

Fat Poor Renal Angiomyolipoma Showing Collapsed Shape: A Case Report

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Abstract

Fat-poor renal angiomyolipoma (fpAML) and renal cell carcinoma (RCC) are difficult to differentiate and misdiagnosis can lead to unnecessary nephrectomy. We experienced a case showing a “collapsed shape” which reflected a fpAML tissue type. A renal tumor was incidentally discovered in a 42-year-old female during an abdominal ultrasound. RCC was suspected according to CT and MRI imaging results, and a partial nephrectomy was performed. However, the pathologic diagnosis was fpAML. Upon reevaluation of preoperative images, morphological change to the tumor due to contact with surrounding tissues: the collapsed shape was observed and could be identified by CT, which is the gold standard test for differentiating renal tumors. In cases where the collapsed shape is observed in a renal tumor, fpAML should be considered.

Keywords

Fat Poor Renal Angiomyolipoma, Renal Cell Carcinoma, Differential Diagnosis, Computed Tomography

1. Introduction

With the wide availability of imaging examinations, the chance of incidentally finding small renal tumors has increased [1]. Among these tumors, most solid tumors are renal cell carcinomas (RCC), but 20% - 30% are benign, including angiomyolipomas (AMLs) and oncocytomas [2]. Particularly among these tumors, fat poor renal angiomyolipoma (fpAML) and RCC are difficult to differentiate, and misdiagnosis can lead to an unnecessary nephrectomy. Previous reports have described findings such as hyperattenuating on unenhanced CT as

well as hyperenhancement during the corticomedullary phase and washout on dynamic contrast-enhanced CT are characteristics of fpAML. However, these findings overlap with renal cell carcinoma, and no single CT feature can reliably diagnose fpAML [3]. We report a tumor that showed a “collapsed shape” which reflected a fpAML tissue type. This collapsed shape may help make the diagnosis of fpAML.

2. Case

A 42-year-old female presented to our hospital after a tumor was incidentally discovered in the patient’s left kidney by abdominal ultrasonography during a health checkup. The patient had an unremarkable family history and medical history. A dynamic contrast-enhanced CT scan revealed a 20 mm mass protruding laterally from the inferior half of the left kidney in the unenhanced phase (Figure 1(a)). The tumor showed a moderate and uniform contrast enhancement during the corticomedullary phase, which decreased slightly in later phases (Figures 1(b)-(d)). On MRI, a low signal on T2-weighted imaging was observed (Figure 2(a)) and traces of fat were not detected by chemical shift imaging (Figure 2(b) and Figure 2(c)). Pseudocapsule-like structures were not observed. Both fpAML and renal cell carcinoma were included in the initial differential diagnosis. Since the possibility of renal cell carcinoma could not be ruled out based on imaging alone, a partial nephrectomy was performed.



Figure 1. Dynamic contrast-enhanced CT scan. Noncontrast CT showed a high attenuating 20 mm mass protruding laterally from the inferior half of the left kidney (a) (arrow). No fat components were detected. Dynamic contrast-enhanced CT showed a moderate and uniform contrast enhancement from the corticomedullary phase (b) (arrow), which decreased slightly in the nephrographic phase (c) (arrow) and pyelographic phase (d).

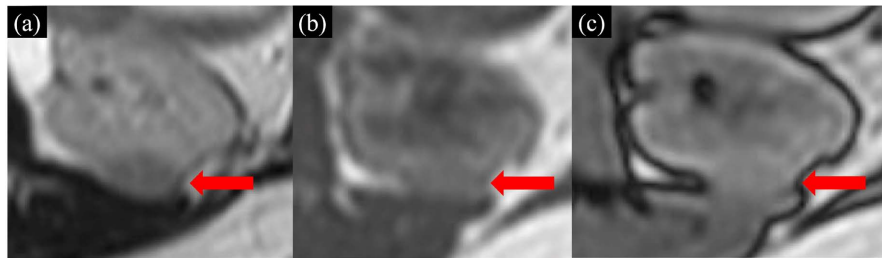


Figure 2. MRI. (a) T2WI, (b) and (c) Chemical shift imaging ((b) In phase, (c) Out of phase). MRI showed low signal on T2-weighted imaging (a) (arrow) and traces of fat was not detected by chemical shift imaging (b and c) (arrows).

The excised specimen is shown in **Figure 3**. A well-defined mass 20 mm in size protruding outside the kidney was grossly observed. Histologically, the tumor consisted of abundant smooth muscle and blood vessels. Fat cells were hardly observed (**Figure 4(a)** and **Figure 4(b)**). Immunohistologically, the tumor was positive for HMB-45 (**Figure 4(c)**) and α SMA (**Figure 4(d)**). The final pathological diagnosis was fpAML. Upon detailed reevaluation of preoperative images, morphological change to the tumor due to contact with surrounding tissue: the collapsed shape was observed.

The patient's postoperative course was good with no complications. The patient was followed up for 7 years after the operation and developed no recurrence.

3. Discussion

The following two points could be shown in this case. In fpAML, the collapsed shape can be shown to reflect the tissue type, and collapsed shape can be indicated by CT.

