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Case Report
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ABDOMINAL LYMPHANGIECTASIA

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ABSTRACT

We report a series of four cases of intra-abdominal lymphangiectasias diagnosed in our hospital using contrast enhanced CT scan of abdomen & pelvis. One patient had peri-renal lymphangiectasia, one retroperitoneal lymphangiectasia, one retroperitoneal & mesenteric lymphangiectasia and one patient had involvement of mesentery, retroperitoneum, extra-peritoneal tissues of pelvis and muscular & subcutaneous compartments of right thigh. We present the clinical details and the imaging findings, followed by discussion of the etiology, imaging and management options of this disorder.

KEYWORDS: Lymphangiectasia, peri-renal, retroperitoneal, CT.

INTRODUCTION

Lymphangiectasia is regarded as a developmental malformation of the lymphatic system characterized by generalized or localized dilatation of lymphatics leading to formation of thin walled cystic masses. Commonest locations of lymphangiectasias are neck & axillae (95% of cases) but they can occur anywhere in the body.[1] Lymphangiomatosis is a rare disease with multifocal lymphatic proliferation that has usually presents clinically during childhood and involves multiple parenchymal organs including the lung, liver, spleen, kidney, bone, and skin.

USG, CT scan & MRI are the commonly used imaging modalities to evaluate intra-abdominal lymphangiectasia. The diagnosis is confirmed by aspiration of chylous fluid.

CASE REPORTS

Case 1: A 45 years old male who was previously treated for filariasis presented with vague abdominal pain and with a normal USG report. He was referred for CT scan of abdomen and pelvis, which revealed non-enhancing lobulated hypodense tissue in the retroperitoneum completely encasing aorta & IVC (Figure 1A). This tissue was also encasing left renal vessels with significant narrowing of left renal vein (Figure 1B). However, left kidney showed normal nephrogram.

The diagnosis of retroperitoneal lymphangiectasia was made based on the above findings.

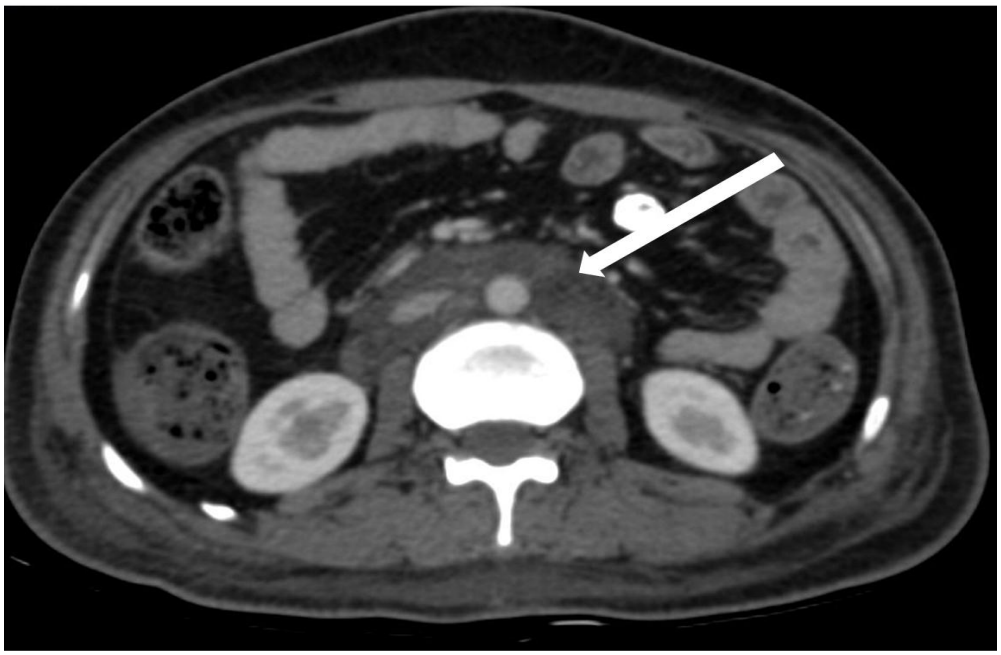

Figure 1A

Figure 1A: CT scan of abdomen in hepatic venous phase, axial section showing non-enhancing lobulated hypodense tissue in the retroperitoneum (white arrow) completely encasing aorta & IVC.

