Case Report: **Renal Lymphangiectasia**

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Abstract:

A young female presented with complaints of abdominal pain and feeling of abdominal fullness for the last 5 years. Her abdominal ultrasound showed bilateral perinephric cystic collections. CT with IV contrast revealed presence of bilateral renal lymphangiectasia. It is a rare benign condition considered as differential in cases of renal cystic masses in adults and children.

Key words: renal lymphangiectasia, cystic mass, benign

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Introduction:

Renal lymphangiectasia is a rare benign form of cystic mass and till now only 42 cases have been reported [1]. Its pathogenesis remains unclear and most of information is based on case reports [2]. It can be primary (dilative) or secondary (obstructive) in nature [3]. Familial case have been reported as well [4]. We, hereby, describe a case of young girl with bilateral renal lymphangiectasia.

Case Presentation:

An 18yrs old female presented with complaints of abdominal pain and fullness for the last 5yrs. Initially the symptoms were mild but over a past few months she reported an aggravation of symptoms. Now the pain wasn't resolving despite oral and intramuscular analgesics.

Apart from having a thin, lean physique, her physical examination was normal including her blood pressure. Baseline investigation showed normal blood counts and normal serum urea (30mg/dl) and creatinine (0.8mg/dl). Ultrasound abdomen suggested bilateral perinephric cystic collections. CT with IV contrast showed bilateral perinephric cystic collections with a few internal septations and a radio density of 1.5-3 HU, with marked distortion of the renal contours suggestive of bilateral renal lymphangiectasia.

Approximately 2000ml of fluid was aspirated from right sided perinephric collection. Analysis of the fluid revealed a colorless fluid with low protein content (0.5 g/dl) and few lymphocytes (17/cmm). Fluid creatinine level was 0.6mg/dl that was similar to serum creatinine levels .Fluid cultures were negative for any organism and ZN staining was negative for AFB.

All the investigations were suggestive of bilateral renal lymphangiectasia. Patient improved symptomatically after fluid aspiration and was managed conservatively afterward.

Discussion:

Renal lymphangiectasia is a rare disorder of lymphatic vessel characterized by dilatation of the perirenal, peripelvic or intrarenal lymphatics. The proposed mechanism leading to dilation is the abnormal drainage of small lymphatic vessels into the major lymphatics [5]. The definitive pathophysiology remains unknown.

Renal lymphangiectasia can be an incidental finding and patient may remain asymptomatic. Others may present with abdominal pain, abdominal distension, hematuria, hypertension and occasional reversible deterioration of renal functions. Hemorrhagic complication such as intracystic hemorrhage and renal vein thrombosis have also been reported [3]. Exacerbation during pregnancy has been seen as well[4].

The differential diagnosis of renal lymphangiectasia includes cystic renal dysplasia, ADPKD, ARPKD, nephroblastomatosis, lymphoma and multilocular cystic nephroma according to the age of presentation[3,6].

Diagnosis is based on radiological finding on ultrasound and CT scan or MRI. Ultrasound findings may show enlarged renal size (involvement of intra renal lymphatics), perirenal collection and peripelvic cysts, retroperitoneal fluid collection, ascites, poor corticomedullary differentiation and diffuse echogenic renal parenchyma. CT may show a perinephric fluid with a radiodensity of usually 0-10 HU. Distortion of renal contours may also be seen due to pressure effects. Fluid collection in peritoneal or retroperitoneum compartments may be noted.

No definitive treatment algorithm exists for treating this condition due to its rarity. However, treatment can be conservative with anti-hypertensive medication and diuretics. Surgical

intervention may range from percutaneous drainage and sclerotherapy, marsulpialization of collection to nephrectomy. [7,8]

Conclusion:

Renal lymphangiectasia is a rare disorder and radiological findings are diagnostic.

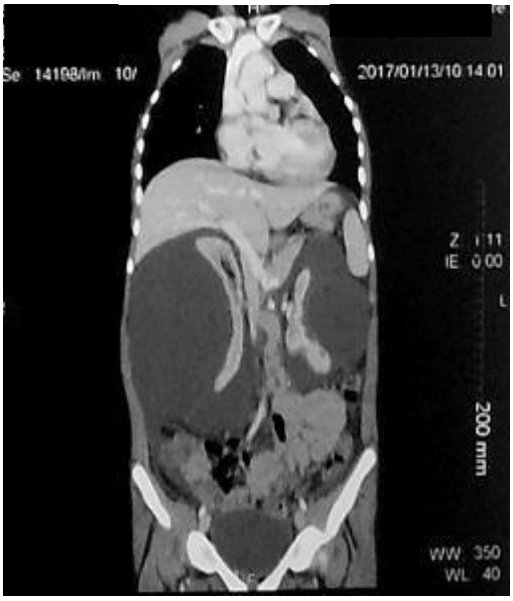

Figure 1: CT scan abdomen with IV contrast showing low attenuating, non-enhancing bilateral perinephric collections with a density of 1.5-3 HU suggestive of bilateral renal lymphangiectasia.

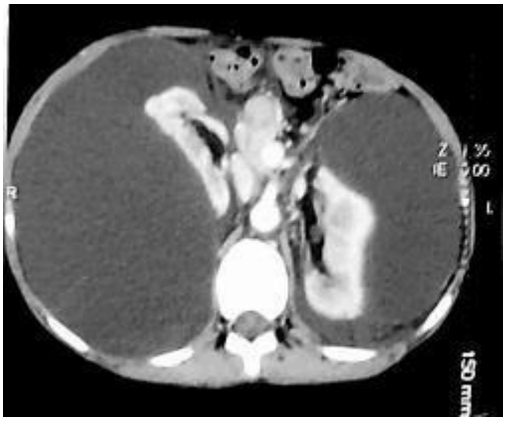

Figure 2: CT scan abdomen with IV contrast showing bilateral distortion of renal contours due to pressure effects.

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