오인된 다방성 낭종 형태의 신장의 유상피 혈관근지방종: 증례 보고

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Renal epithelioid angiomyolipoma (EAML) is a rare variant of angiomyolipoma (AML), with a prominent epithelioid component. EAML usually presents as a large heterogeneous soft tissue lesion with intratumoral hemorrhage and variable necrosis or cystic changes. We present a case of multiloculated cystic renal EAML mimicking renal cell carcinoma in a 64-year-old female. Intracystic massive hemorrhage, hyperattenuating wall and septa on an unenhanced study, and enlarged intratumoral vessels can be helpful imaging features for distinguishing renal EAML from renal cell carcinoma.

Index terms Angiomyolipoma; Computed Tomography, X-Ray; Magnetic Resonance Imaging; Renal Cell Carcinoma

INTRODUCTION

Renal epithelioid angiomyolipoma (EAML) is a rare variant of angiomyolipoma (AML), which is a potentially malignant mesenchymal neoplasm (1). Rarely, renal EAML can present as multilocular cystic type. Herein, we report an unusual case of multiloculated cystic type renal EAML mimicking renal cell carcinoma in a 64-year-old female.

CASE REPORT

A 64-year-old female presented with a palpable huge mass in the left abdominal region which was first noticed 1 year ago. During a physical examination, the mass was firm, non-tender and fixed. The patient stated that the mass was gradually increasing in size. She had no significant medical history other than hypertension and dyslipidemia.

CT scan revealed a 13.2 cm × 11.3 cm × 19.8 cm size well-defined multilocular cystic mass in the interpolar region of the left kidney, compressing the residual renal parenchyma medially, and abutting to the pancreas tail portion. On unenhanced scan, the lesion showed thickened wall and septa, which was hyperattenuating of about 50 Hounsfield unit (HU) (Fig. 1A, left). CT densities of cystic parts of variably sized each locules are higher than that of water, ranging from 20–40 HU, suggesting hemorrhage and/or proteinaceous fluid. Neither areas of calcification nor macroscopic fat were detected within the lesion. On enhancement study, irregular thickened wall and septa revealed strong enhancement in the corticomedullary phase. There were multiple prominent enlarged vascular structures along the wall and cystic septa (Fig. 1A, middle). On MRI scan, the intracystic components were mostly occupied by high signal intensities on T1-weighted images, corresponding to areas of hemorrhage (Fig. 1B, upper left). Contrast-enhanced MRI showed marked enhancement of the wall and septa with thick distorted vessels along the cystic septa (Fig. 1B, upper right). Vascular flow voids along the cystic septa were noted on T2-weighted images (Fig. 1B, lower right). Some parts of the lesion showed the restricted diffusion on diffusion weighted images. Considering the large size, irregularly thick enhancing wall and septa of the mass, our initial differential diagnosis was clear cell renal cell carcinoma with extensive cystic changes.

Radical nephrectomy was performed. During the surgery, a huge yellowish mass arising from the middle pole of the left kidney was noted. Grossly, the tumor had multilocular cystic changes with intra cystic blood clots, measuring 17.0 cm × 14.0 cm × 5.5 cm (Fig. 1C, left). Microscopically, the tumor was composed of predominantly epithelioid cells with dense eosinophilic cytoplasm, nuclear pleomorphism. Immunohistochemical analysis revealed diffuse positive staining of HMB-45 marker, but was negative for S-100 protein, epithelial membrane antigen, and cytokeratin (Fig. 1C, middle, right). These pathological features were consistent with EAML. Post-operative recovery was uneventful, and the patient was discharged on the tenth postoperative day. She remained well at three years of follow-up with no evidence of tumor recurrence.

The study protocol conformed to the ethical guidelines of the 1975 Declaration of Helsinki.