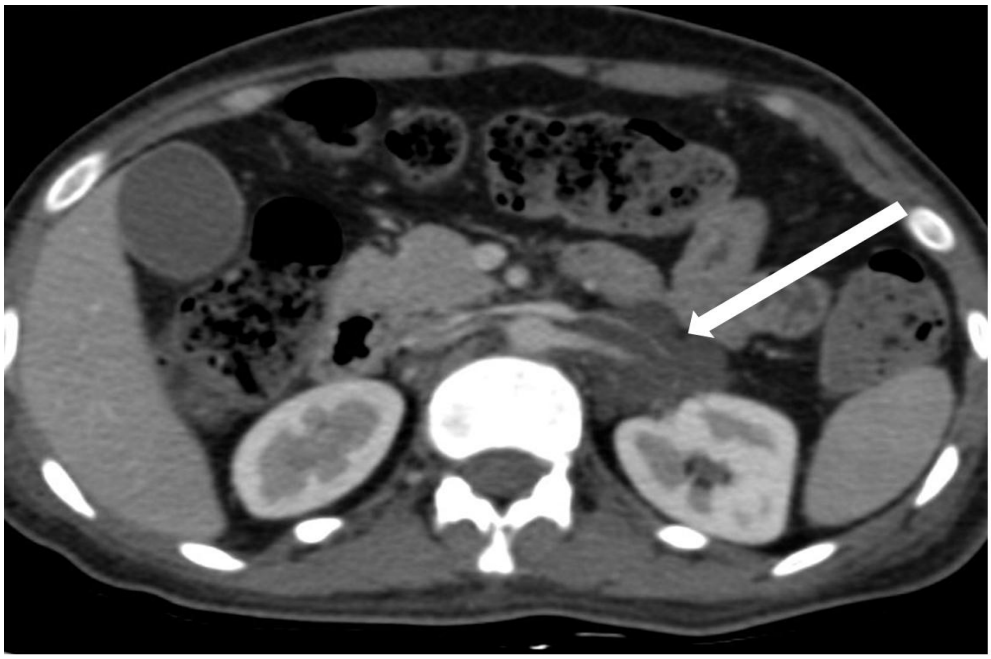

Figure 1B

Figure 1 B: CT scan of abdomen in hepatic venous phase, axial section showing non-enhancing lobulated hypodense tissue in the retroperitoneum (white arrow) encasing left renal vessels with significant narrowing of left renal vein However, left kidney showed normal nephrogram.

Case 2: A 2 years old male child presented with gradually progressive swelling of abdomen and right lower limb. USG revealed multiple ill-defined anechoic collections in muscles of right thigh and ascites. CT revealed multiple non-enhancing fluid density collections involving muscles of right thigh and in extraperitoneal tissues of pelvis (Figure 2A). There were also few ill-defined collections and mild stranding in subcutaneous tissues of right thigh (Figure 2B). CT also revealed diffuse ill-defined non-enhancing hypodense tissue in the mesentery & retroperitoneum and mild ascites (Figure 2B). Aspiration confirmed chylous nature of the collections.

