Case Report

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Renal Epithelioid Angiomyolipoma with Epithelial Cysts Mimicking Cystic Renal Cell Carcinoma: A Case Report of Combination of Two Rare Entities

상피낭종을 동반한 신장의 상피모양 혈관근지방종: 두 희귀 질환의 조합에 대한 증례 보고

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Renal angiomyolipomas (AMLs) are typically solid tumors, but there have been few reports of a rare cystic variant of AML. AML with epithelial cysts, where the epithelial cyst has a cuboidal epithelial lining, account for the majority of them. Next, epithelioid AML (EAML) with cystic changes due to hemorrhage and necrosis, which is composed of epithelioid cells with abundant eosinophilic cytoplasm, have also been reported. These rare cystic types of AML can be mistaken for other cystic tumors, such as cystic renal cell carcinoma, in preoperative imaging. We report the imaging findings of a rare case of EAML with epithelial cysts.

Index terms Angiomyolipoma; Kidney; Neoplasms; Computed Tomography, X-Ray; Magnetic Resonance Imaging

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INTRODUCTION

Angiomyolipoma (AML) is a common triphasic neoplasm composed of various proportions of dysmorphic blood vessels, smooth muscle, and adipose tissue. It is usually diagnosed by detecting its fat component on a CT or MRI scan (1). Epithelioid AML (EAML) is an extremely rare subtype of AML that is characterized predominantly by proliferation of epithelioid cells and has the potential for malignancy unlike other subtypes (1, 2).

Renal AML, including EAML, is representative of a solid renal neoplasm, and cystic change in it is very rare. Therefore, it is usually excluded from the differential diagnosis of predominantly cystic lesions (3). A rare variant of AML called AML with epithelial cysts (AMLEC) or cystic AML was first reported in two case series in 2006 (4, 5). To date, fewer than 30 cases of renal AMLEC have been reported in 10 publications (3). In addition, EAML with cystic changes has rarely been reported. Most of the cystic portion of EAML is caused by hemorrhage or necrosis (2, 6). To our knowledge, there has been only one case of the rare combination of EAML and AMLEC, and no report has focused on the imaging findings of the lesion (2).

Herein, we report a case of an EAML with epithelial cysts that was misdiagnosed as cystic renal cell carcinoma (RCC) in a 58-year-old male.

CASE REPORT

A 58-year-old male was referred to our hospital for further evaluation of a right renal tumor discovered incidentally during a routine health checkup. He had no urinary symptoms and no related medical history. Ultrasonography and CT had been performed at another medical center. The US scan revealed a 2 cm predominantly cystic lesion in the interpolar area of the right kidney, with an eccentric solid area in the posterior aspect of the lesion (Fig. 1A).

CT revealed a 2.3 × 2.2 × 2.9 cm^3 predominantly cystic and partially exophytic lesion in the right kidney, with a well-defined margin. The CT density of the cystic portion was approximately 15 Hounsfield units (HU), and the solid portion had an isodensity of 44 HU without a gross fat component or calcification. In addition, the solid portion had segmental and nodular wall thickening in the posterior aspect of the lesion, with a maximum thickness of approximately 9 mm. Dynamic enhanced CT of the solid portion showed strong enhancement in the corticomedullary and nephrogenic phases and washout in the excretory phase (Fig. 1B).

Contrast-enhanced MRI was performed for further characterization. T2-weighted MRI of the cystic portion of the lesion revealed a unilocular feature with homogeneous hyperintense fluid signal intensity (Fig. 1C, left). T1- and T2-weighted MRI showed that the solid portion had iso-signal intensity. The signal intensity did not drop on opposed-phase T1-weighted images, suggesting that the lesion was a fat-invisible tumor (Fig. 1C, middle and right). The enhancement pattern of the solid portion on the dynamic enhanced T1-weighted MRI scan was similar to that on the CT scan (Fig. 1D). The solid portion of the lesion showed restricted diffusion on diffusion-weighted images (b-value of 800 s/mm^2) and apparent diffusion coefficient values (Fig. 1E). The diagnosis of the predominantly cystic lesion as Bosniak classification IV

was based on these imaging findings, and the first radiological impression was cystic RCC.

