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Case Report

Computed tomography in the diagnosis of childhood renal lymphangiectasia

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ABSTRACT

A 14-year-old male patient presented with a history of intermittent bilateral flank pain for 2-3 months. There was no history of any urinary complaints or trauma. Renal function test was normal. Computed tomography (CT) scan of the abdomen revealed bilateral perinephric and peripelvic fluid collections, which were almost symmetrical. A provisional diagnosis of bilateral renal lymphangiectasia was suggested which was confirmed by laboratory analysis of aspirated perinephric fluid. Renal lymphangiectasia is a very rare benign disorder of renal lymphatics. CT can noninvasively diagnose this condition resulting in early treatment and reduced morbidity.

Keywords: Computed tomography, Perinephric, Renal lymphangiectasia

INTRODUCTION

Renal lymphangiectasia, also known as renal lymphangiomatosis is a rare benign malformation involving the lymphatic system of the kidneys (perirenal, peripelvic and intrarenal lymphatics).[1] Renal lymphangiectasia may present at any age, with equal incidence in men and women. It arises as a result of failure of renal lymphatic ducts to drain in to larger retroperitoneal lymphatics.[1] This may be a congenital abnormality or an acquired dysfunction of lymphatics. Early diagnosis of this condition noninvasively will result in early treatment and reduced morbidity.

CASE REPORT

A 14-year-old male patient presented with a history of intermittent bilateral flank pain for 2-3 months. There was no history of any urinary complaints or trauma. On examination, he was pale, and his blood pressure was 170/110 mm Hg. Baseline investigations revealed haemoglobin level 9.1 g%, total leucocyte count 5800/mm^3, blood urea 27 mg/dl, serum creatinine 0.9 mg/dl, serum albumin 4.5 g/dl and serum potassium 3.9 mEq/l. Ultrasound (US) examination revealed bilateral, septated, perinephric collection.

To determine the cause of bilateral perinephric collection, a computed tomography (CT) scan was done (Figures 1-3) which revealed bilateral perinephric and peripelvic fluid collections, which were almost symmetrical. The Hounsfield units value of the collections ranged from 0 to +20. Few internal septations and few hyperdense foci of haemorrhage were noted on either side. The renal parenchyma was normal on both sides. The renal pelvis and calyces appeared normal in calibre. No calculi were found in either kidney or ureter, and the renal veins were patent bilaterally. On delayed contrast scan after 10 min (Figure 3), there was no leakage

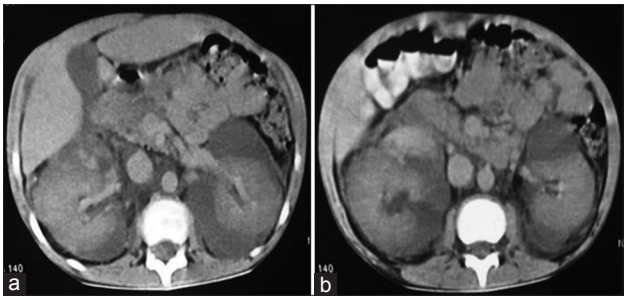

Figure 1: Renal lymphangiectasia. Non-contrast computed tomography abdomen axial images show perinephric fluid collection bilaterally, with hyperdense focus of haemorrhage in the collection anterior to midpole of right kindney.

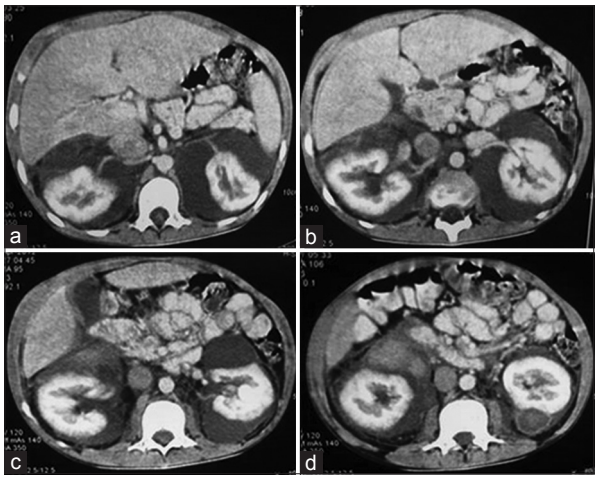

Figure 2: Renal lymphangiectasia. Contrast enhanced computed tomography abdomen axial images show perinephric and peripelvic fluid collection bilaterally, with hyperdense foci of haemorrhage in the collection anterior to midpole of right kidney and posterior to lower pole of left kidney.