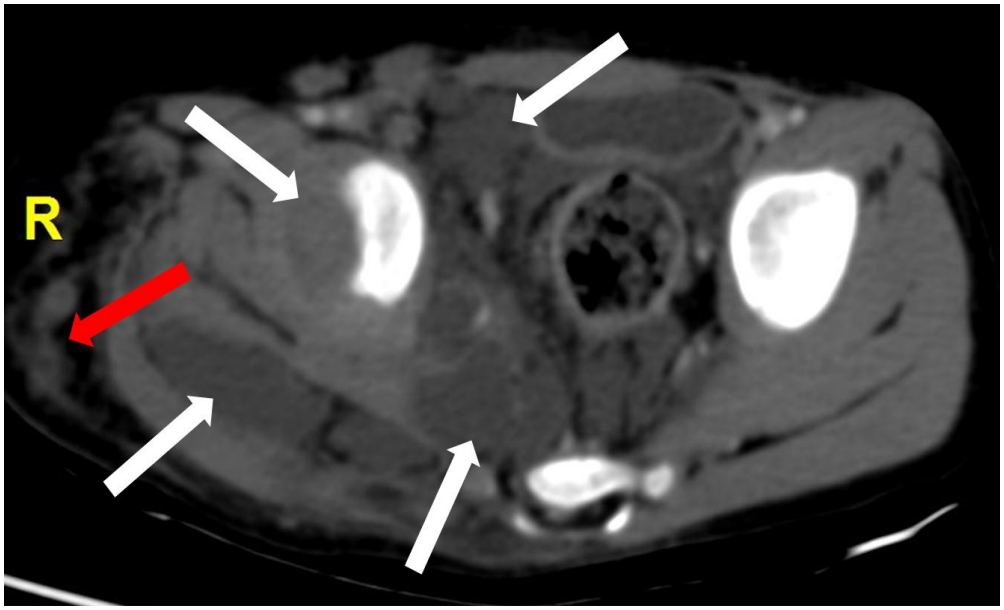

Figure 2A

Figure 2A: CT scan of abdomen & pelvis in hepatic venous phase, axial section showing multiple non-enhancing fluid density collections involving muscles of right thigh and in extraperitoneal tissues of pelvis (white arrows). There were also few ill-defined collections and mild stranding in subcutaneous tissues of right thigh (red arrow).

