Case report: renal lymphangiectasia

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Renal lymphangiectasia is a rare renal condition. We present a case of bilateral renal lymphangiectasia in a 52-year-old man who presented with bilateral flank pain and gross hematuria. The clinical features, diagnosis and characteristic imaging findings are reviewed. This condition should be considered in the differential diagnosis of cystic renal masses in both children and adults.

Key Words: renal lymphangiectasia, cystic renal masses, benign, ultrasound, computerized tomography, magnetic resonance imaging

Introduction

Renal lymphangiectasia is a benign, rare renal disorder that has been reported in both children and adults.[1,2] Most information about this condition is based on isolated case reports. In this condition, the perirenal space and renal sinus are filled by multiseptated cystic masses that may demonstrate extrarenal extension.[3,4] Patient symptoms and characteristic imaging findings using a variety of imaging modalities have been described. We report on a 52-year-old man who presented with bilateral flank pain and gross hematuria, and was subsequently found to have bilateral lymphangiectasia based on characteristic ultrasound (US), computerized tomography (CT) and magnetic resonance imaging (MRI) findings.

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Case report

A 52-year-old man with a history of intermittent bilateral flank pain and gross hematuria was referred to our institution for assessment. CT scan from the referral center demonstrated a right perirenal collection with bilateral parapelvic cysts. The flank pain was described as a "pressure" sensation. There was no history of trauma or infection. The hematuria had resolved spontaneously without intervention. The patient was asymptomatic when he was seen by our service. Past medical history was unremarkable for the exception of previous abdominal surgery for a pancreatic pseudocyst. There was no pertinent family history. Physical examination was unremarkable including a normal blood pressure. Serum creatinine was normal at 95 umol/L.

The patient underwent an abdominal US, triphasic CT of the abdomen and pelvis and an MRI of the abdomen with gadolinium-enhanced MR urography. Based on the imaging findings, a diagnosis of renal lymphangiectasis was made. Since the patient was asymptomatic and not hypertensive, conservative treatment with follow-up imaging to exclude progression was recommended.

Radiology

Imaging plays an important role in the differentiation of renal lymphangiectasia from other perinephric renal diseases. The diagnosis of renal lymphangiectasia can be confirmed with aspiration of chylous fluid from the perinephric fluid collections.[5] However, with the classic radiologic findings, a diagnosis of renal lymphangiectasia can be made confidently. The imaging findings are characteristic for this disease and allow for a specific and non-invasive diagnosis.

Ultrasound demonstrates perirenal and parapelvic hypo or anechoic lesions.[4] These lesions will demonstrate through transmission and a well delineated far wall, Figure 1. There may be debris demonstrated within, which likely indicates intra cystic hemorrhage.[4]

The cystic lesions may extend into the parapelvic region where the large perirenal lymphatics drain via the renal pedicle. Particularly in pediatric patients, ultrasound may demonstrate enlargement of the involved kidney with increased renal cortical echogenicity.[6] There may also be loss of the normal corticomedullary differentiation.[6]

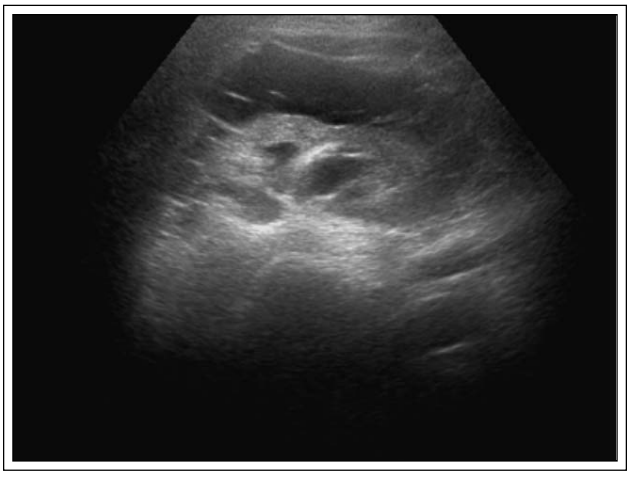

Figure 1. Sagittal ultrasound image of right kidney demonstrates perinephric hypoechoic collection with thin septations. Central hypoechogenicity is also present.