The patient underwent a radical right nephrectomy. Grossly, the largest diameter of the tumor was 2.5 cm, and no cortical invasion was suspected. Macroscopically, a well-demarcated mass was observed in the renal cortex (Fig. 1F, upper left). Microscopically, the tumor was composed of predominantly epithelioid cells with abundant eosinophilic cytoplasm (Fig. 1F, upper middle). Immunohistochemically, epithelioid tumor cells showed positive immunoreactivity for human melanoma black-45 and Melan A, but negative immunoreactivity for cyto-

Fig. 1. Imaging findings of epithelioid angiomyolipoma with epithelial cysts in a 58-year-old male.
A. Transverse ultrasonography shows a predominantly cystic lesion in the interpolar area of the right kidney. An eccentric solid portion is seen in the posterior aspect of the cystic lesion (arrows).
B. An axial unenhanced CT image shows a partially exophytic cystic lesion with an isodense solid portion (upper left). The axial corticomedullary phase (upper right), nephrogenic phase (lower left), and excretory phase (lower right) contrast-enhanced CT images show an arterial-enhancing solid portion with delayed washout (arrows).
C. Axial T2-weighted MRI (left) shows a predominantly cystic lesion with an iso-signal intensity of the solid portion. The eccentric solid portion shows no signal loss on the opposed-phase image (middle), compared with the in-phase T1-weighted image (right).