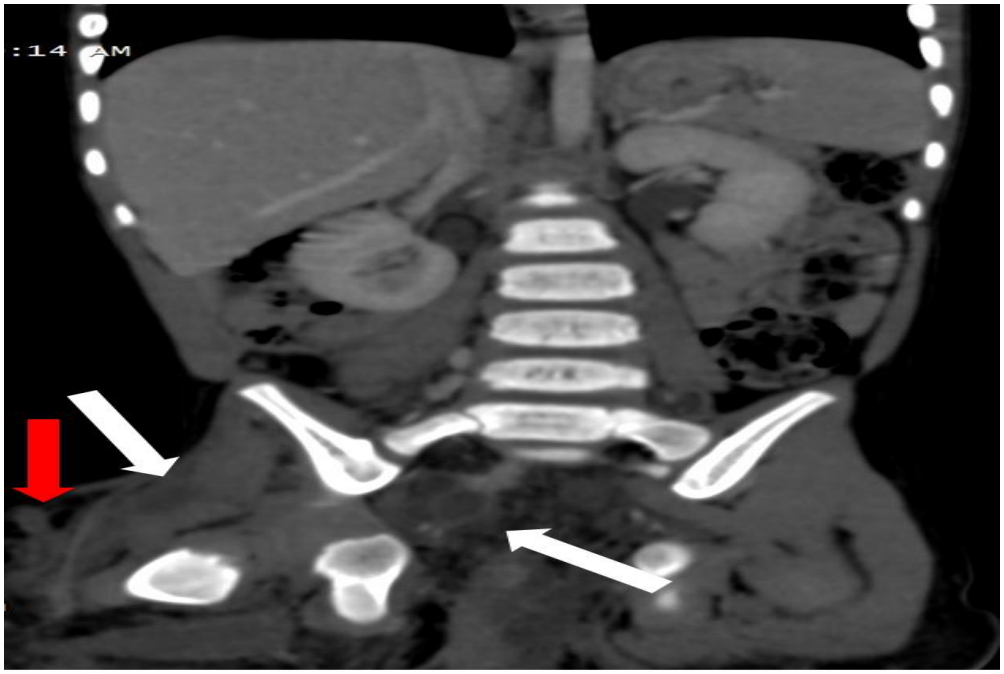

Figure 2B

Figure 2B: CT scan of abdomen & pelvis in hepatic venous phase, coronal reconstruction showing multiple non-enhancing fluid density collections involving muscles of right thigh and in extraperitoneal tissues of pelvis (white arrows) with a collection in the subcutaneous tissue (red arrow).

Case 3: A 25 years old male presented with left flank pain and occasional hematuria. USG revealed mild enlargement of left kidney and loss of cortico-medullary differentiation. There were loculated, septated collections in peripelvic and perinephric location with mild dilataion of renal pelvis. Liver, spleen & pancreas were normal.

CT revealed loculated, fluid density collections in the peripelvic & perinephric space with suboptimal nephrogram of left kidney (Figures 3A & 3B).

Aspiration revealed chylous fluid confirmatory for renal lymphangiectasia.

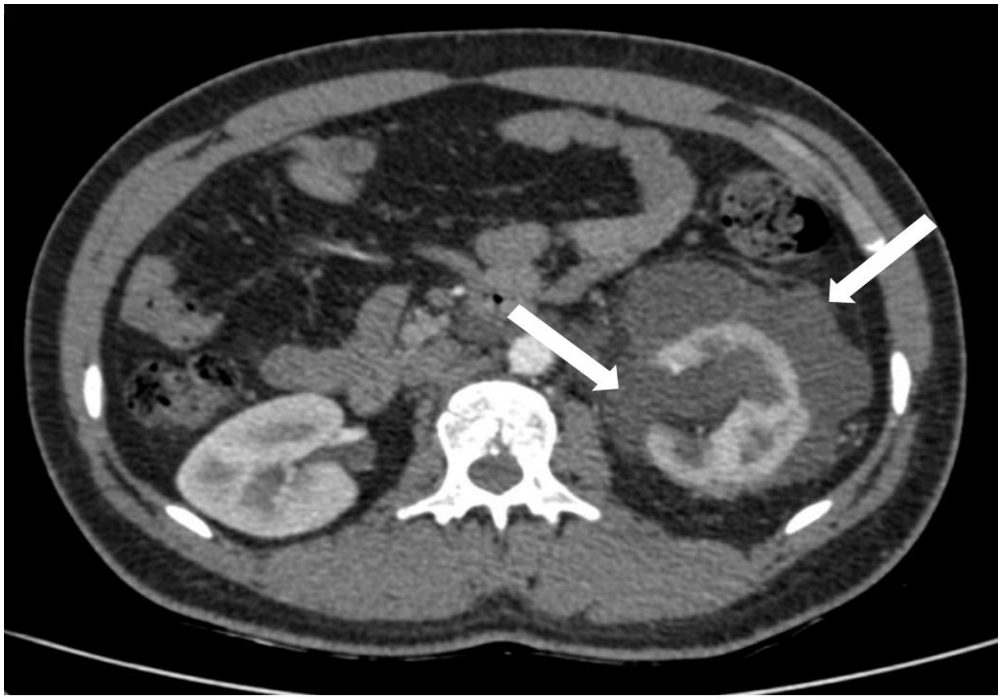

Figure 3A

Figure 3A: CT scan of abdomen & pelvis in nephrographic phase, axial section showing loculated, fluid density collections in the peripelvic & perinephric space (white arrows) with suboptimal nephrogram of left kidney.