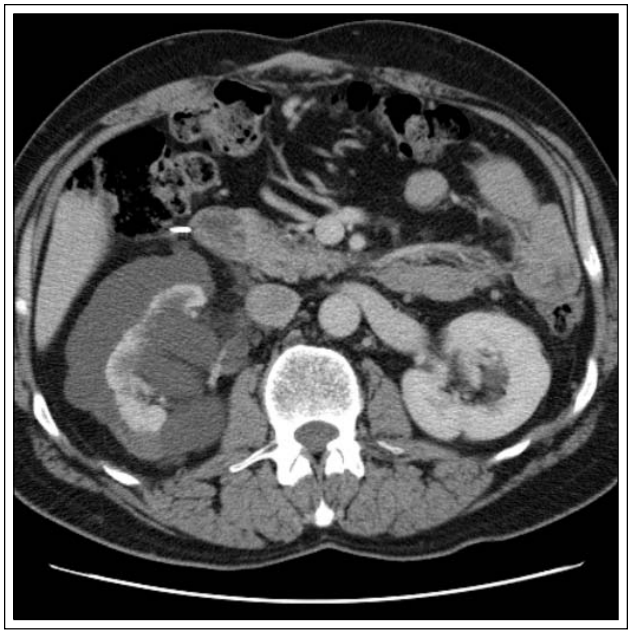

Figure 2. Axial contrast enhanced CT image through the mid kidneys demonstrates lobulated perinephric and parapelvic fluid density collection. The underlying kidney demonstrates mild atrophy.

CT examination reveals parapelvic/pararenal multilocular cystic lesions.[7] The walls and septations of these lesions are usually thin and irregular but may also be thick and irregular. The CT density is typically that of fluid (0-10), Figure 2. Higher attenuation values may be seen with intra-cystic hemorrhage. Rarely, retroperitoneal fluid collections, likely due to dilated lymphatic channels, may be present. Free intraperitoneal fluid may be seen in severe cases.[3]

Although both ultrasound and CT have been used previously to image patients with renal lymphangiectasia, MR imaging findings include low T1 weighted imaging (WI) and high T2WI signal intensity in the cystic disease,[8] Figures 3 and 4. The perirenal cysts demonstrate thin septations. Additionally, the involved kidney(s) may be enlarged and the parenchyma may show increased cortical intensity and decreased medullary intensity on T2WI. The findings of corticomedullary reversal and increased medullary signal intensity may be secondary to obstructed intrarenal lymphatics and subsequent edema. This mechanism might also explain the similar ultrasound findings of increased echogenicity and corticomedullary distinction loss. Rarely, retroperitoneal perivascular tubular lesions may be present, in keeping with enlarged lymphatic vasculature.

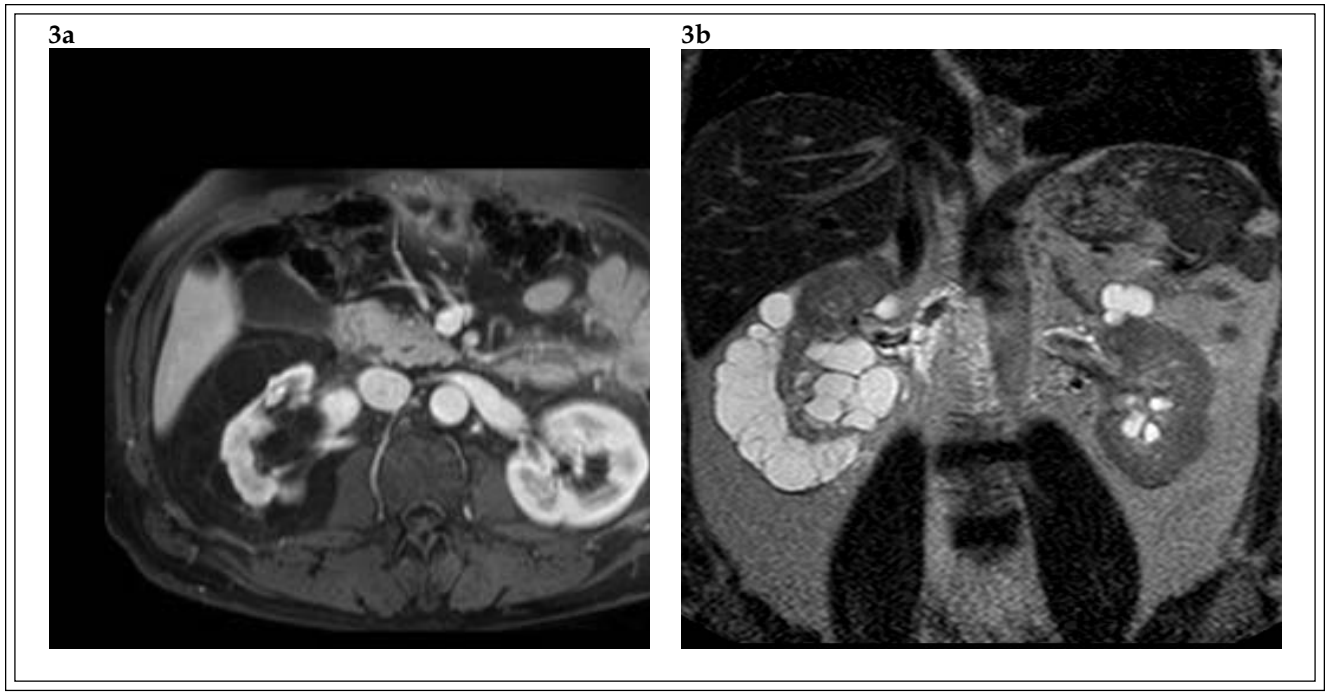

Figure 3a and 3b. Axial fat saturated T2 and coronal non-fat saturated T2 WI demonstrate lobulated perinephric and parapelvic fluid signal intensity collection with thin septations involving the right kidney. Similar changes are also identified in the contralateral kidney to a lesser degree.

