A Case of Renal Angiomyolipoma with Minimal Fat Mimicking Renal Cell Carcinoma

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SUMMARY
We present a case of renal angiomyolipoma (AML) with minimal fat mimicking renal cell carcinoma (RCC). AML is composed of variable amount of fat, smooth muscle and abnormal blood vessels. In general, AML can be differentiated from RCC with great accuracy using modern radiological techniques due to the fat component of the renal mass. In the present case, the renal tumor did not demonstrate intratumoral fat on radiological studies. Surgery was performed and the renal tumor was removed. Histologically, the renal tumor showed abundant muscle that occupied almost the entire lesion, which demonstrated HMB-45 antigen. The tumor was diagnosed as renal AML.

Key Words: Renal angimyolipoma, Renal cell carcinoma

INTRODUCTION
Angiomyolipoma (AML) is the most common benign tumor of the kidney. In evaluating of AML, the more common problem is differentiating it from renal cell carcinoma (RCC) because the recommended approaches to management for these two diseases are significantly different. In general, AML can be accurately diagnosed by identifying intratumoral fat on radiological studies. However, there is a subset of AML that does not demonstrate intratumoral fat on radiological studies, namely, so-called AML with minimal fat, mimicking RCC. Herein, we report a case of AML with minimal fat.

CASE REPORT
A 40-year-old woman was referred to our hospital for evaluation of microscopic hematuria. She did not have tuberous sclerosis. In the right kidney, ultrasonography (US) demonstrated an iso-echoic mass relative to the renal parenchyma (Fig. 1). Unenhanced CT showed a homogeneous hyperdense renal mass compared with the renal parenchyma (Fig. 2a). The

Figure 1 Ultrasonography demonstrated a right renal mass that was iso-echoic relative to the renal parenchyma.
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due to the fat component of the renal mass. However, there are some problems in differentiating between AML and RCC. First, there is a subset of RCC with intratumoral fat. RCC showing fat density on CT can be caused by the engulfment of peripheral or renal sinus fat into the tumor, intratumoral bone metaplasia with fatty marrow elements, or the presence of cholesterol necrosis being misconstrued as fat. Second, there is a subset of AML with minimal fat. Jinzaki et al. first reported AML with minimal fat in 1997 and indicated that this unusual manifestation account for 4.5% of all AMLs\(^1\). Because AML with minimal fat histologically shows abnormally thickened blood vessels and predominance of smooth muscle with only a small amount of fat, differentiating between AML with minimal fat from RCC on radiological findings is very difficult. Although it has been reported that AML with minimal fat possesses at least 3% adipose tissue on histologic analyses (mean 4.1%, range 3%~10%)\(^2\), AML with minimal fat does not demonstrate intratumoral fat on

hyperdense mass demonstrated homogeneous enhancement (Fig. 2b) and an early washout enhancement pattern after administration of contrast material (Fig. 3). T1-weighted magnetic resonance image (MRI) demonstrated a right renal mass that was slightly less intense than the renal parenchyma (Fig. 4a). On T2-weighted image, the lesion was hypointense (Fig. 4b). We considered the right renal mass to be malignant. A staging workup demonstrated no other evidence of disease. Surgery was performed and the renal tumor was removed. Histologically, the renal tumor showed abundant muscle that occupied almost the entire lesion (Fig. 5a), which demonstrated HMB-45 antigen (Fig. 5b). The tumor was diagnosed as renal AML. The patient has remained free of disease for 105 months since surgery.

**DISCUSSION**

Generally, AML can be differentiated from RCC with great accuracy using modern radiological techniques due to the fat component of the renal mass. However, there are some problems in differentiating between AML and RCC. First, there is a subset of RCC with intratumoral fat. RCC showing fat density on CT can be caused by the engulfment of peripheral or renal sinus fat into the tumor, intratumoral bone metaplasia with fatty marrow elements, or the presence of cholesterol necrosis being misconstrued as fat. Second, there is a subset of AML with minimal fat. Jinzaki et al. first reported AML with minimal fat in 1997 and indicated that this unusual manifestation account for 4.5% of all AMLs\(^1\). Because AML with minimal fat histologically shows abnormally thickened blood vessels and predominance of smooth muscle with only a small amount of fat, differentiating between AML with minimal fat from RCC on radiological findings is very difficult. Although it has been reported that AML with minimal fat possesses at least 3% adipose tissue on histologic analyses (mean 4.1%, range 3%~10%)\(^2\), AML with minimal fat does not demonstrate intratumoral fat on

![Figure 2](image2.jpg) (a) Unenhanced CT showed a homogeneous hyperdense renal mass compared with that of the renal parenchyma. (b) The hyperdense mass showed a pattern of homogeneous enhancement.

