Unilateral renal lymphangiectasia – the role of Ultrasound, CT and MRI. Case report

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ABSTRACT
Renal lymphangiectasia is a rare disorder characterized by ectasia of perirenal, peripelvic and intrarenal lymphatic vessels. A disturbance in the drainage of renal sinus lymphatics results in ectasia of peripelvic, intrarenal and perirenal lymphatics, and leads to the formation of renal lymphangiectasia. Multiple cysts with various sizes are seen in the perinephric space, renal parenchyma and in the renal sinus. The presence of fluid, density lesions in the retroperitoneum and around the great vessels, crossing the midline at the level of the origin of renal vessels are the diagnostic criteria. We report a case of a 27-year old female with multiple tiny cysts in the perinephric space and renal cortex, and a large cyst in the fat of the renal sinus, discovered with sonography, confirmed with CT Urography and MRI.

Keywords: renal lymphangiectasia, peripelvic cyst, renal lymphangioma, intrarenal and perinephric cyst.

CASE REPORT
A 27-year old female patient presented with vague pain in the right lumbar region for the past 2-3 months. There was no h/o fever/burning micturition/hematuria. No previous h/o Hypertension/Diabetes mellitus/Surgery/Renal calculus. Urine sample, renal function tests were within normal limits.

Ultrasonography of abdomen (Figure 1) showed multiple anechoic cystic lesions in various sizes (0.5-3.1 cm) in the fat of the renal sinus, not communicating with each other and with the pelvicalyceal system. Multiple small anechoic cystic lesions (3-5 mm) were seen in the renal cortex and perinephric space adjacent to the upper and lower pole and regions. The left kidney and the urinary bladder were normal. Ascites or retroperitoneal lymphadenopathy were not seen.

CT Urography was performed by obtaining arterial phase, corticomedullary phase, nephrographic phase and excretory phase after plain study (Figures 2-5). The size of the left kidney was 9.3x4.4 cm and appeared normal. The right kidney was 10.5x5.3 cm. CT showed a large cystic lesion, the size of approx. 3.7x3.2x4.5 cm in the fat of the right renal sinus, causing extrinsic compression on adjoining renal pelvis. The cystic lesion was extending to adjoining renal parenchyma alongside of infundibula and calyces in the upper, interpolar and lower pole calyces, which were effaced. Tiny hypodense cysts, the size of approx. 2-3 mm were noted in the renal cortex. Multiple small cysts, the size of 2-6 mm were noted in the perinephric space, more along its lower pole. No extensions of calyceal contrast were noted in these cystic lesions. The right kidney showed normal function. The right ureter appeared normal. These cystic lesions appeared to be displacing the intrarenal arteries.

MRI of the abdomen was performed for kidneys by obtaining T1WI, T2WI and T2 fat saturated sequences in axial, coronal and sagittal planes (Figures 6,7). Multiple cystic lesions were noted in the right kidney – in the peripelvic region, extending into renal parenchyma, involving cortex. These appeared hypointense on T1WI and hyperintense on T2WI. Multiple cystic lesions were also seen in the perinephric space. Cystic lesions were extending along the right upper ureter and in adjoining retroperitoneum posteriorly, right lateral and anterior aspects of IVC, extending to interaortocaval region. Thin hyperintense fluid
was also noted in the retroperitoneum, on right side, and in the right anterior pararenal space. MRI has an advantage of detecting the location and the content of cysts, extensions of the disease and associated involvement of retroperitoneal lymphatics and thoracic duct. The diagnosis of renal lymphangiectasia/lymphangioma/lymphangiomatosis was made. Histopathology report of USG Guided aspirated fluid showed lymphocytes, occasional eosinophils, macrophages, mesothelial cells and few degenerated cells in the background of proteinaceous fluid. No signs of malignancy were seen in the specimen.

Figure 1. USG of the Right Kidney in transverse (A), and longitudinal (B-D) showing multiple anechoic cystic lesions in the fat of the renal sinus, extending around the pelvicalyceal system, small cysts in the renal parenchyma and in the perinephric space (C).
Figure 2. CT of the ABDOMEN – Axial Plain CT (A), Axial corticomedullary phase (B-D) showing hypodense fluid cyst in the fat of the right renal sinus, extending to the renal parenchyma, small cortical cysts in the lower pole (C), small cysts in the perinephric space adjacent to the lower pole (C), hypodense fluid collection in the retroperitoneum extending anteriorly to the aorta and IVC (D).

