

Bilateral peri-renal lymphangiectasia

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Case Report

Abstract: Renal lymphangiectasia is a very rare benign disorder where there is dilatation of the perirenal and peripelvic lymphatics. The condition is commonly misdiagnosed as other cystic lesions of kidney. We present USG and CT findings of bilateral peri renal lymphangiectasia in a 24 year old male patient, which was diagnosed, based on the typical imaging findings and the lab reports and the patient was managed conservatively.

Keywords: Renal lymphangiectasia, cystic renal masses, benign, ultrasound.

Introduction

Case history

A 24 year old male patient presented with the complaints of pain abdomen and abdominal distension since 2 years. He had one episode of hematuria one week back. Patient had no history of fever, diabetes mellitus, hypertension or any previous abdominal surgeries. On clinical examination patient was afebrile and had blood pressure of 150/110 mm Hg. His abdominal examination demonstrated the presence of ascites. Routine investigations revealed hemoglobin level of 15.6mg%, Total leucocyte count and the differential leucocyte counts within normal limits, ESR 12 mm/hour, Random blood sugar 80mg/dl, Blood urea levels 20mg/dl and serum creatinine 0.9mg/dl. Urine examination was insignificant indicating that there was no renal function compromise. Further, the patient underwent imaging evaluation. On plain X-ray there was homogenous soft tissue density mass in bilateral renal fossa and it was displacing the bowel loops and the adjacent structures. Further to know the origin of the mass we did an ultrasound examination of the patient which demonstrated bilaterally enlarged echogenic kidneys and corticomedullary differentiation was not maintained. Interestingly there was large clear multiseptated anechoic fluid collection located in the perinephric space indenting the surface of the kidney and mild hydronephrosis on right side. Ascites was present. No cysts were seen in other organs. To know the extent and relation of the fluid collection to adjacent structures, Contrast enhanced CT scan on a siemens somatom emotion 6 slice helical scanner was done. Both kidneys were enlarged with normal contrast enhancement and excretion. However the reniform shape of the kidneys was maintained.

Hydronephrosis and blunting of the calyces on right side was seen. The left Pelvicalyceal system was normal. Renal arteries and the renal veins on both sides were normal. Ureters were delineated all along their course and appeared normal. No other significant findings were seen on CT scan. The fluid collection seen on the ultrasound was localized in the perinephric space. It was seen to cross the midline anterior to the inferior vena cava and the aorta to communicate with the opposite side. The Hounsfield units of the fluid collection were ranging from 3 to 10 HU. No soft tissue density mass or fat attenuation mass were seen within the fluid collection. To know the nature of fluid collection it was aspirated under ultrasound guidance and was sent for analysis. It was clear straw coloured clear fluid and showed Urea 14.7 mg/dl, Total protein 770mg/dl, Protein electrophoresis selective protein band in albumin region, Triglycerides were negative, few cells were seen and all were lymphocytes. No organisms on gram stain and acid fast stain and no growth on culture were seen. Based on the typical imaging findings and the laboratory analysis of the fluid patient was diagnosed to have bilateral perirenal lymphangiectasia. Patient was conservatively managed with Antihypertensives and diuretics following which patient condition improved.

Discussion

Bilateral perirenal lymphangiectasia is a very rare benign condition which is often mistaken for the other forms of cystic diseases of the kidney [2]. It is known by different synonyms such as renal lymphangiomatosis [2], hygroma renale [4] and polycystic disease of the renal sinus [3]. The differential diagnosis to be considered in this condition are polycystic kidney disease [8], multilocular cystic nephroma [8], urinoma or abscess and bilateral peri renal liposarcomas [5]. The cysts in the polycystic kidney disease and the multilocular cystic nephroma are parenchymal and perirenal liposarcomas show the presence of macroscopic fat [5]. The exact pathogenesis of this condition is not known and is thought to be due to failure of the lymphatics to communicate with the main lymphatic