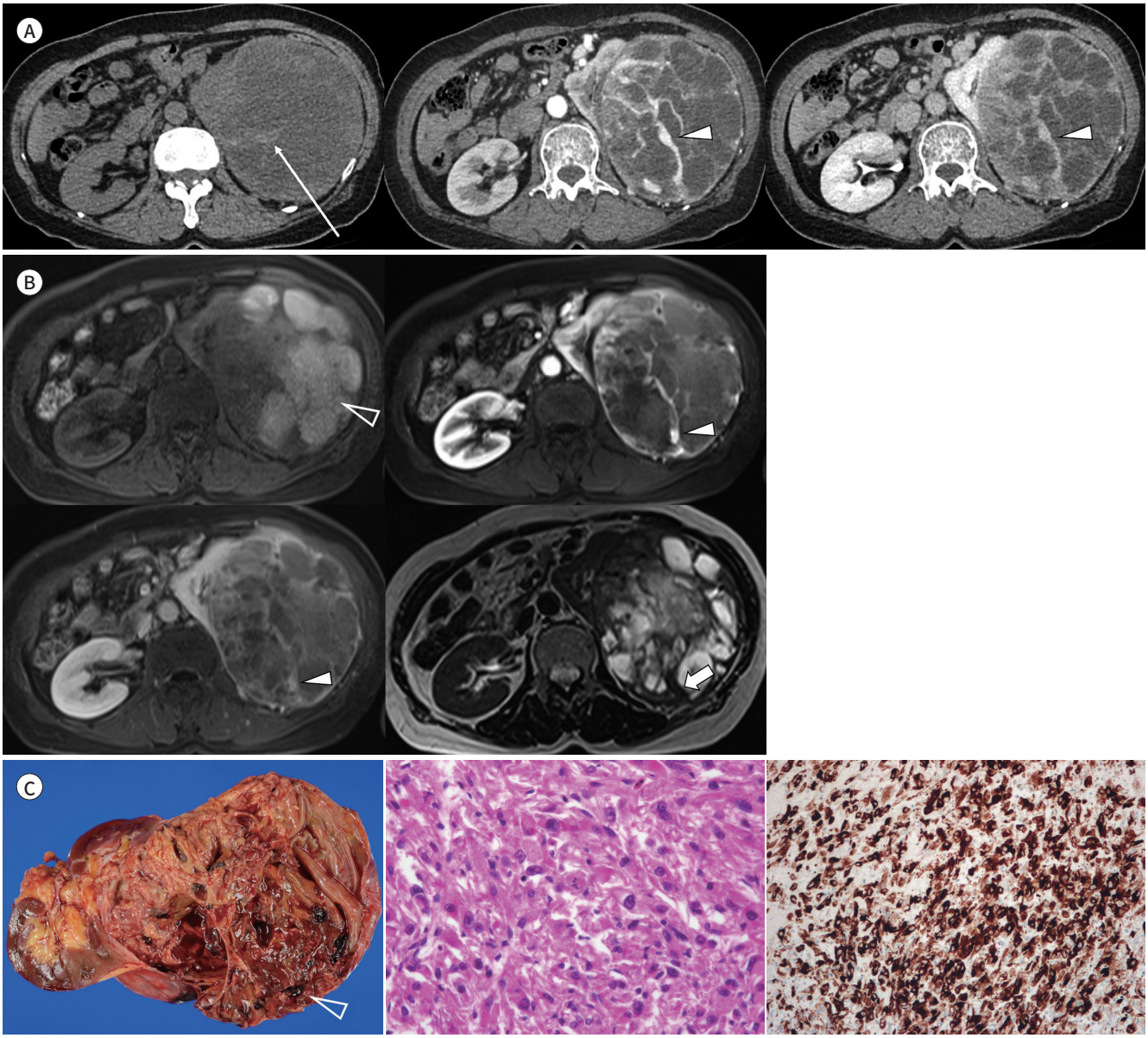
DISCUSSION

Renal AML is the most common benign mesenchymal renal tumor composed of varying amount of dysmorphic blood vessels, smooth muscles cells and adipose tissue. Renal EAML is a rare variant of AML with histologically predominant epithelioid cells and minimal or no adipose tissue (1). Renal EAML was classified as a new category of mesenchymal tumor in the 2016 World Health Organization classification of renal tumors (2). It is considered potentially malignant neoplasm with possible metastasis and local recurrence (1). It belongs to a

family of perivascular epithelioid cell tumors (PEComas), which may occur sporadically or in association with tuberous sclerosis complex (TSC) (3).

Renal EAML can present as heterogeneous solid-type, homogeneous solid-type or multi-

Fig. 1. A 64-year-old female with multiloculated cystic type renal epithelioid angiomyolipoma, mimicking renal cell carcinoma.
A. CT scans show a large well-defined multilocular cystic mass arising from the interpolar region of the left kidney. Pre-contrast axial CT (left image) shows hyperattenuating walls and septa (arrow). Dynamic enhanced CT shows marked enhancement with enlarged vascular structures (arrowheads) along the septa during the corticomedullary phase (middle image) and washout during the excretory phase (right image).
B. On T1-weighted MR images (upper left image), the intracystic components (open arrowhead) show high signal intensities. The cystic septa show marked enhancement with enlarged vessels (arrowheads) during the corticomedullary phase (upper right image) and washout during the nephrogenic phase (lower left image). On T2-weighted images (lower right image), vascular flow voids of engorged intratumoral vessels are observed along the cystic septa (arrow).
C. Gross specimen (left image) shows a large multilocular cystic mass filled with blood clots (open arrowhead). Histologic examination (middle image) shows the proliferation of epithelioid cells with abundant eosinophilic cytoplasm (hematoxylin and eosin stain, × 400). Immunohistochemical staining (× 400) for HMB-45 (right image) is diffusely positive.



locular cystic type. Heterogeneous solid type lesions are typically accompanied by intratumoral hemorrhage and variable necrosis or cystic changes (4). On unenhanced CT scan, most EAML are hyperattenuating due to their abundant epithelioid components. The enhancement patterns are heterogeneous with rapid wash-in and slow wash-out (5). On MRI scan, the tumor has tendency to be low signal intensity on T2-weighted images, due to their muscle components. On the dynamic enhancement study, it shows inhomogeneous marked enhancement in the corticomedullary phase and rapid wash-in and wash-out pattern. It shows restricted diffusion on diffusion weighted images. Cong et al. (6) suggested the presence of hypointensity on T2-weighted images, microscopic fat component, reticular enhancement pattern and lower apparent diffusion coefficient value are helpful imaging findings to differentiate from clear cell renal cell carcinoma.

