JMS 2021; 4(3):166-170
UDC:616.423-007:[616.61-073.756.8:004

RENAL LYMPHANGIECTASIA- AN UNUSUAL MIMICKER OF RENAL HYDRONEPHROSIS

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Abstract

Renal lymphangiectasia is a rare malformation of the renal lymphatics. It occurs in both children and adults and can be unilateral or bilateral with no gender predilection. It is a condition characterized by different degrees of dilatation of the lymphatic ducts. Because of the rarity, it can be easily misdiagnosed for other cystic masses, most commonly peripelvic cysts, renal cysts or hydronephrosis. Usually is asymptomatic and incidental finding, but in severe cases can lead to hypertension and renal failure.

We report a case of unilateral renal lymphangiectasia in adult male patient with medical history of hydronephrosis and characteristic radiologic CT findings. From imaging methods, we conducted ultrasound (US), contrast enhanced computed tomography (CT) and CT angiography, because they have an important role as diagnostic procedures to recognize renal lymphangiectasia.

Kidney cystic lesions revealed on ultrasound and confirmed on CT as hypodense intrarenal multiloculated findings, as well as fluid attenuation collections, not always go in favor of hydronephrosis. Knowledge of the radiological findings associated with renal lymphangiectasia can contribute to better differentiation from other conditions and with the right diagnosis, successful management and treatment can be provided for the patient.

Keywords: renal lymphangiectasia, hydronephrosis, computer tomography

Introduction

Renal lymphangiectasia (RLM) is a rare, benign entity of the renal lymphatics. It occurs in both children and adults and can be unilateral or bilateral with no gender predilection. [1]. It is a condition characterized by different degrees of dilatation of the lymphatic ducts. In rare occasions it develops in the kidney, where it is believed to be due to an abnormal communication between the renal lymphatics and the larger retroperitoneal lymphatics [2].

Knowledge about this condition is limited and based on isolated case reports [6]. Because of the rarity, it can be easily misdiagnosed for other cystic masses, most commonly peripelvic cysts, renal cysts or hydronephrosis [1].

Usually is asymptomatic and incidental finding, but in severe cases can lead to hypertension and renal failure [1].

Imaging with US, CT and MRI has an important role in differential diagnosis and the diagnosis can be confirmed with needle aspiration of chylous fluid [6].

The objective of this study is to show the role of contrast enhanced computed tomography in differentiating Renal Lymphangiectasia.

Case report

We present a case of a 30-year-old male patient with past medical history of essential hypertension, allergic asthma and right-sided hydronephrosis. On the previous ultrasound was detected cystic dilatation of the renal calyxes in the right kidney and there was also presence of a large kidney calculus. There is no other information about any illnesses or hospitalizations due to urinary tract infections.

Clinical symptoms of the patient were: increased blood pressure, fatigue, headache and lumbar pain on the right side. He was sent for further laboratory investigations and all of the lab analysis were within normal referential values.

The patient was also sent for ultrasound, CT urography and CT angiography. A grayscale ultrasound scan of the abdomen showed that the right kidney was enlarged with dimensions of 160x74mm, granulated parenchymal structure, hydronephrosis grade 3 (moderate) and calculus in the pyelon. The left kidney's dimensions were 112x44mm, also with granulated parenchymal structure, but there were no signs of hydronephrosis or calculus in the pyelocaliceal system.

CT urography showed that the left kidney was with regular postcontrast imbibition, but there was a calculus, 4mm (axial plane), in the renal calyx of the middle posterior group, without repercussions in eliminating the contrast. The CT images revealed a voluminously enlarged right kidney with thinned cortex. There were fluid attenuation collections of 0-15 HU, peripelvic, intrarenal, multiple lobulated, which suppressed some of the calyxes, therefore they were flattened (Fig.1). On delayed scans in the excretory phase there was non-opacification of these lesions (Fig.2)

The contrast normally eliminates through the pyelocaliceal system, without repercussions on the structures. The right renal vein was smaller in caliber and perirenal venous collaterals of blood vessels were seen on the same side. From the renal sinus towards the infrarenal, paraaortic and retroperitoneal part were seen changes which diagnostically associate with ectopic and enlarged lymph nodes. Imaging features were suggestive of peripelvic renal lymphangiectasia on the right side. Case follow up was required with CT angiography. The rest of the abdominal organs were normal.