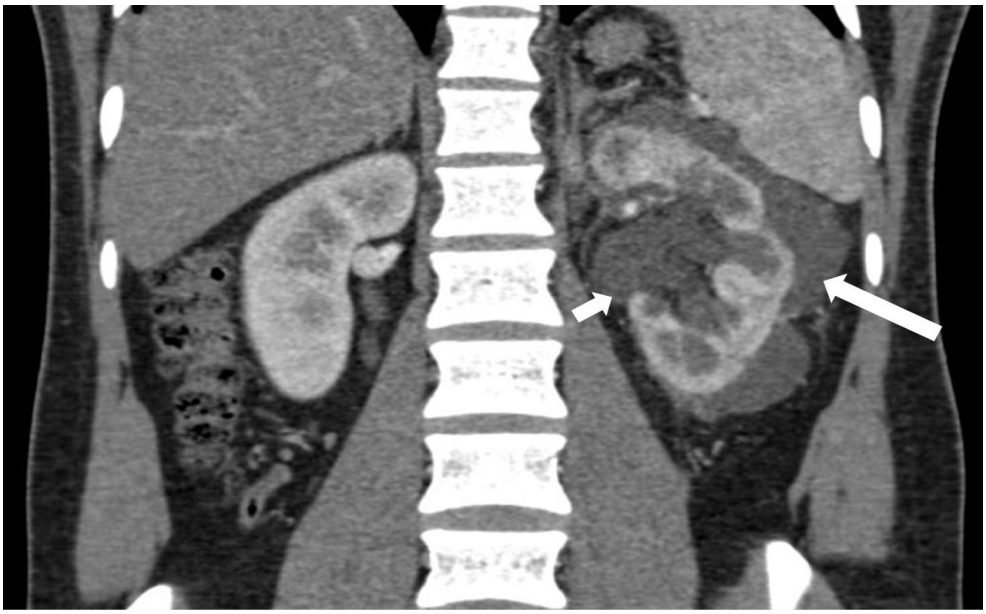

Figure 3B

Figure 3B: CT scan of abdomen & pelvis in nephrographic phase, coronal reconstruction showing loculated, fluid density collections in the peripelvic & perinephric space (white arrows) with suboptimal nephrogram of left kidney.

Case 4: A 15 years old boy presented with vague abdominal pain. USG revealed multiple ill-defined loculated anechoic intra-abdominal collections. CT revealed diffuse mesenteric haziness with diffuse ill-defined non-enhancing hypodense tissue in the mesentery & retroperitoneum and mild ascites (Figures 4A & 4B).

Aspiration confirmed chylous fluid.