Firstly, in fpAML, the collapsed shape may reflect AML tissue type. Typical AML is a benign tumor composed of fat, smooth muscle, and vascular components [4]. For typical AML, diagnosis by detecting macroscopic fat within the tumor on non-contrast CT is possible. However, in fpAML, differentiation from renal cell carcinoma is difficult due to the lack of macroscopic fat. The ratio of fpAML with a composition mainly of smooth muscle is high, and this is reflected by their higher attenuation on non-contrast-enhanced CT compared to the renal parenchyma, as well as a uniform contrast enhancement, and a low signal on T2-weighted imaging in MRI [5] [6]. However, differential diagnosis is difficult due to diverse findings depending on the proportion of tissues involved and many findings overlap with renal cell carcinoma [7]. In this case, although fpAML was included in the initial differential diagnosis, partial nephrectomy was performed because renal cell carcinoma could not be ruled out. Since the pathologic diagnosis was fpAML, a detailed reevaluation of the preoperative images was performed, and collapsed shape was shown to reflect AML tissue type. The collapsed shape is a morphological change in which the tumor collapses due to contact and compression from the surrounding tissue. In the sagittal section, the

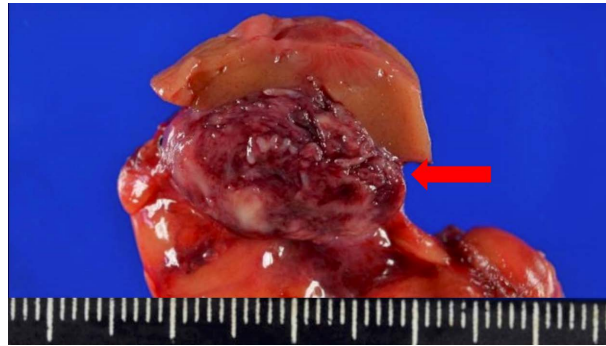


Figure 3. Macroscopic image of the excised specimen. The surgically resected specimen showed a well-defined mass 20 mm in size protruding from the kidney (arrow).

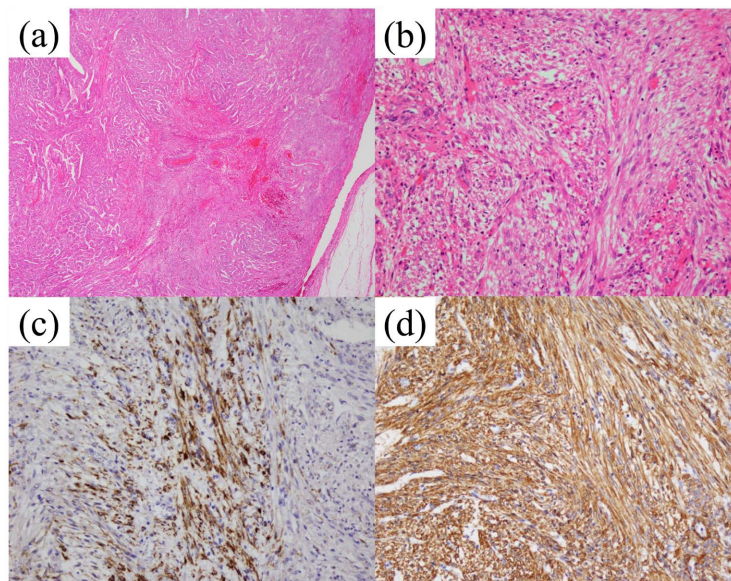


Figure 4. Histologic and immunohistochemical findings. Hematoxylin and eosin stain (×20) (a) and (×200) (b) showed a tumor composed of smooth muscle, and thick-walled blood vessels, with minimal fat. Immunohistochemistry demonstrated positivity for HMB-45 (×200) (c) and α SMA (×200) (d).

tumor was confirmed to be in wide contact with the dorsal muscle which could be considered to cause the deformation (**Figure 5**). This is assumed to reflect the tissue elasticity and stiffness of smooth muscle comprising fpAML. While still in the research phase, measuring tissue elasticity and stiffness by ultrasound elastography has been reported to be useful to differentiate between AML and renal cell carcinoma [8] [9]. This supports the mechanism that fpAML shows collapsed shape. To our knowledge, this is the first report which focuses on tissue elasticity and stiffness of fpAML and points out the morphological changes caused by contact with the surrounding tissue.

Secondly, the collapsed shape could be pointed out on CT images. CT is widely accepted as first choice imaging for preoperative diagnosis of renal tumors [10], and an MRI scan is added when a definite diagnosis is difficult. Additional information obtained by MRI includes trace amounts of fat, pseudo-capsules,



Figure 5. Collapsed shape. Sagittal reformatted contrast-enhanced CT showed a morphological change to the tumor presumably due to contact with the dorsal muscle (arrow).

muscle tissue, fibrous components, and hemosiderin. However, these findings overlap with other renal tumors and are not highly specific. Therefore, MRI does not provide definitive diagnostic findings in many cases. Since the collapsed shape is a morphologic change, it could be detected by CT, which is the gold standard test for differentiating renal tumors. In this respect, the collapsed shape could be a useful finding suggestive of AML.

In this case, the focus was placed on the collapsed shape which reflected a fpAML tissue type, but the usability of an angular interface as a morphological characteristic of fpAML has previously been reported [11] [12]. The angular interface is described as a tumor having a tapered pyramidal outline or clear apex with the renal parenchyma. However, differentiation between collapsed shape and the angular interface was made according to the following two points. Firstly, findings for collapsed shape reflected tissue elasticity and hardness. On the other hand, for the angular interface, differences in growth patterns between benign and malignant masses were assumed to affect the shape of the boundary with the renal parenchyma. However, no convincing pathophysiological explanation was provided [12]. Secondly, collapsed shape is an interaction between the tumor and the surrounding tissue, whereas an angular interface is caused by an interaction between the tumor and the kidney.

In conclusion, for fpAML, the collapsed shape was shown to reflect the tissue type, and collapsed shape could be pointed out by CT, which is the gold standard test for differentiating renal tumors. When the collapsed shape is observed in a renal tumor, fpAML should be considered and the recommendation of a follow-up or biopsy may lead to a reduction in unnecessary nephrectomies.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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