![Figure 3](image3.jpg) Renal mass showed an early washout enhancement after administration of contrast material. (a) early arterial phase (b) late arterial phased (c) static phase
A Case of Renal Angiomyolipoma with Minimal Fat Cell RCC was a valuable CT finding. To our knowledge, the present case is the second report to describe the enhancement pattern after the administration of contrast material. Data from more patients should be accumulated in order to establish a definitive conclusion. In our case, the renal tumor was isoechoic ultrasonographically. The coexistence of fat and muscle would result in hyperechogenicity due to the difference between the acoustic impedances of each component. Therefore, a homogeneous muscle component with a small amount of fat would account for the iso-echoic pattern on US. MRI was unable to detect the fat component within this tumor and the tumor showed low intensity on T2-weighted images.

Simpfendorfer et al. investigated whether counts of pixels on CT scans could aid in the diagnosis of AML radiological finding.

In the present case, attenuation of the tumor on unenhanced CT images was high compared with that of the renal parenchyma and similar to that of muscle. This finding suggests that the lesion contained a rich muscle component. Previous series have also reported similar findings. Kim et al. reported homogeneous hyperdense findings of AML with minimal fat on unenhanced CT in 53% of their cases. Jinzaki et al. pointed out that the AMLs reported by Kim et al. might include cases of AML with diffusely scattered fat. After the administration of contrast material, AML with minimal fat in the present case showed an early washout enhancement pattern similar to that of clear cell RCC (CCRCC), although Kim et al. reported that a prolonged enhancement pattern similar to that of papillary cell RCC was a valuable CT finding. To our knowledge, the present case is the second report to describe the enhancement pattern after the administration of contrast material. Data from more patients should be accumulated in order to establish a definitive conclusion. In our case, the renal tumor was isoechogenic ultrasonographically. The coexistence of fat and muscle would result in hyperechogenicity due to the difference between the acoustic impedances of each component. Therefore, a homogeneous muscle component with a small amount of fat would account for the iso-echoic pattern on US. MRI was unable to detect the fat component within this tumor and the tumor showed low intensity on T2-weighted images.

Simpfendorfer et al. investigated whether counts of pixels on CT scans could aid in the diagnosis of AML.

Figure 4  (a) On MRI examination, T1-weighted axial image view demonstrated a right renal mass that was slightly hypo-intense relative to the renal parenchyma. (b) T2-weighted axial image view demonstrated a hypo-intense mass in the right kidney.

Figure 5  Histologically, the renal tumor showed abundant muscle that occupied almost the entire lesion (Fig. 5a), which demonstrated HMB-45 antigen (arrow) (Fig. 5b).
with minimal fat\textsuperscript{7}. They concluded that AML with minimal fat could not be reliably identified based on an absolute pixel count. The most striking feature of AML with minimal fat is its high attenuation similar to that of muscle on unenhanced CT. However, high attenuation is nonspecific, as 22\% of RCCs also show this feature on unenhanced CT\textsuperscript{5}. Catalano et al. investigated pixel-by-pixel histogram analysis of unenhanced CT images of histologically proven AMLs with minimal fat and CCRCCs\textsuperscript{6}. They reported that once lesions with macroscopic fat have been excluded, pixel attenuation histogram analysis cannot be used to distinguish AML with minimal fat from CCRCC. Kim et al. reported that double-echo gradient-echo chemical shift MRI can be used to differentiate AML with minimal fat from other renal neoplasms\textsuperscript{8}. However, distinguishing AML with minimal fat from leiomyoma may be very difficult, because both are composed of rich muscle. Recently, some leiomyomas have been shown to focally express HMB-45, suggesting a relationship to AML\textsuperscript{10}.

Almost all AMLs with minimal fat are surgically resected because malignancy is diagnosed on preoperative evaluation\textsuperscript{1-6}. However, in renal lesions showing homogeneously high attenuation on unenhanced CT images, homogeneous enhancement on contrast-enhanced CT images, homogeneous isoechogenicity on US and hypointensity on T2-weighted MRI diagnosis and decisions regarding management should be considered AML with minimal fat. In such cases, biopsy is recommended.

REFERENCES