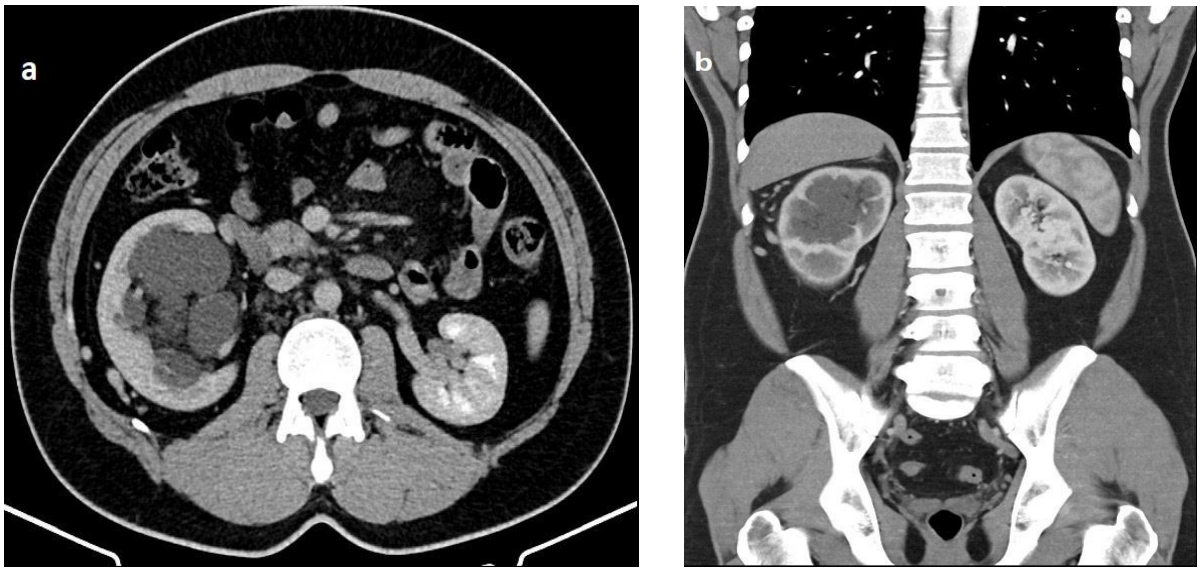

Figure 1: Axial (a) and coronal plane (b) of contrast enhanced CT of the abdomen in the arterial-venous phase demonstrates enlarged voluminously right kidney, with fluid attenuation collections, hypodense lesions that are multiple lobulated, peripyelonic and intrarenal. On the right perirenal side venous collaterals of blood vessels are seen and ectopic and remarkable lymphatic nodes

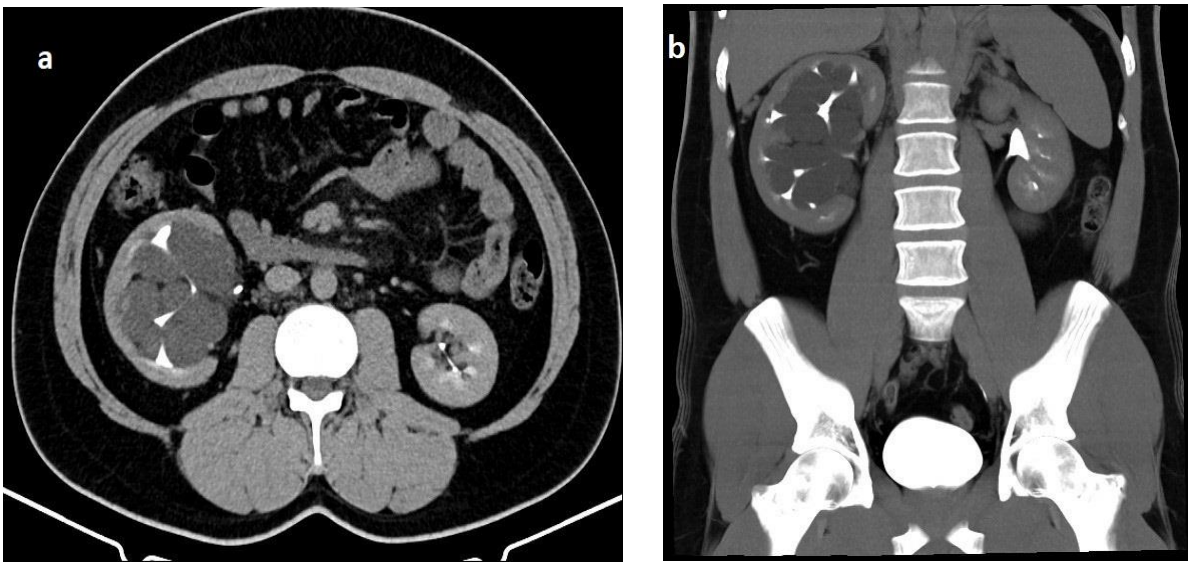

Figure 2: Axial (a) and coronal (b) plane of contrast enhanced CT of the abdomen in the delayed phase demonstrates no opacification with iv contrast of the fluid collections

The following CT angiography showed a tortuous network of perirenal varicose dilated veins which were entering the right kidney vein, with remarkable lymphatic nodes and lymphatic ducts retroperitoneally, around aorta and v. cava inferior. All indicates that is renal lymphangiectasia of the right kidney. The patient has been treated conservatively and he is on follow up appointments. (Fig.3)

