An Incidental Diagnosis of Microscopic Renal Angiomyolipoma Completely Excised on Renal Biopsy: A Case Report

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Patient: Female, 44-year-old
Final Diagnosis: Angiomyolipoma
Symptoms: Asymptomatic proteinuria
Medication: —
Clinical Procedure: —
Specialty: Pathology

Objective: Rare disease
Background: Microscopic tumor foci have been detected incidentally on renal biopsy, including renal cell carcinoma and renomedullary interstitial cell tumor (medullary fibroma). A report is presented of a case of an incidental finding of microscopic renal angiomyolipoma that was diagnosed and completely excised on core needle biopsy.

Case Report: A 44-year-old woman was referred to our hospital for evaluation of persistent mild proteinuria. Three years previously, she was diagnosed with Cushing’s syndrome associated with a right adrenal cortical adenoma, which was successfully treated with unilateral adrenalectomy. During the present admission, a renal biopsy was performed that showed minimal changes in the renal glomeruli and interstitium. Immunofluorescence showed weakly positive staining for IgM in the glomeruli and no dense deposits. A microscopic focus of a predominantly spindle-cell tumor was found in the corticomedullary region. Immunohistochemistry showed positive immunostaining for HMB-45, Melan-A, and alpha-smooth muscle actin (ASMA), which supported a diagnosis of angiomyolipoma. Abdominal ultrasound at one-year follow-up showed no evidence of residual renal tumor.

Conclusions: To our knowledge, this is the first reported case of a completely excised incidental microscopic renal angiomyolipoma. This case demonstrated that even when imaging findings are normal, renal biopsy may detect microscopic foci of primary renal tumors.

MeSH Keywords: Angiomyolipoma • Kidney Neoplasms • MART-1 Antigen

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Background

Angiomyolipoma is a benign mesenchymal tumor that is currently classified as a perivascular epithelioid cell tumor, or PEComa [1]. Histologically, angiomyolipoma is a triphasic tumor composed of smooth muscle cells, mature adipocytes, and thick-walled and dilated blood vessels, in varying proportions. However, in some tumors, one component may exclusively predominate, and such tumors are termed atypical angiomyolipoma. Immunohistochemistry shows that the myogenic cells in angiomyolipoma express smooth muscle markers, including alpha-smooth muscle actin (ASMA) and desmin, and melanocytic markers including HMB-45 and Melan-A. Most cases of angiomyolipoma involve the kidney and occasionally involve the liver, and the involvement of other organs and anatomical sites is extremely rare [1].

Angiomyolipoma typically forms a tumor mass that can be identified on renal imaging, and may cause clinical effects. A tumor size of more than 4 cm has been the main indication for surgical intervention [2–4]. However, angiomyolipoma is often detected incidentally on imaging due to the increasing use of cross-sectional imaging and advances in imaging techniques [4]. A recent report described an asymptomatic small renal angiomyolipoma that was incidentally detected on abdominal magnetic resonance imaging (MRI) and was confirmed by core needle biopsy, which highlighted the importance of histology to confirm the diagnosis [5]. Also, a series of autopsy cases identified asymptomatic and small angiomyolipoma ranging in size from 0.1 mm to 10 mm in a minority of patients [6].

A report is presented of a case of an incidental finding of microscopic renal angiomyolipoma detected on core needle biopsy. In this case, the renal angiomyolipoma was completely excised on diagnostic biopsy.

Case Report

A 44-year-old woman was referred to our institution with chronic proteinuria. Three years before her present admission, she was diagnosed with Cushing's syndrome associated with a right adrenal cortical adenoma. Unilateral adrenalectomy was successfully performed. The clinical manifestations of Cushing's syndrome, including central obesity, moon face, and hypertension, improved after surgery. However, asymptomatic proteinuria persisted for a further two years. On admission at our institution, no pretibial edema was observed. Her blood pressure was 108/58 mmHg, and her urine protein excretion, according to the protein/creatinine ratio, was 1.07 g/gCr. She had no microscopic hematuria, serum creatinine was 0.64 mg/dL, and blood urea nitrogen (BUN) was 8.9 mg/dL.