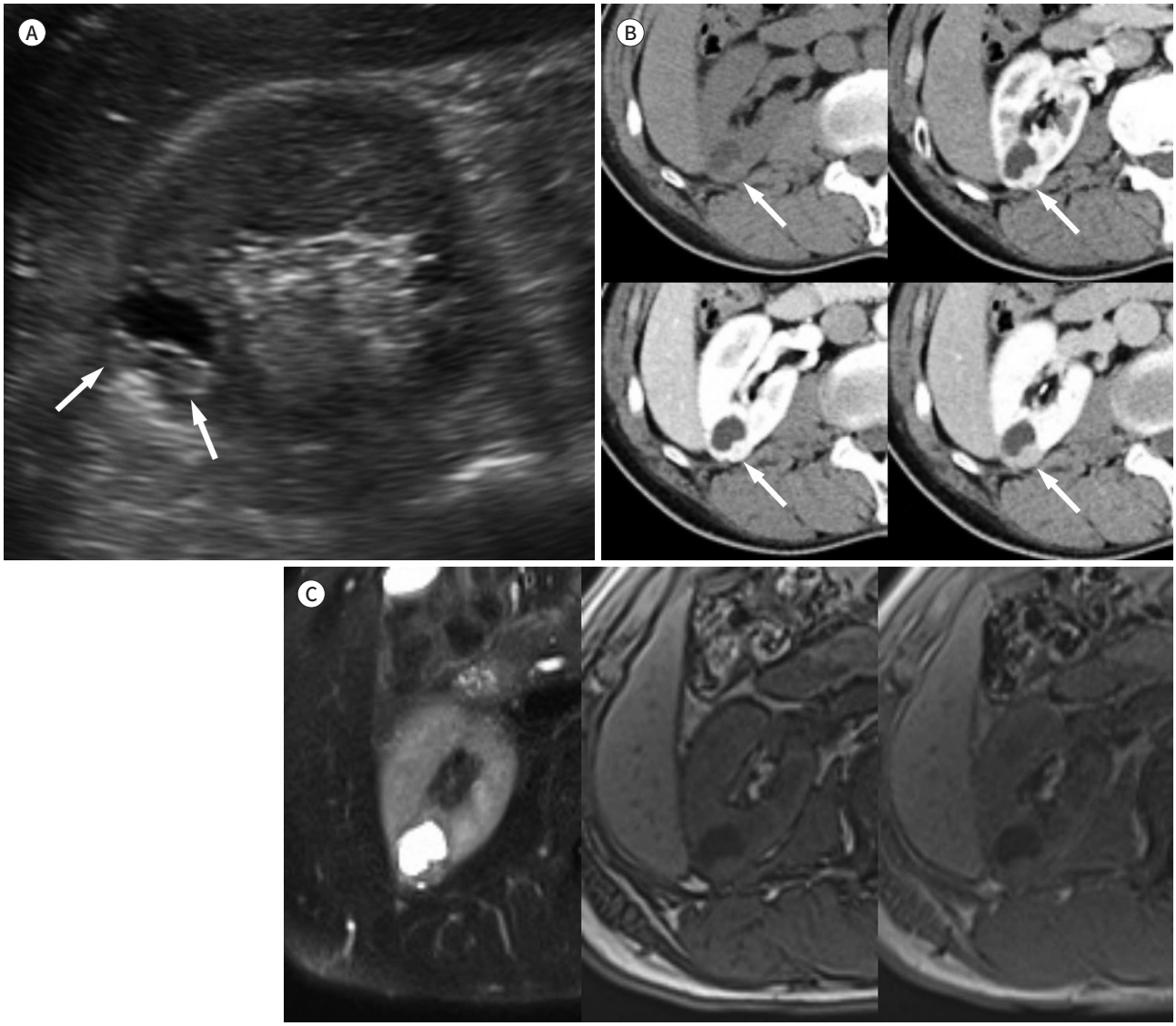

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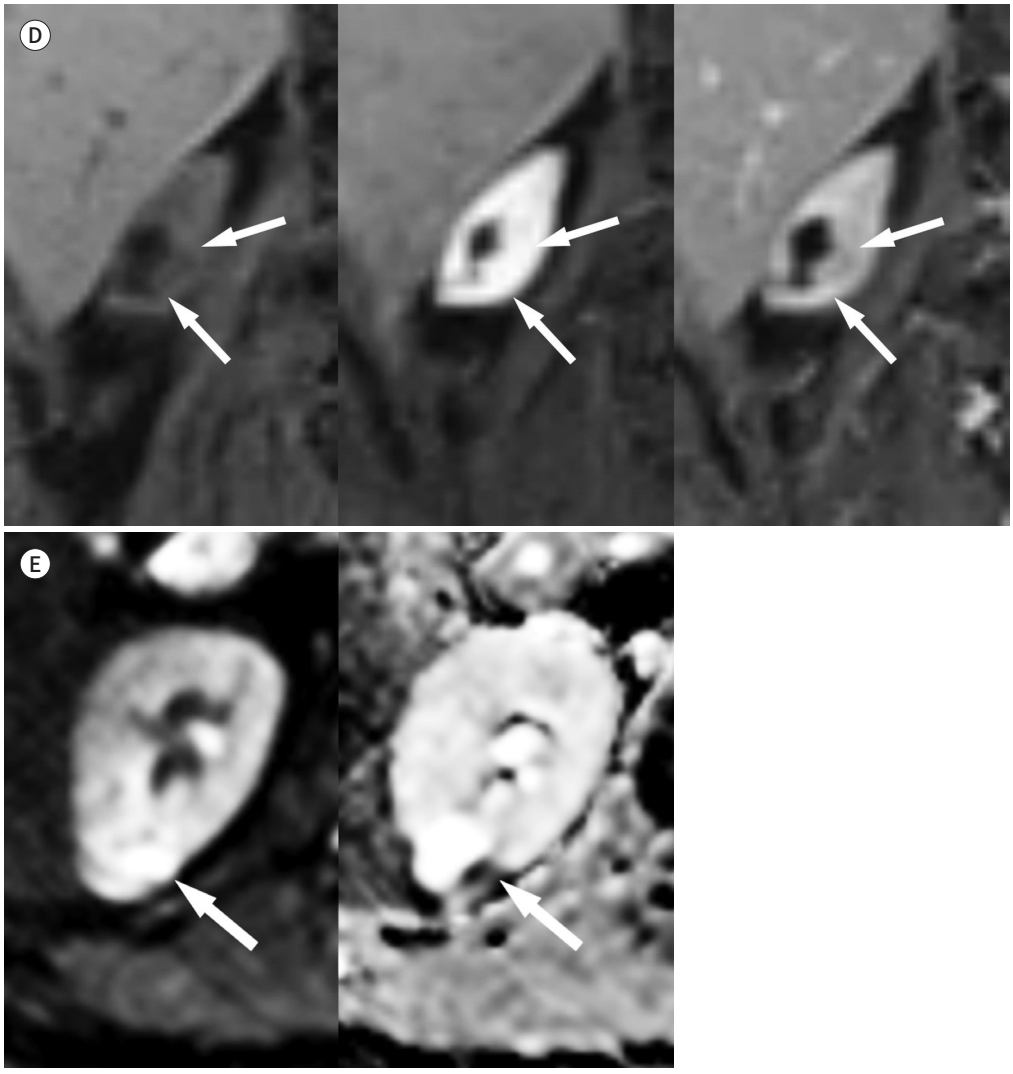
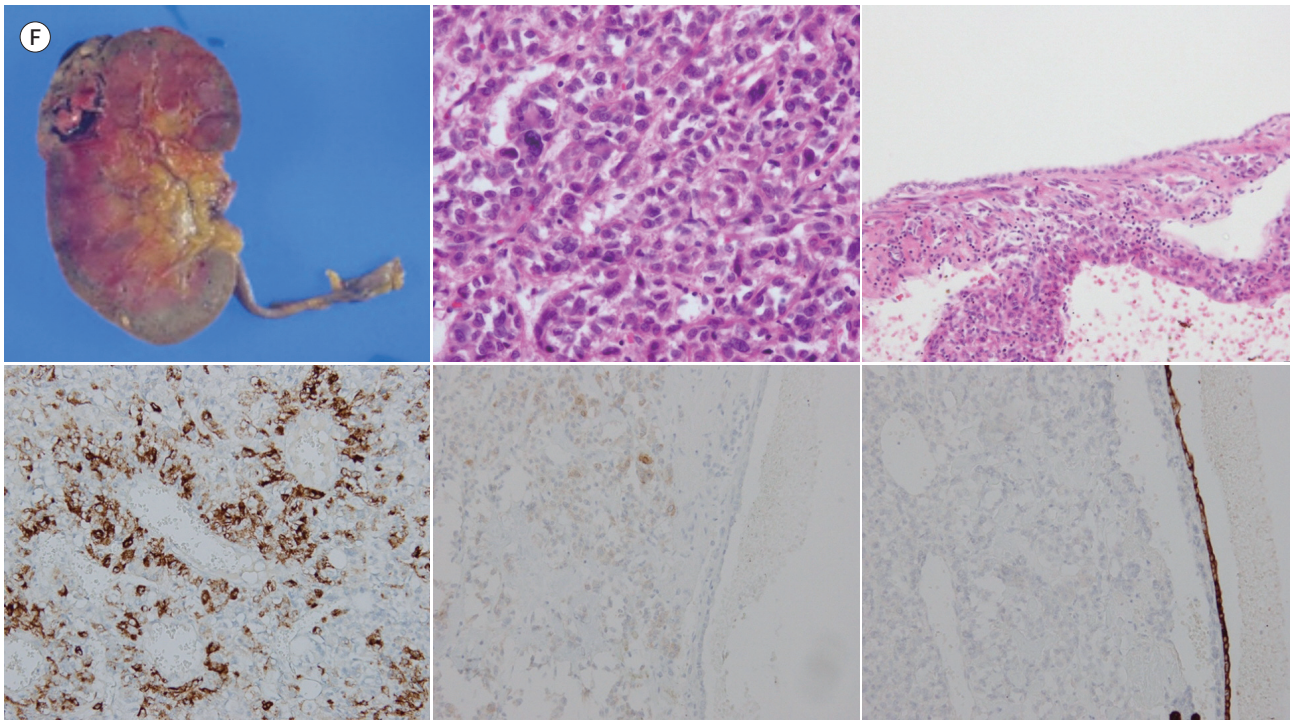
Fig. 1. Imaging findings of epithelioid angiomyolipoma with epithelial cysts in a 58-year-old male.
D. Coronal unenhanced T1-weighted image (left), corticomedullary phase image (middle), and excretory phase image (right) of the enhancement pattern of the solid portion of the lesion are similar to the CT scans (arrows).
E. The axial diffusion-weighted image obtained using a b-value of 800 s/mm^2 (left) shows the solid portion of the lesion with slightly high signal intensity (arrow). The axial apparent diffusion coefficient map (right) shows low values in the corresponding area (arrow).