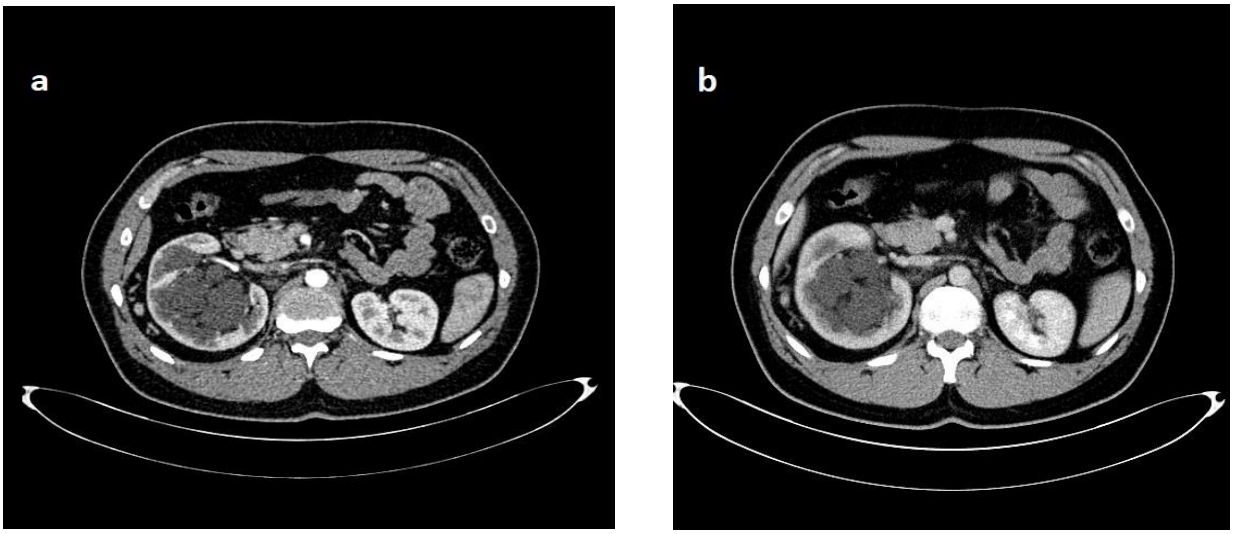

Figure 3: Ct angiography axial (a,b) planes demonstrate the attenuated caliber of the right renal vein.

Discussion

Renal lymphangiectasia is a rare benign disease that happens because of miscommunication between the two lymphatic drainage systems, the renal and the retroperitoneal lymphatic system [9].

The most characteristic feature is the accumulation of lymph in the renal lymph ducts, forming simple or multiloculated, dilatated, collections in the pyelocaliceal, perirenal, peripelvic, and intrarenal lymphatic vessels [6,7,9].

It can be manifested as unilateral and bilateral as well as in focus and diffuse [1,7]. Renal lymphangiectasia is being diagnosed as an accidental disease because it is usually asymptomatic, unrelated to gender or age, it can affect both children and adults [2,10].

In literature this disease is known with different names such as renal lymphangiomatosis, renal lymphangioma, peripelvic lymphangiectasia, renal sinus polycystic disease, renal hygroma [2]. The etiology of the disease can be genetic, (developmental and acquired) [1].

Symptoms of this disease are: abdominal pain, flank pain, bloating of the stomach, fatigue, fever, weight loss, hematuria, proteinuria [3, 4].

In the most severe cases it is manifested with high blood pressure and ascites, renal insufficiency and renal vein thrombosis or these can be complications when the disease will not be very well threated [4, 5]. But in literature hypertension associated with renal lymphangiectasia is rarely reported [10].

Hypertension is presumed to be due to excessive renin release, when subcapsular collection is causing compression of renal parenchyma [7].

In our case all the imaging methods were in favor of unilateral renal lymphangiectasia. Ultrasound imaging revealed right-sided hydronephrosis grade 3 (moderate).

But, in hydronephrosis there is dilatation of the collecting system, furthermore, CT examination showed fluid attenuation collections of 0-15 HU, mimicked cystic lesions, which suppressed some of the calyxes and only displace and minimum flattened collecting system, on contrast - enhanced CT there is opacification of the collecting system of delayed scans, but there is no opacification of cyst on delayed scans in contrast- enhanced CT, as it is mentioned in literature for RML [4], these findings confirmed our diagnose for RML.

The differential diagnoses of this disease are: hydronephrosis, which is more common, polycystic kidney disease, nephroblastomatosis, lymphoma, multilocular cystic fibrosis [2,5]. The disease is also confirmed by aspiration and by the cytology of the aspirated fluid [2].

Treatment of the disease in asymptomatic cases is not necessary but regular control is required due to complications that may occur as a result of the disease. For a conservative treatment there are antihypertensive medications for patients with arterial hypertension, and diuretics for ascites [8].

In symptomatic cases, percutaneous aspiration is performed in many situations, or nephrectomy for severe cases such as Page kidney and renal vein thrombosis [2, 4, 8]

Conclusion

Kidney cystic lesions revealed on ultrasound and confirmed on CT as hypodense intrarenal multiloculated findings, as well as fluid attenuation collections, not always go in favor of hydronephrosis. Even more, associated with dilated lymphatic channels are findings which conduct radiologist for correct diagnose of renal lymphangiectasia.

Knowledge of the radiological findings associated with renal lymphangiectasia can contribute to better differentiation from other conditions and with the right diagnosis, successful management and treatment can be provided for the patient.

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