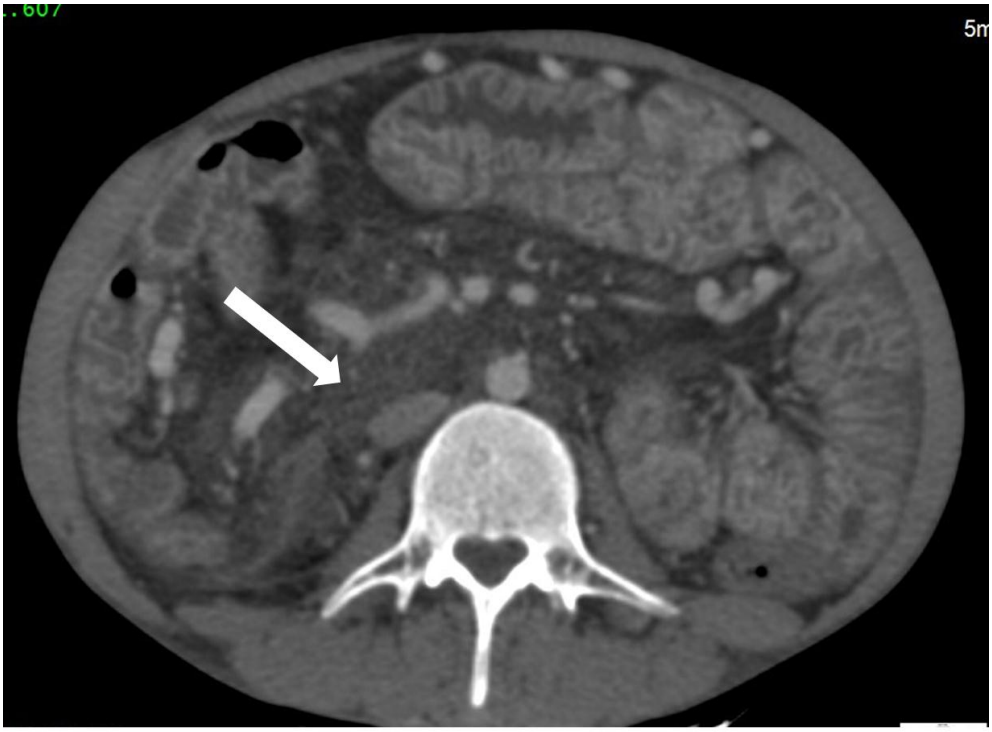

Figure 4A

Figure 4A: CT scan of abdomen & pelvis in hepatic venous phase, axial section showing diffuse mesenteric haziness with diffuse ill-defined non-enhancing hypodense tissue in the mesentery & retroperitoneum (white arrow).

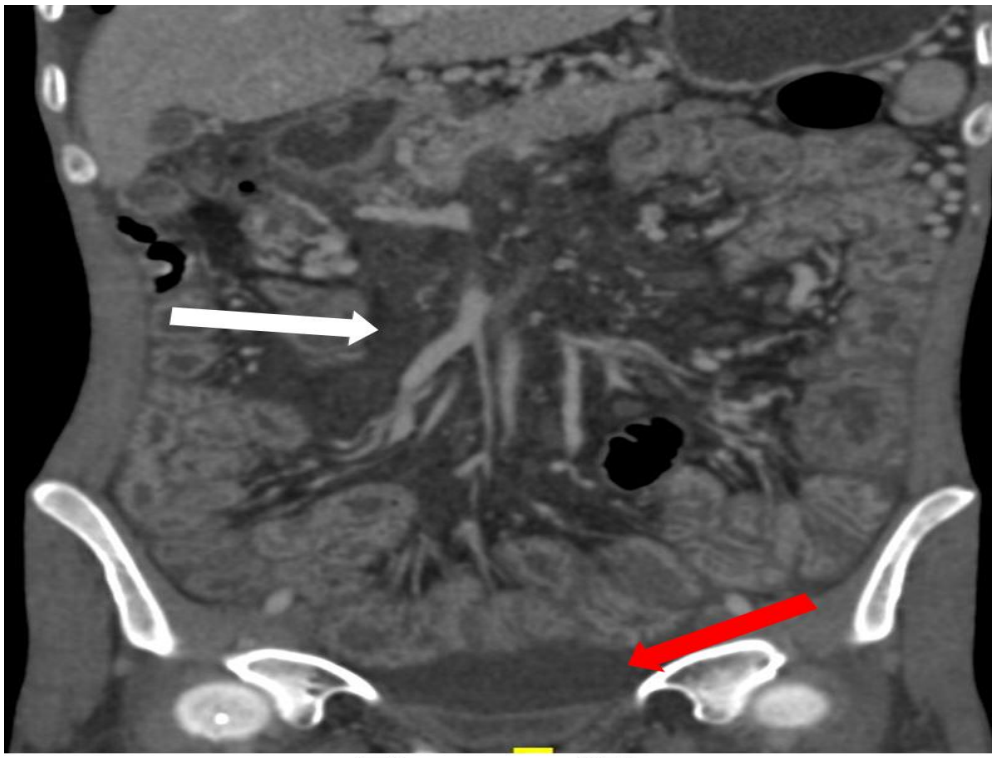

Figure 4B

Figure 4B: CT scan of abdomen & pelvis in hepatic venous phase, coronal reconstruction showing diffuse mesenteric haziness with diffuse ill-defined non-enhancing hypodense tissue in the mesentery & retroperitoneum (white arrow) and mild ascites (red arrow).