Figure 3. CT of the ABDOMEN Axial arterial phase (A-D), showing hypodense fluid cyst in the fat of the right renal sinus, extending in the renal parenchyma, displacing the intrarenal arteries, small cortical cysts in the lower pole (C), small cysts in the perinephric space adjacent to the lower pole (C), hypodense fluid collection in the retroperitoneum, extending anteriorly to the aorta and IVC (D).
Figure 4. CT of the ABDOMEN Axial excretory phase (A-D), showing hypodense fluid cyst in the fat of the right renal sinus, extending in the renal parenchyma alongside of the infundibula and calyces in the upper, interpolar and lower pole calyces, which were effaced, small cortical cysts in the lower pole and in the perinephric space, adjacent to the lower pole (D).

Figure 5. CT of the ABDOMEN Coronal (A), Sagittal (B-D), excretory phase showing hypodense fluid cyst in the fat of the right renal sinus, extending in the renal parenchyma alongside of the infundibula and calyces in the upper, interpolar and lower pole calyces, which were effaced, small cortical cysts in the lower pole and in the perinephric space, adjacent to the lower pole.
Figure 6. MRI of the ABDOMEN Axial T2 fat sat. (A-F) showing cysts in the fat of the right renal sinus, extending in the renal parenchyma in the upper, interpolar and lower pole regions (A-C), small cortical cysts in the lower pole and in the perinephric space, adjacent to the lower pole (C-D), hyperintense fluid collection in adjoining retroperitoneum, extending anteriorly to the IVC and in the interaortocaval region (D-F), pole appearing hyperintense on T2WI.

Figure 7. MRI of the ABDOMEN – Coronal T2WI (A), Sagittal T2WI (B), Coronal T1WI (C) - showing cysts in the fat of the right renal sinus, extending to the renal parenchyma in the upper, interpolar and lower pole region, small cortical cysts in the lower pole and in the perinephric space, adjacent to the lower pole, appearing hyperintense on T2WI and hypointense on T1WI.
INTRODUCTION

Renal lymphangiectasia is characterized by the ectasia of perirenal, peripelvic and intrarenal lymphatic vessels. Renal lymphangiectasia is a more appropriate term than other terms used, as renal lymphangioma. It is usually bilateral, but can be unilateral and may be asymptomatic or present with flank pain, hematuria, abdominal distension, and proteinuria. Neck (75%) and axilla (20%) are common sites for lymphangioma. 5% of cases occur in the retroperitoneum, mediastinum, mesentery, omentum, pelvis and colon. Retroperitoneal lymphangiectasia accounts for 1% of all lymphangiectasias (1). Renal lymphangiectasia is rare. Unilateral lymphangiectasia is extremely rare with only 3 out of 21 reported cases of renal lymphangiectasia in a literature review, done in the last decade (2).

DISCUSSION

The lymphatics of the renal capsule and renal parenchyma drain into the renal sinus lymphatics, which empty into the paracaval, paraortic and interaortocaval lymph nodes. Though pathophysiology of renal lymphangiectasia is unclear, both congenital and acquired obstructive inflammatory processes are suggested, and etiological factors. A disturbance in the drainage of renal sinus lymphatics results in the ectasia of peripelvic, intrarenal and perirenal lymphatics leading to the formation of renal lymphangiectasia.

Hypertension is seen in about 50% of unilateral cases and 15% of bilateral cases, and usually occurs due to compressions, caused by parapelvic, intrarenal and perirenal cysts in the intrarenal arterial circulation, which leads to the renin dependent hypertension. The natural history of renal lymphangiectasia is unclear. It can appear suddenly, grow rapidly, squeeze or regress spontaneously (3).

It can exacerbate in pregnancy. The dilated lymphatic ducts can decompress the urinary tract leading to chyluria. Hemorrhages, ruptures, ascites and hypertension are the most common complications secondary to perirenal fluid collection. Venous thrombosis is a less reported complication (1). It is usually seen in children and less frequently occur in adults. Microscopically, they can be capillary or cavernous depending on size of the lymphatic spaces. Abnormal lymphatic channels can be unilocular or multilocular. Usually it occurs due to obstruction of the lymphatic ducts through the renal pedicle (4).

