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Case Report

Computed Tomography Imaging of an Unusual Case of Bilateral Renal Lymphangiectasia

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Abstract

Renal lymphangiectasia is a very rare disorder characterized by dilatation of the perirenal and peripelvic lymphatics. Only 22 cases of renal lymphangiectasia have been described during the past 25 years in the literature [1-6]. It is commonly misdiagnosed as hydronephrosis or polycystic kidney disease. Here, we present a case of 57 years old male patient diagnosed as bilateral renal lymphangiectasia presenting with complaints of left flank pain since 2 weeks and decreased appetite. Case was evaluated on Contrast Enhanced CT (CECT).

Keywords: Renal lymphangiectasia; Cysts; Lymphatic malformation; Multimodality imaging; Lymphoscintigraphy; Management

Introduction

Retroperitoneal lymphangiectasia is a rare condition accounting for ~1% of all lymphangiectasias [1]. Among them, renal lymphangiectasia is a very rare benign disorder of the kidney lymphatic malformation with no specific clinical presentations. It is non-communication of the perirenal and peripelvic lymphatics with the main lymphatics. The condition is often misdiagnosed as other cystic lesions such as polycystic kidneys and hydronephrosis. Clinically, it may be asymptomatic or present as abdominal pain, hematuria or a palpable mass. Here we report a case of bilateral renal lymphangiectasia diagnosed on the basis of Ultrasound (US) and Computed Tomography (CT) imaging findings.

Case Report

A 57 years old patient presented to the emergency department with complaints of left flank pain and decreased appetite from 2 weeks. On clinical examination, patient was a febrile but found hypertensive with a blood pressure of 150/100 mmHg. Physical examination was normal. Laboratory tests were unremarkable. His haemoglobin, total leucocyte count and differential leucocyte counts were within normal limits. His urea level was 27 mg/dl (normal range: 15.0 – 45.0 mg/dl), creatinine was 1.0 mg/dl (normal range: 0.5 – 1.2 mg/dl) and uric acid was 6.7 mg/dl (normal range for males: 3.5 – 7.2 mg/dl).

Then he underwent an ultrasound abdomen for evaluation left flank pain which shows the dilatation of left pelvicalyceal system and appears as a hydronephrosis and minimal dilatation was noted on the right side. No obstruction of bilateral ureters revealed on ultrasound so patient was evaluated further with a Contrast Enhanced CT (CECT) imaging. CECT reveals no significant enlargement of kidney sizes (Right kidney measures 8.2 x 4.7 cm and left kidney measures 9.6 x 6.0 cm). A hypodense cystic non-enhancing lesion measuring 3.0 x 6.0 cm seen filling the left renal sinus. Similar smaller cystic non-enhancing hypodense lesion measuring 3.6 x 1.0 cm seen filling the right renal sinus both the cystic lesions are seen stretching the bilateral renal sinus.

Also, multiple small cortical cysts noted in both the kidneys largest measuring ~ 1.0 cm.

Based on the imaging findings diagnosis a of bilateral renal lymphangiectasia was made. The patient was managed conservatively.

Discussion

Renal lymphangiectasia also known as renal lymphangiomatosis is a rare benign malformation involving the lymphatic system of the kidneys (perirenal, peripelvic and intrarenal lymphatics). The exact pathogenesis of renal lymphangiectasia is not known but is thought to be due to failure of the perinephric lymphatics to communicate with the main lymphatic system. It is observed that only cortex is involved and it is characterized by dilatation of endothelial-lined spaces without glomerular or tubular abnormalities [2]. Renal lymphangiectasia is not specific to any age group. Familial association of the condition have been observed in few cases. It is mainly asymptomatic or if symptomatic, patients present with flank pain, hematuria, abdominal distension, fever but rarely impaired renal function. However, there can be sudden appearance and rapid growth or cessation of growth and even spontaneous regression of symptoms. The differential diagnosis of renal lymphangiectasia includes hydronephrosis, polycystic kidney disease, urinoma, and lymphoma.