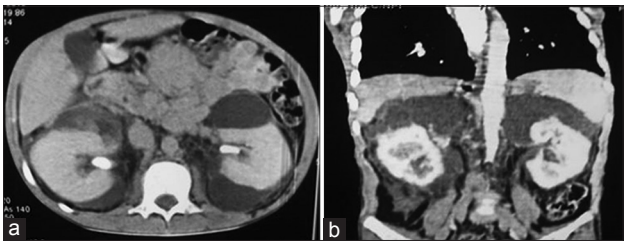

Figure 3: Renal lymphangiectasia. Delayed computed tomography (CT) abdomen axial image shows no leakage of intraveneous contrast into bilateral perinephric collection. Contrast enhanced CT coronal image shows bilateral perinephric fluid collection.

of intravenous contrast into bilateral perinephric collection. No free fluid was noted in the peritoneal cavity. Based on the clinicoradiologic features, a provisional diagnosis of bilateral renal lymphangiectasia was suggested.

Fluid aspiration and cytology followed by drainage under US guidance was performed for confirming the diagnosis and as part of treatment. Approximately 500 mL of chylous fluid was drained from both sides. Laboratory analysis revealed that the aspirated fluid was rich in protein and contained abundant renin. Microscopic examination revealed many red blood cells and only few lymphocytes. No organisms were isolated on culture of aspirated fluid. Thus, the diagnosis based on CT findings was confirmed.

The patient was put on diuretics and antihypertensives and was followed-by serial US examinations for 1 year, which did not reveal any significant increase in perinephric collection. The antihypertensives were withdrawn after 6 months since the patient's blood pressure had returned to normal limits because of therapeutic percutaneous drainage of perinephric collection and diuretic treatment.

DISCUSSION

Renal lymphangiectasia is a rare benign malformation involving the lymphatic system of the kidney subsequently leading to formation of unilocular or multilocular cystic spaces in the pelvic sinus and perinephric spaces.[1]

Renal lymphangiectasia may be seen at any age. Clinically, it is usually asymptomatic. When symptomatic, the most common presentations are abdominal pain (42%), abdominal distension (21%), followed by fever, hematuria, fatigue, weight loss, hypertension and occasional deterioration in renal function (mostly reversible).[1,2]

On CT scan, renal lymphangiectasia appears as a well contained, fluid attenuating collections in the peripelvic or perinephric space, as in our case, with or without demonstrable septations with normal renal parenchyma.[1,3] The pelvicalyceal system is normal with no evidence of any renal or ureteric calculi, as in our case. There is no leakage of intravenous contrast into perinephric collections on delayed CT. All the other differential diagnosis like nephroblastomatosis, lymphoma and multilocular cystic nephroma can be distinguished from renal lymphangiectasia on CT scan by the fact that all of them involve the renal parenchyma and the former two are solid in attenuation.[3,4] The other differential diagnoses of perirenal collections are urinoma, hematoma and abscess. Urinoma, hematoma and abscess are usually unilateral. In urinoma, there is leakage of intravenous contrast into the perinephric collection on delayed CT. Haematoma can be identified by its hyperdense character on non-contrast CT and associated renal injury in the presence of relevant history of trauma. In abscess, peripheral enhancement is seen around the fluid collection with associated pyelonephritis in renal parenchyma.

Complications of undiagnosed/untreated cases include hematuria, ascites, hypertension, renal vein thrombosis and impairment of renal function.[2] Hypertension due to renal compression by perinephric fluid collection in renal lymphangiectasia is reported in literature.[2,5] Fluid aspiration and cytology of the perinephric collection not only confirms the diagnosis but also excludes the presence of urinoma and abscess. Renal origin of lymph can be confirmed

by demonstrating high levels of rennin in the aspirated perinephric fluid collection as seen in our case.[2-5]

Asymptomatic cases or small collections are treated conservatively while percutaneous drainage is indicated in large and symptomatic collections causing pressure symptoms or other complications related to renal lymphangiectasia.[4] Our patient's blood pressure normalized gradually after percutaneous drainage, suggesting hypertension secondary to renal compression. Similar observation is reported in the literature.[5] Deterioration has been described during pregnancy. Recurrent collections can be treated by marsupialization where a communication is made between the collection and the peritoneal cavity.[2] Ascites and hypertension can be treated with diuretics and antihypertensives, as in our case. Nephrectomy is reserved for cases with recurring or complicated collections and uncontrollable complications.[4]

CONCLUSION

Fluid collections around the kidneys may be caused by urine, blood, pus, lymph, or plasma. CT can not only show and characterize the fluid, but also may help determine the underlying cause of the perinephric fluid collection, such as ureteric obstruction, kidney injury, infection, or renal lymphangiectasia. CT scan can play a very important role in the diagnosis of renal lymphangiectasia, thus reducing the morbidity associated with this condition. Definitive diagnostic test is aspiration of perinephric fluid collection and its laboratory analysis including renin estimation.

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