Rarely, renal EAML can present as multilocular cystic type like our case. It is still unclear whether this type of lesion was initially a multilocular figure or the result of necrosis or recurrent hemorrhage. Multilocular cystic type EAML tends to show intracystic high signal intensity on T1 weighted image due to massive hemorrhage. Tsukada et al. (4) suggested that intracystic massive hemorrhage and hyperattenuating wall or septa on unenhanced CT images are useful imaging features to distinguish the cystic renal cell carcinoma. In our patient, intracystic components are mostly high signal intensity on T1 weighted image and the wall and septa showed hyperattenuating on unenhanced CT images, which are consistent with the previous studies.

Additionally, enlarged intratumoral vessels could be characteristic imaging features of renal EAML (7). Those radiologic features are also seen in hepatic EAML, suggesting unique feature of epithelioid subtype (8). In our case, prominent intratumoral vessels along the cystic septa are noted.

Surgical resection is treatment of choice due to potentially malignant behavior of EAML. Because it is challenging to distinguish EAML from renal cell carcinoma by preoperative imaging findings, surgical removal is usually performed. Recent studies have found that EAML is activated through the mechanistic target of rapamycin (mTOR) pathway, and there have been case reports with tumor responding dramatically to mTOR inhibitors such as sirolimus or temsirolimus (9).

In summary, we present a case of multilocular cystic type renal EAML mimicking renal cell carcinoma. Intracystic massive hemorrhage, hyperattenuation of the wall and septa, and prominent intratumoral vascularity can be helpful imaging features to distinguish from the renal cell carcinoma.

Author Contributions

Conceptualization, S.J.W.; data curation, S.J.W.; investigation, K.B.; project administration, S.J.W.; resources, S.J.W.; supervision, S.J.W.; visualization, K.B.; writing—original draft, K.B.; and writing—review & editing, S.J.W.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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REFERENCES

1. Park JH, Lee C, Suh JH, Kim G, Song B, Moon KC. Renal epithelioid angiomyolipoma: histopathologic review, immunohistochemical evaluation and prognostic significance. *Pathol Int* 2016;66:571-577
2. Moch H, Cubilla AL, Humphrey PA, Reuter VE, Ulbright TM. The 2016 WHO classification of tumours of the urinary system and male genital organs—part A: renal, penile, and testicular tumours. *Eur Urol* 2016;70:93-105
3. Martignoni G, Pea M, Reghellin D, Zamboni G, Bonetti F. PEComas: the past, the present and the future. *Virchows Arch* 2008;452:119-132
4. Tsukada J, Jinzaki M, Yao M, Nagashima Y, Mikami S, Yashiro H, et al. Epithelioid angiomyolipoma of the kidney: radiological imaging. *Int J Urol* 2013;20:1105-1111
5. Liu Y, Qu F, Cheng R, Ye Z. CT-imaging features of renal epithelioid angiomyolipoma. *World J Surg Oncol* 2015;13:280
6. Cong X, Zhang J, Xu X, Zhang M, Chen Y. Renal epithelioid angiomyolipoma: magnetic resonance imaging characteristics. *Abdom Radiol (NY)* 2018;43:2756-2763
7. Zhong Y, Shen Y, Pan J, Wang Y, An Y, Guo A, et al. Renal epithelioid angiomyolipoma: MRI findings. *Radiol Med* 2017;122:814-821
8. Ji JS, Lu CY, Wang ZF, Xu M, Song JJ. Epithelioid angiomyolipoma of the liver: CT and MRI features. *Abdom Imaging* 2013;38:309-314
9. Wolff N, Kabbani W, Bradley T, Raj G, Watumull L, Brugarolas J. Sirolimus and temsirolimus for epithelioid angiomyolipoma. *J Clin Oncol* 2010;28:e65-e68

신세포암으로 오인된 다방성 낭종 형태의 신장의 유상피 혈관근지방종: 증례 보고

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신장의 유상피 혈관근지방종은 혈관근지방종의 드문 변이형으로 현저한 유상피세포로 구성되어 있다. 유상피 혈관근지방종은 일반적으로 종양 내부의 출혈과 다양한 정도의 괴사 혹은 낭성 변화를 동반한 크기가 큰 불균일한 연조직 종괴로 나타난다. 우리는 64세 여성에서 발견된 신세포암으로 오인된 다방성 낭종 형태의 신장의 유상피 혈관근지방종에 대해 발표하려 한다. 낭종 내 다량의 출혈, 조영 전 검사에서 고음영의 벽과 격막, 그리고 비대된 종괴 내 혈관들이 신세포암을 감별하는 데 있어 도움이 될 수 있다.

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