Renal biopsy was performed to identify the cause of her persistent proteinuria. Histology of the renal core needle biopsy identified 22 glomeruli, three of which were globally sclerotic. In the remaining glomeruli, there was no glomerular hypertrophy or proliferative change. Atrophic renal tubules were observed in 5% of the renal cortex, but there was no interstitial fibrosis or inflammation. Immunofluorescence showed weak IgM positivity in the mesangium, and electron microscopy did not identify dense deposits. In the corticomedullary region, there was a small focus of spindle cells with eosinophilic cytoplasm and enlarged nuclei without mitoses or necrosis (Figure 1A, 1B). The microscopic tumor focus measured 1,800 μm in maximum diameter. Adipocytes were sparse, and thick-walled vessels were not observed within the lesion. Immunohistochemistry showed that the tumor spindle cells were diffusely positive for Melan-A and ASMA (Figure 1C, 1D). Some scattered cells were immunopositive for HMB-45, and no estrogen receptor immunoreactivity was detected. Based on the histological findings, the patient was diagnosed with a microscopic focus of angiomyolipoma. Abdominal ultrasound at one-year follow-up showed no evidence of residual renal tumor.

Discussion

Renal biopsy specimens rarely contain small neoplasms that are too small to be detected by imaging methods. In a retrospective study that examined 11,800 renal biopsies, neoplasms were detected in only 25 biopsies (0.2%) biopsies, which included papillary neoplasms in 22 cases, clear cell renal cell carcinoma in two cases, and collecting duct carcinoma in one case [7]. Also, a previous study examined the incidence of incidental renal neoplasms of autopsy and found that papillary adenoma was detected most frequently (18.7%), followed by adrenal rest tumor (7.7%), oncocytoma (1.2%), and renal cell carcinoma (0.5%) [8]. Renomedullary interstitial cell tumor (medullary fibroma) has also been reported as an incidental finding in renal biopsy specimens obtained from a patient with asymptomatic proteinuria and hematuria [9]. However, to the best of our knowledge, the present case is the first report of a completely excised incidental microscopic renal angiomyolipoma on diagnostic renal needle core biopsy.

In the present case, abdominal computed tomography (CT) before adrenalectomy did not detect any neoplastic lesion in the kidney. The reasons for this were that the tumor measured only 1,800 μm in maximum diameter, which was smaller than the recommended slice thickness of CT scans (1.5–3 mm) [10]. Also, this tumor had a lack of lipomatous cells, and the lipid component of angiomyolipoma is homogenously isoechoic on ultrasound examination and manifests as a high-density area on enhanced CT [11,12]. The angiomyolipoma in this patient was a fat-poor angiomyolipoma, which is also known as...
a hyper-attenuating subtype [11]. Immunohistochemical staining showed diffuse ASMA immunopositivity, which is consistent with a diagnosis of predominant smooth muscle cell proliferation in fat-poor angiomyolipoma [13].

In the present study, the cause of proteinuria remained unclear. Hematological malignancies and solid tumors are occasionally associated with proteinuria, reflecting renal manifestations of paraneoplastic syndrome [14]. However, considering the small size of the tumor, a causal association between the tumor and proteinuria seems unlikely.

Sex steroids may play a role in the pathogenesis of angiomyolipoma. Hormone receptor expression, including estrogen and progesterone receptor expression, have been detected in resected angiomyolipoma [15]. In 2004, Yu et al. showed that a cell line obtained from a human angiomyolipoma showed estradiol-dependent cell growth in vitro [16]. Also, pregnancy and oral contraceptive therapy are associated with the growth of angiomyolipoma [17,18]. In the present case, tumor cells were negative for the estrogen receptor, which may have contributed to the small size of the tumor.

Conclusions

This report presented a case of the incidental finding of a microscopic renal angiomyolipoma detected on core needle biopsy. In this case, the renal angiomyolipoma was completely excised on diagnostic biopsy. To the best of our knowledge,
this is the first reported case of completely excised incidental microscopic renal angiomyolipoma. The findings highlight that renal biopsy may identify microscopic tumors that may not be detected on imaging.

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Conflict of interest

None

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