system [3]. There is no particular age or sex predilection for this condition and familial association is seen in few cases [2]. Clinical course of this condition can be varied with sudden appearance, rapid growth, cease growth, or even spontaneous regression [1]. Most commonly this condition is asymptomatic which is diagnosed incidentally. When symptomatic abdominal pain, hematuria, hypertension and renal failure can be presenting features [3]. Exacerbations during pregnancy can be seen [2]. On imaging it is usually seen as large homogenous soft tissue mass displacing the adjacent structures on plain radiography. There can be hydronephrosis and distortion of the Pelvicalyceal system on intravenous excretory urography[8]. On ultrasound examination it can be seen as bilateral multiseptated thin walled fluid collection localized in the perinephric and peripelvic space with normal renal parenchyma[5]. Kidneys may be enlarged, echogenic and corticomedullary differentiation may be lost[8]. It can be seen as a solid mass when the smaller intra renal lymphatics are blocked[6]. Ascites can also be present. CT examination shows the presence of low density fluid collection (0 to 10 HU) in the perinephric space and septa may not be delineated very clearly [4]. There will be no invasion or erosion of the adjacent structures. No evidence of cysts in other organs are seen [7]. Laboratory analysis of the fluid shall be helpful by demonstrating very low proteins and fat levels [4], sparse cells which will be lymphocytes. Hematuria, ascites, hypertension and deterioration of the renal function are the known complications of this condition; however hypertension and renal failure are known to improve following treatment [4, 7]. Most of the asymptomatic cases need no treatment [8]. The treatment options available are marsupilisation [4] or nephrostomy when collections become large. Antihypertensives and diuretics can be used to reduce blood pressure and fluid secretion [8]. Nephrectomy can be done in complicated cases [8]. To conclude we present a rare case of bilateral perirenal lymphangiectasia in a young male patient with ascites, which was diagnosed based on the typical imaging findings and lab investigations. Knowledge of this condition shall be helpful in the differential diagnosis of cystic diseases of the kidney and arriving at correct diagnosis.

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Legends



Figure 1: Plain radiography showing homogenous soft tissue density in region of kidney bilaterally (arrow head) displacing bowel loops (solid arrow) and adjacent structures.



Figure 2A: USG image of the right kidney showing multiseptated perinephric collection (arrow heads). Cortico-medullary differentiation is lost (solid black arrow).



Figure 2B: USG image of the left kidney showing multiseptated perinephric collection (arrow heads) with indentation of the surface (white arrow). Cortico-medullary differentiation is lost.



Figure 3: Plain axial CT scan showing bilateral low density perinephric collection (arrow heads)



Figure 4: contrast enhanced axial CT scan showing bilateral low density perinephric collection (black arrows) with normal enhancement of the renal parenchyma and communication (arrow heads) with the opposite side in the midline.

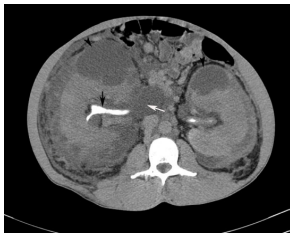


Figure 5: Perinephric (arrow heads) and the Para pelvic (white arrow) collection with minimal right sided hydronephrosis (black arrow) and normal excretion of the contrast bilaterally.

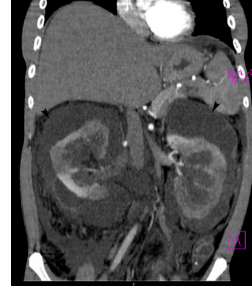


Figure 6: Reformatted coronal image showing bilateral enlarged kidneys with perinephric collection (arrow heads). Renal vessels are normal. Ascites is present (black arrow).

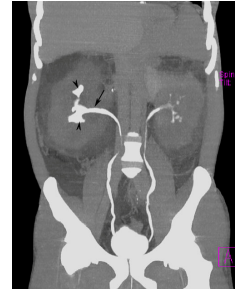


Figure 7: Reconstructed CT excretory urography showing right sided mild hydronephrosis (solid black arrow) with blunting of the calyces (arrow heads) and bilateral normal ureters.