Renal Lymphangiomatosis Mimicking Polycystic Kidney Disease In An Adult, A Case Report and Literature Review

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Renal lymphangiomatosis is an unusual disorder. It may develop due to the abnormality of the intrarenal, peripelvic and perirenal lymphatics. The differential diagnosis contains renal lymphoma, polycystic kidney disease, multicystic dysplasia and renal tumors. We report a case of renal lymphangiomatosis, previously diagnosed as autosomal dominant polycystic kidney disease, to emphasize that these two diseases can be easily confused. It should be kept in mind that RL is in the differential diagnosis of polycystic renal disease to prevent overtreatment.

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Case Report

INTRODUCTION

Renal lymphangiomatosis (RL) is a rare disorder whose etiopathogenesis is not clear.[1] It may develop due to malformation of the perirenal lymphatic drainage. Here we present a case of a 30-year-old male patient who was diagnosed with autosomal dominant polycystic kidney disease (ADPKD) depending on renal ultrasound findings 10 years ago. Many years later, he was diagnosed with RL with magnetic resonance imaging (MRI) and overtreatment was prevented.

CASE REPORT

A 30-year-old man was diagnosed with hypertension during a routine examination10 years ago. His blood pressure controlled with perindopril/indapamide combination. There were no signs of impaired renal function and family history of ADPKD. Renal ultrasound showed the enlargement of the right kidney with multiple anechoic cysts. The left kidney was normal and had no cysts. He was diagnosed with ADPKD and followed up with annual ultrasound imaging in another hospital. When he admitted to our hospital, MRI performed to calculate kidney volume for tolvaptan treatment. MRI revealed unilateral multiple cysts and enlargement of the right kidney. The majority of the cysts were located peripherally consistent with RL (Figure1 and 2). Therefore an invasive method was not used to confirm the diagnosis and the patient followed up conservatively.

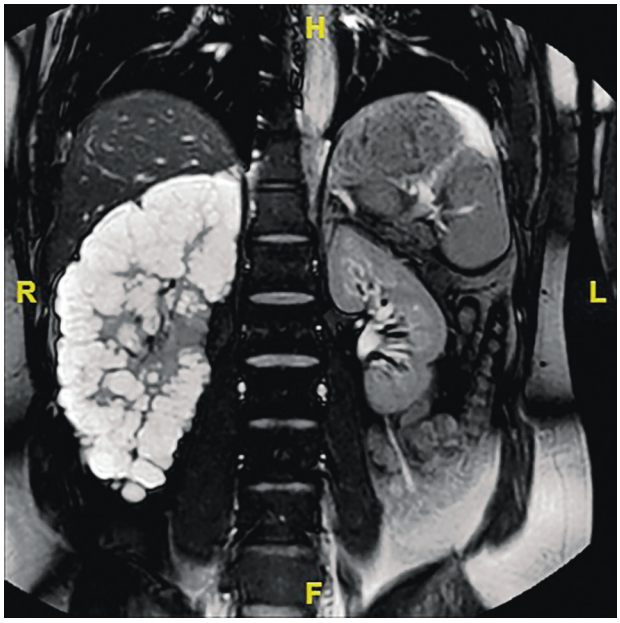

Figure 1. Axial T2-weighted MR images show unilateral multiple cysts in the right kidney. Note that majority of the cysts are located peripherally consistent with lymphangiomatosis.

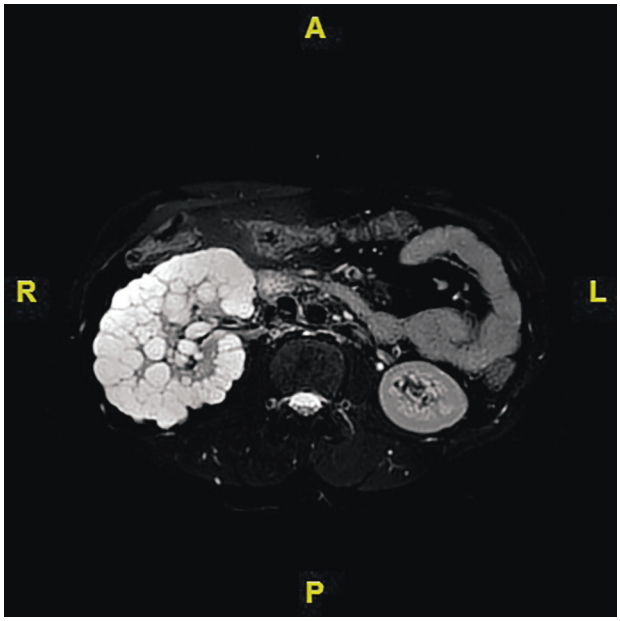

Figure 2. Coronal T2-weighted MR images show unilateral multiple cysts in the right kidney. Note that majority of the cysts are located peripherally consistent with lymphangiomatosis.

DISCUSSION

RL is a benign tumor of the kidney, and this tumor is also known as renal lymphangioma or renal lymphangiectasia.[1] RL usually presents sporadically but familial cases are also described in the literature.[2,3] There is no differece between the genders and ages. Abdominal distention and the mass in the examination are common symptoms in childhood. In adults, it is generally asymptomatic and needs no treatment, but if it is symptomatic, patients present with flank pain, hematuria, hypertension or obstructive uropathy depends on compression.[4] Nephrectomy may be required in case of obstructive uropathy and uncontrolled secondary hypertension.[5] There are few cases reported with exacerbation during pregnancy. Perirenal lymphatic collection may cause acid and impaired kidney function during pregnancy and may need treatment with percutaneous drainage and sclerotherapy.[2,6] Lymphangiomatosis is arranged in classes as cavernous, capillary and cystic according to the dimension of the lymphatic space microscopically.[7] Most cases in the literature have bilateral lymphangiomatosis, but RL can be unilateral as in our case.[7]

The differential diagnosis contains renal lymphoma, polycystic kidney disease, multicystic dysplasia, and renal tumors. RL can be limited only in the hilar region or expanded to the renal parenchyma. It is generally present in the parapelvic and renal hilar region. If it has renal parenchymal involvement, it may be confused with polycystic kidney diseases.[8] Although ultrasound can report the bilateral or unilateral multiple cysts as ADPKD, there are differences between ADPKD and RL. On ultrasound images, the cysts are parenchymal in ADPKD, but cysts are parapelvic with normal renal parenchyma in lymphangiomatosis.[9] Another distinguishing feature is that the cystic structures are covered with endothelium in lymphangiomatosis, and the cyst content is a chylous fluid with high albumin and lipid content.[1] Computed tomography (CT) and MRI have a significant role in diagnosis. CT imaging reveals cysts leading to enlargement in the hilar region and perirenal multiple capsular cysts. On MRI, multiple lesions in perirenal and peripelvic area appear hyperintense on T2 weighted images and hypointense on T1 weighted images typically.[1,10] Our case was also diagnosed with multiple peripheral cysts in the right kidney from MRI.

To our knowledge, one pediatric and two adult cases have been reported with renal lymphangiomatosis which were followed up with polycystic kidney disease previously. Both adult patients were female in their forties and diagnosed with ADPKD by ultrasound incidentally. CT and MR images revealed multiple cysts with normal parenchyma and they were diagnosed with RL as in our case.[9,11,12]

CONCLUSION

In conclusion, we present a case with an uncommon disease that mostly requires no treatment, but this condition can be confused with polycystic kidney disease which can cause end-stage renal disease. It should be kept in mind that RL is in the differential diagnosis of polycystic renal disease to prevent overtreatment.

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