DISCUSSION

Although most cases of abdominal lymphangiectasia are considered developmental abnormalities of the lymphatic system, acquired cases may result secondary to obstruction of lymphatics following infections like filariasis, lymphatic metastasis or following surgery or radiotherapy.[2]

Abdominal lymphangiomas most commonly occur in the mesentery, followed by the omentum, mesocolon, and retroperitoneum. It commonly presents as multilocular cystic masses. The cysts may contain chylous, serous, hemorrhagic, or mixed fluid. Lymphangiomas have an insinuating nature hence complete surgical excision is difficult in some cases.

On USG, lymphangiomas may appear anechoic or may contain echogenic debris.

On CT, the fluid component is typically homogeneous and has low attenuation values. Occasionally, it may have negative attenuation values due to presence of chyle. Calcification may occur but is uncommon.[3] Cyst wall & septae may show enhancement.

Uncomplicated lymphangiomas are T1 hypointense and T2 hyperintense on MRI, similar to fluid. Haemorrhage into a cyst or presence of infection may alter the signal characteristics.[3]

Most important point differentiating large mesenteric lymphangiomas from ascites is lack of fluid in the dependent recesses of the peritoneum such as the paracolic gutters, subhepatic spaces and between the folds of mesentery.[1] It is often difficult to differentiate lymphangiomas from enteric duplications, mesothelial cysts, and pseudocysts as the imaging features of these lesions overlap.

Complications like intestinal obstruction or volvulus, and infarction may occur with mesenteric lymphangiomas.

The commonest abdominal organ affected is the kidney with dilatation of the lymphatics located in the kidneys and the retroperitoneal tissue. It has also been reported as "renal hygroma"[4], "renal lymphangioma" and "renal peripelvic multicystic lymphangiectasia"[2], the preferred name being "Renal lymphangiectasia". As there is failure of renal lymphatic ducts in draining into larger retroperitoneal lymphatics, there is dilatation of perirenal, peripelvic, and intrarenal ducts and formation of unilocular or multilocular cystic spaces in the pelvic sinus as well as perinephric spaces.

The common minor variant, renal peripelvic multicystic lymphangiectasia or polycystic disease of the renal sinus manifests as peripelvic cysts. These flat, fan like cysts which do not communicate with each other are easily diagnosed by ultrasonography.[4]

The major variant, which affects the entire kidney and retroperitoneal tissue, is an uncommon manifestation that is often associated with significant enlargement of the kidneys. Renal Lymphangiectasia occurs as loculated, cystic, septated collection in peripelvic and/or perinephric region causing posterior acoustic enhancement. There is distortion of the pelvicalyceal system (in intrapelvic collection), distortion of renal contour (in peripelvic collection)[5] in addition to thickening of perirenal fasciae.

Renal Lymphangiectasia is a rare disorder and may present with hematuria, abdominal pain and flank pain. Children may present with only bilateral nephromegaly and sonographically echogenic kidneys.[6]

On ultrasonography, bilateral nephromegaly is a common finding. Hydronephrosis, as evident in the form of dilated structures in both renal sinuses may also be found without any calculi or distal ureteric obstruction.[6] The most important differential diagnosis on ultrasonography is adult polycystic kidneys and hydronephrosis. The cysts in ADPKD vary in size and are scattered non-uniformly throughout renal parenchyma whereas the cysts in retroperitoneal lymphangiectasia are perinephric with normal renal parenchyma.[7] In multilocular cystic nephroma, adult polycystic kidney disease or von Hippel-Lindau disease, cystic lesions may additionally be seen in liver and pancreas.[8]

Before the advent of CT scan, the patients were usually diagnosed at the time of surgery i.e. exploratory laparotomy or after nephrectomy. However, currently the diagnosis is mainly based on CT scan findings. On CT scan, renal lymphangiectasia appears as a well contained, fluid attenuating collections(0-10 HU) in the peripelvic or perinephric region with or without demonstrable septations with normal renal parenchyma and normal pelvicalyceal system.[5] The diagnosis of renal lymphangiectasia can be confirmed with aspiration of chylous fluid from the perinephric fluid collections which reveals increased renin due to its renal origin.