Renal lymphangiomatosis, renal lymphangioma, peripelvic lymphangiectasia, renal peripelvic multicystic lymphangiectasia are the other terms used for renal lymphangiectasia. Renal lymphangiectasia is a preferred name and has replaced the others, as there is the ectasia of perirenal, intrarenal and peripelvic lymphatics, renal polycystic disease, renal hygroma (5,6). Two different patterns of cystic lesions are observed in the renal sinus. The first pattern is called peripelvic - characterized by multiple small confluent cysts in the renal sinus, which are benign, usually bilateral and intraparenchymal. These usually occur due to lymphatic duct obstruction with resultant renal sinus lymphangiectasis. These mimic hydrenephrosis, but do not fill with excreted contrast and usually cause extrinsic compression and displacement of the collecting system. The second pattern is called parapelvic cyst and correspond to a large single cyst in the renal sinus, which originate from the medial renal parenchyma encroaching into the renal sinus. These appear like the renal cortical cyst. They may cause hydrenephrosis due to compression of the renal collecting system. In the absence of radiological and pathological data, the term cystic lesions of the renal sinus can be used. Multiple small cystic lesions are seen in the perinephric space surrounding the kidney, the view represents capsular lymphatic dilative perirenal lymphangiectasia. Sometimes perinephric fluid accumulation occurs in renal lymphangiectasia and is not surrounded by a wall. This occurs due to continuous secretion of fluid by the perirenal lymphatics, associated with altered retroperitoneal lymphatic pressure balance, that prevents reabsorption of the fluid. Multiple dilated tortuous cystic lesions are seen in the retroperitoneum, around the great vessels, and represents the ectasia of the lymphatic channels (6).

Renal lymphatic aspirate is not chylous or milky
like, the thoracic lymphatic duct as well as renal lymphatics are outside the pathway of mesenteric drainage. They contain lymphocytes and small amounts of fat and protein material. Imaging is based on Ultrasonography, the involved kidney may be normal or enlarged in size. Renal cortical echotexture may be normal or increased. Corticomedullary differentiation can be normal or lost. Multiseptated thin walled anechoic cystic lesions can be seen in the renal parenchyma, the fat of the renal sinus, in the peripelvic region and perinephric space. It can present as a solid mass, when small intrarenal lymphatics are obstructed. Retroperitoneal cystic lesions may be seen adjacent to the aorta and Inferior vena cava along with ascites. In the CT scan, they are seen as multiple well defined hypodense fluid lesions, the CT value (0-20HU), in the perinephric region. These can be unilocular or multilocular, high density fluid represents intracystic hemorrhage. Intervening renal parenchyma shows normal enhancement with normal excretion of contrast, mass effect can be seen in adjoining pelvicalyceal system.

The presence of fluid density lesions in adjoining retroperitoneum around the great vessels and crossing the midline at the level of origin of the renal vessels represents dilated renal lymphatics, draining into the larger retroperitoneal lymphatics, and is a typical sign of renal lymphangiectasia (7). Renal lymphangiectasia may show certain genetic mutations like Trisomy 7Q, monosomy X chromosome and mutations in VHL gene (Von-hippel Lindau gene) (8). On MRI, the cyst appears hypointense on T1WI and hyperintense on T2WI, and shows septations. These cysts can be seen in the perinephric, intrarenal and peripelvic regions. Extension of cysts can be seen in adjoining retroperitoneum. The signal intensity of cysts may vary, if there is high in protein content or hemorrhage within the cyst (3).

The treatment is not required in asymptomatic cases, especially those, which are detected incidentally. In symptomatic cases, percutaneous cyst aspiration is the first line treatment. However, high relapse rates are known in larger lesions, as they are multiseptated. Aspiration and sclerosis of the cystic lesions of the renal sinus is also useful. Sclerotherapy is contraindicated for peripelvic cysts due to the risks of leading to stenosis. Due to the leakage of sclerosing agent (6). Symptomatic management includes antihypertensives for arterial Hypertension and diuretics for ascites. In severe cases, laparoscopic ablation and nephrectomy can be performed.

Differential Diagnosis includes polycystic kidney disease, nephroblastomatosis, multilocular cystic nephroma, lymphoma and urinoma, polycystic renal disease (9).

CONCLUSION

Unilateral renal-lymphangiectasis is rare. It should be suspected if cystic lesions are seen in the perinephric space, within the renal parenchyma and in the renal sinus. The presence of fluid density lesions in adjoining retroperitoneum, around the great vessels and crossing the midline at the level of origin of the renal vessels represents dilated renal lymphatics draining into the larger retroperitoneal lymphatics, and is a typical sign of renal lymphangiectasia.
REFERENCES


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