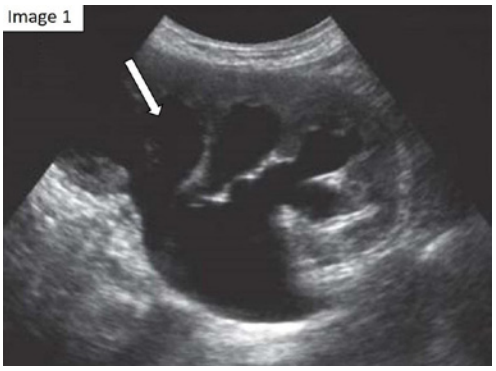

Image 1: Ultrasound image of left kidney showing the dilatation of left pelvicalyceal system and appears as a hydronephrosis (Arrow).

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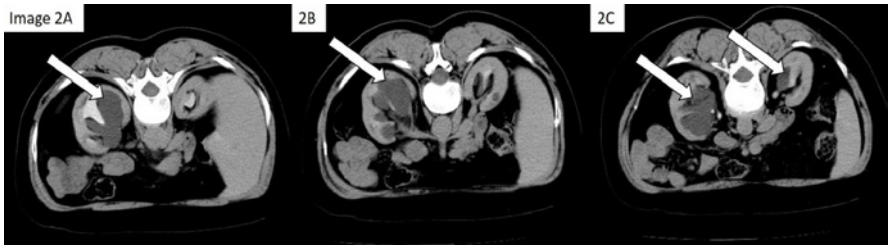

Image 2: A, B and C: Axial contrast enhancing CT urogram phase shows cystic density along the renal pelvicalyceal systems (Arrows).

Clinical manifestations and its correlation with typical imaging findings are helpful to differentiate renal lymphangiectasia from other similar conditions. Prior to the advent of CT scan, the diagnosis was usually made at the time of exploratory laparotomy or after nephrectomy. Isolated renal sinus lymphangiectasia on US can closely mimic pelviureteric junction obstruction [3]. Scattered intra-parenchymal cysts of varying size are seen in Adult Polycystic Kidney Disease (APKD) associated with or without liver and pancreatic cysts. Focal masses with soft tissue attenuation are seen in lymphomas, while in renal lymphangiectasia cysts with fluid attenuation are found [4]. Nephroblastomatosis is also a differential diagnosis made in children with renomegaly and soft-tissue masses.

Ultrasound examination in renal lymphangiectasia demonstrates perirenal or parapelvic hypo or anechoic lesions. These lesions are usually appears as loculated, cystic, septate collection. There may be debris within the lesions which most likely indicates intra cystic haemorrhage. These cystic lesions may extend to parapelvic region where large perirenal lymphatics drain via renal pedicle [7]. Kidney may be enlarged or normal in size. There may also be loss of corticomedullary differentiation.

CT examination reveals peripelvic or perirenal multiloculated cystic lesions with lower fluid attenuation (0 to 10 Hounsfield units) but the septations may not be delineated as clearly as in Ultrasound [8]. Higher attenuations are generally secondary to intracystic haemorrhage. Rarely, retroperitoneal fluid collections may be present. Free intraperitoneal fluid may be seen in severe cases [9].

Magnetic Resonance Imaging (MRI) can also be used to diagnose renal lymphangiectasia which demonstrates cystic lesions that are hypointense on T1-Weighted Image 1 (T1WI) and hyperintense on T2-Weighted Image 2 (T2WI). Corticomedullary intensity reversal will be seen in which cortex will appear hyper intense and medulla will be hypointense on T2WI because of obstructed intra-renal lymphatics and subsequent edema [10].

Nowadays, lymphoscintigraphy is also used to detect abnormal lymphatic flows and helps in differentiating renal lymphangiectasia from other similar conditions [11].

Conservative management is usually preferred choice of treatment for the asymptomatic patients. If patients are symptomatic then percutaneous drainage can be done to relieve symptoms. However, surgery (cyst marsupialization) is considered as the last therapeutic measure and it may result in nephrectomy because these lesions tend to bleed easily during operation [7].

Conclusion

Renal lymphangiectasia is a rare benign condition of lymphatics malformation which closely mimics the hydronephrosis and bilateral polycystic disease of kidney. Radiologist can easily nearly conclude the diagnosis based on the imaging findings on contrast enhanced CT or MRI with urogram, and MRI if needed. CT has a very important role in diagnosing the extent of this condition and following up. This condition is mostly managed conservatively.

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