keratin 7 (Fig. 1F, lower left, middle, and right, respectively). These findings are consistent with those for EAML. In addition, the tumor was accompanied by epithelial cysts lined with a single layer of cuboidal cells (Fig. 1F, upper right). In contrast to the solid portion of the tumor, the cuboidal cells of the epithelial cysts were negative for Melan A and positive for cytokeratin 7 on immunohistochemical staining (Fig. 1F, lower middle and right, respectively). From all these findings, we diagnosed the predominantly cystic tumor as EAML with epithelial cysts.

This retrospective study was approved by the Institutional Review Board of Jeju National University Hospital, and the requirement for informed consent was waived (IRB No. 2021-05-013).

Fig. 1. Imaging findings of epithelioid angiomyolipoma with epithelial cysts in a 58-year-old male.
F. The gross specimen (upper left) has a well-demarcated mass in the renal cortex. A histological examination (upper middle and right) shows predominantly epithelioid cells with abundant eosinophilic cytoplasm and epithelial cysts lined with cuboidal cells (hematoxylin and eosin stain: upper middle at × 400 and upper right at × 200). An immunohistochemical examination shows epithelioid tumor cells with positive immunoreactivity for human melanoma black-45 and Melan A but negative immunoreactivity for cytokeratin 7 (lower left, middle, and right, respectively; IHC stain, × 200).