Magnetic resonance imaging can also diagnose RLM, as the lymphatic collections appear hyperintense on T2 weighted half-Fourier acquisition single-shot turbo spin-echo images and there is reversal of corticomedullary intensity.[5]

Renal Lymphangiectasia has been associated with hypertension, renal vein thrombosis with /without collateral formation. Lymphangiectasia can become quite extensive and ultimately lead to 'decompensated lymphangiectasia' with massive lymph collections in pre-existing cavities. Other complications include hematuria, proteinuria and hemorrhage into lymphatic cyst. Renal and retroperitoneal lymphangiectasia may cause renin-dependent hypertension which is secondary to mechanical compression and resultant reduced

perfusion of the kidney (Page's kidney).[4] The natural history of this disorder is not completely understood. Pregnancy is associated with exacerbation in the symptoms and complications whereas in a neonatal case, partial regression has been reported.[2,9]

Treatment is usually not necessary in asymptomatic cases. Percutaneous drainage is indicated in symptomatic and large collections causing pressure symptoms and other complications. Recurrent collections need marsupialization whereas nephrectomy is reserved for collections with uncontrollable complications or recurring, complicated collections.

CONCLUSION

Retroperitoneal lymphangiectasias are rare disorders which can be asymptomatic or can cause significant symptoms. A differential should be kept in mind while interpreting dilated pelvicalyceal system without demonstration of a calculus, or while evaluating peripelvic cysts. Management should be tailored to the severity of the patient's symptoms. Complications, though rare, imply the need for active treatment.

REFERENCES

1. Lugo-Olivieri CH, Taylor GA. CT differentiation of large abdominal lymphangioma from ascites. Pediatric radiology., 1993; 23(2): 129-30. PubMed PMID: 8516037.
2. Ramseyer LT. Case 34: renal lymphangiectasia. Radiology., 2001 May; 219(2): 442-4. PubMed PMID: 11323470.
3. Ko S-F, Ng S-H, Shieh C-S, Lin J-W, Huang C-C, Lee T-Y. Mesenteric cystic lymphangioma with myxoid degeneration: unusual CT and MR manifestations. Pediatric radiology., 1995; 25(7): 525-7.
4. Riehl J, Schmitt H, Schäfer L, Schneider B, Sieberth H. Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis. Nephrology Dialysis Transplantation., 1997; 12(8): 1701-3.
5. Rastogi R, Rastogi V. Computed tomographic scan in the diagnosis of bilateral renal lymphangiectasia. Saudi Journal of Kidney Diseases and Transplantation., 2008; 19(6): 976.
6. Sarikaya B, Akturk Y, Bekar U, Topaloglu S. Bilateral renal lymphangiomatosis mimicking hydronephrosis: multidetector CT urographic findings. Abdominal imaging., 2006; 31(6): 732-4.
7. Rajeev T, Barua S, Deka P, Hazarika S. Bilateral peri-renal lymphangiomatosis. Indian Journal of Urology., 2006; 22(1): 73.
8. Rastogi R, Rastogi U, Sarikwal A, Rastogi V. Renal lymphangiectasia associated with chronic myeloid leukemia. Saudi Journal of Kidney Diseases and Transplantation., 2010; 21(4): 724.
9. Meredith WT, Levine E, Ahlstrom N, Grantham J. Exacerbation of familial renal lymphangiomatosis during pregnancy. American Journal of Roentgenology., 1988; 151(5): 965-6.