The differential diagnosis of renal lymphangiectasia includes polycystic kidney disease, urinoma, and lymphoma. Adult polycystic kidney disease demonstrates scattered intra-parenchymal cysts, which vary in size, and liver and pancreatic cysts may be present. Lymphoma would demonstrate soft tissue characteristics on CT and MRI as opposed to the fluid attenuation seen with renal lymphangiectasia. Clinical history combined with typical imaging findings makes it possible to arrive at a diagnosis with certainty.

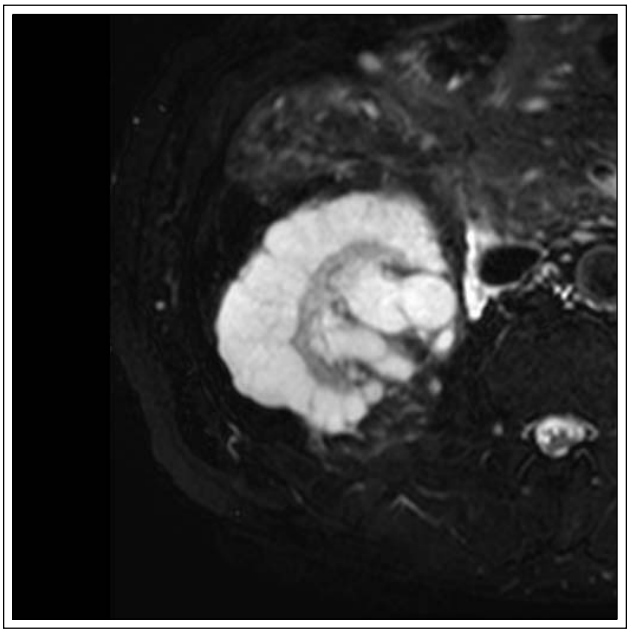

Figure 4. Axial fat saturated T1 WI post contrast demonstrates no enhancement within the cystic abnormality.

Comment

Renal lymphangiectasia is a benign, rare renal condition. The condition is characterized by ectatic perirenal, peripelvic, and intrarenal lymphatic vessels.[4,9] The pathophysiology of this condition is unclear. The lymphatics of the renal capsule and renal parenchyma drain into the renal sinus lymphatics. These lymphatic channels then empty into the paraaortic, paracaval, and interaortocaval lymph nodes. A disturbance in renal sinus drainage results in the characteristic ectatic perirenal, peripelvic, and intrarenal lymphatic vessels which is responsible for the typical imaging findings.[8] Suggested etiologic factors include both congenital conditions and acquired obstructive inflammatory processes.[1,3] Some have suggested that renal lymphangiectasia represents a benign neoplastic process.[1,3]

Patient symptoms and signs described in the

literature for renal lymphangiectasia include hematuria, abdominal and flank pain, abdominal mass, lower extremity edema, and hypertension in any age group.[4,7] Some cases have been reported with renal insufficiency, renal vein thrombosis, large perinephric fluid collections and ascites.[1,10] Hypertension has been reported in about 50% of unilateral cases and in about 15% of bilateral cases. The cause of hypertension is presumed to be due to the compressive effect of perirenal and parapelvic cysts on the intrarenal arterial circulation stimulating a renin-dependent hypertension.[11]

The natural history of this benign condition is unknown. Renal lymphangiectasia can appear suddenly, grow rapidly, cease growth or even regress spontaneously. Partial regression has been reported in a neonatal case with bilateral nephromegaly.[12] Spontaneous resolution in a 30-year-old man with a large perirenal collection and ascites has been reported following 6 years of observation. This latter case, however, was also associated with increasing nephromegaly because of worsening intrarenal lymphatic ectasia.[3]

The diagnosis of renal lymphangiectasia is based on clinical information combined with characteristic imaging findings from US, CT and MRI. The diagnosis can be confirmed with needle aspiration of chylous fluid from the perinephric fluid collections.[5] Needle aspiration, however, is seldom required as a diagnosis can be made confidently based on radiologic findings.

Treatment options for renal lymphangiectasia vary according to the symptoms, complications and site of involvement. Treatment includes percutaneuous drainage, marsupialization or nephrectomy.[5] Asymptomatic cases do not require treatment.

In the case presented here, the diagnosis of renal lymphangiectasis was based on the characteristic imaging findings. No invasive procedures to confirm the diagnosis were deemed appropriate in this patient. Conservative management was elected as the patient's symptoms resolved spontaneously. Follow-up imaging was recommended to rule out progression of this condition. □

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