DISCUSSION

EAML is an extremely rare subtype of AML that is composed of epithelioid tumor cells, which have abundant eosinophilic granular cytoplasm, and little or no fat. It has the potential to be malignant and have metastatic and locally aggressive behavior. Like other AMLs, EAML has two types: one related to tuberous sclerosis complex (TSC) and one not related to TSC, and it belongs to a family of perivascular epithelioid cell tumors (1, 6). Compared with renal parenchyma, unenhanced CT shows that EAML is mainly a hyperattenuating mass, which is thought to be due to the abundant muscle component. Contrast-enhanced CT of EAML reveals homogeneous enhancement and sometimes heterogeneous enhancement due to hemorrhage or necrosis. T2-weighted MRI shows that EAML has a low signal intensity, which also is thought to be due to the muscle component. Among solid renal tumors, it is difficult to differentiate EAML from papillary RCC, chromophobe RCC, and minimal-fat AML because they all have low signal intensity on T2-weighted MRI and similar imaging findings (7, 8).

A few cases of cystic AML have been reported, most of which were AMLEC. In addition, few cases of cystic changes in EAML and fat-poor AML caused by hemorrhage or necrosis have been reported (1, 4-10). AMLEC, a rare variant of AML, has epithelial cysts lined with a single layer of low cuboidal epithelium. AMLEC also contains minimal fat tissue (5). Definitive imaging findings of AMLEC have not yet been established. Imaging of one case of AM-

LEC showed cystic and solid features, and the eccentric solid portion showed hyperdensity on CT and hypointensity on T2-weighted MRI (1). In addition, there have been cases in which the multilocular cystic mass did not have a distinct enhancing septum or wall or it had a slightly thick wall (1, 10). Most of the few reported cases of cystic EAML were multilocular cystic EAML with a hyperattenuating cystic wall or septum, and characterized by massive hemorrhage or necrosis (6, 8). These cystic AMLs require differentiation from other cystic renal tumors. Multilocular cystic RCC is representative of a malignant cystic renal tumor and characterized by an enhanced solid portion, an irregularly thick wall or septum, and intratumoral hemorrhage. However, these characteristics are mostly the same as those of cystic AML (7, 10). Some studies have reported that some radiological findings which favor cystic AML in differential diagnosis for cystic renal neoplasm are that the solid portion of the cystic renal tumor is hyperdense on CT or low signal intensity on T2-weighted MRI, and the cystic portion has massive hemorrhage (7, 8). Mixed epithelial and stromal tumors are more common in women and are associated with hormone therapy (10). Multilocular cystic nephroma may be considered when the mass herniates toward the renal pelvis (10).

A combination of two rare AML variants, i.e., EAML and AMLEC, was first described by Filho Jdo et al. (2) in 2012, but they focused on the immunohistochemical findings of the tumor and did not perform imaging. Therefore, our case is the first report on the imaging of the combination of the two rare variants of AML. In this case, CT of the solid portion of the tumor showed isodensity and T2-weighted MRI showed iso-signal intensity; however, no hemorrhage or multilocular features were seen in the cystic portion of the tumor. These imaging findings were different from the previously reported imaging findings of EAML or cystic AML, which made the differential diagnosis more difficult.

In conclusion, EAML with epithelial cysts is a very rare variant of AML. However, knowledge about this disease will help differentiation among predominantly cystic renal tumors.

Author Contributions

Conceptualization, L.S.H., L.J.S.; data curation, L.S.H., L.J.S., H.C.L.; investigation, L.S.H., K.J.J., K.S.Y., L.K.R.; methodology, L.S.H., L.J.S.; project administration, L.J.S., H.I.K.; supervision, L.J.S., H.I.K.; visualization, L.S.H., K.J.J., K.S.Y., L.K.R., H.I.K., H.C.L.; writing—original draft, L.S.H., L.J.S.; and writing—review & editing, K.J.J., K.S.Y., L.K.R., H.I.K.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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상피낭종을 동반한 신장의 상피모양 혈관근지방종: 두 희귀 질환의 조합에 대한 증례 보고

이상훈[1] · 이정섭[1*] · 김정재[1] · 고수연[1] · 이경렬[1] · 황임경[1] · 현창림[2]

신장의 혈관근지방종은 전형적으로 고형 종괴지만, 낭성 형태의 신장 혈관근지방종이 소수 보고되었다. 그중에서 대다수의 보고는 상피낭종을 동반한 혈관근지방종으로 입방상피조직으로 둘러싸인 낭종을 포함한다. 다음으로 상피모양 혈관근지방종은 호산성 세포질이 풍부한 상피모양세포를 포함하는데, 이것이 출혈이나 괴사로 인해 낭성변화를 일으킨 증례 보고도 있었다. 이런 낭성 형태의 신장 혈관근지방종은 영상검사에서 낭성 신세포암 등의 다른 낭성 종괴로 오인될 수 있다. 이 증례 보고에서는 58세 남성에서 발견된 상피낭종을 동반한 상피모양 혈관근지방종의 영상 소견을 살펴보려고 한다.

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