Case Report

Bilateral renal lymphangiectasia: A case report

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ABSTRACT

Renal lymphangiectasia is a rare entity occurring both in children and adults with no specific age preference. It may be unilateral or bilateral and has no gender predilection. The clinical symptoms are non-specific ranging from no complaints with incidental diagnosis to pain abdomen. Radiological appearance may be mistaken with peripelvic cysts, renal cysts, and perinephric abscess, or collection. Hence, here lies the importance of familiarity with imaging appearance of this rare condition. We describe the case of a 35-year-old male who presented with pain abdomen. The radiological investigations revealed bilateral perinephric collections which led to the diagnosis of bilateral renal lymphangiectasia by exclusion, which was confirmed by aspiration of chylous fluid.

Key words: Chylous, Lymphangiectasia, Parapelvic, Perinephric

Renal lymphangiectasia is a rare entity involving the kidneys; although, the exact prevalence is not mentioned in the previous literature. It accounts for approximately 1% of all lymphangiomas [1]. Ectatic perirenal, parapelvic, and intrarenal lymphatic vessels are seen in this condition [2]. A radiologist must know about its imaging characteristics, radiological presentations, and complications while differentiating it from other possibilities. This rare condition can be confused with hydronephrosis, polycystic kidney disease, multilocular cystic nephroma, urinoma, and abscess [3]. Proper understanding of the imaging appearance of this condition is vital to rule out other conditions with the help of proper clinical history.

CASE REPORT

A 35-year-old male presented with a history of vague pain abdomen for 3 months. The pain was a dull-aching type, remained throughout the day, and relieved by supination. There was no history of abdominal trauma or abdominal surgery.

On examination, his blood pressure was 130/80 mmHg, pulse rate was 80/min, respiratory rate was 18/min, and body temperature was 96^{0}F. Urine analysis was normal, hemoglobin was 13 mg/dl, urea was 22 mg/dl, and creatinine was 1.1 mg/dl. All other blood investigations were within normal limits.

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Ultrasound (USG) abdomen revealed enlarged echogenic kidneys measuring up to 14 cm on the right side and 15 cm on the left side. There was the presence of large perinephric collections on both sides (left >right). No hydronephrosis or calculus was seen. Triple phasic Contrast-enhanced computed tomography (CT) and kidney urinary bladder showed fluid collections in the perinephric space bilaterally, surrounding the kidney cortex (left >right) (Fig. 1a). This fluid collection had a CT value of 0-10HU and also few septa were seen within it (Fig. 1b). There was no evidence of peripelvic cysts. Both the kidneys excreted the contrast normally (Fig. 1c). On imaging, the diagnosis of bilateral renal lymphangiectasia was made.

The USG-guided diagnostic aspiration of the perinephric collection was performed on the left side which yielded straw-colored fluid (Fig. 2). Biochemical analysis revealed a high protein (200 mg/dl) and triglyceride level (11 mg/dl) and a high lymphocyte count (70%) suggestive of chylous aspirate, thus confirming our diagnosis. The patient was managed conservatively with analgesics for symptomatic treatment and relieved and discharged.

DISCUSSION

Retroperitoneal lymphangiectasia is a rare condition accounting for ~1% of all lymphangiectasia [4]. The exact prevalence is not reported in the literature due to the rarity of the disease. The knowledge of this condition is mostly based on solitary case reports.

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Approximately, 40 cases have been reported since 1890 [5]. The other terms used for this entity are renal lymphangiomatosis, renal lymphangioma, renal peripelvic multicystic lymphangiectasia, peripelvic lymphangiectasia, hygroma renale, and polycystic disease of the renal sinus [6]. Some cases have shown familial association [7]. These benign tumors are often seen in children, involving the neck (75%), and axillary (20%) region, but rarely the kidney [8].

The pathogenesis of the disease is due to the disturbance in the development of normal communication between the lymphatic tissue and the rest of the lymphatic system leading to the lack of normal drainage of lymphatic fluid with subsequent cystic dilatation of the lymphatic channels [9]. It is almost always bilateral. Clinically, it is usually asymptomatic and incidentally diagnosed. If symptomatic, the common presentations include flank pain, abdominal distension, hematuria, and proteinuria.

There is little information about the natural history of this disease in the medical literature. The lymphatic drainage of the kidneys, the renal capsule, and the perirenal tissues is interconnected through several large lymphatic trunks located inside the renal sinus. These lymphatic trunks drain into the para-aortic, para-caval, and interaortocaval lymph nodes. There may be a developmental malformation and derangement of the drainage of these lymphatic trunks, leading to their dilatation and the creation of cystic voids adjacent to the renal sinus and in some cases, in the perinephric space [9].

The imaging characteristics depend on the site and extent of the lymphatic involvement. The radiological findings correspond to the site of involvement and include a variety of presentations that comprise peripelvic cysts and perirenal fluid collections, retroperitoneal fluid collections, solid renal lesions, and slight diffusely enlarged kidneys without cystic space. These forms are being explained in Table 1. Imaging plays an important role in differentiating renal lymphangiestasia from other perirenal kidney diseases [2].

Imaging studies give a striking clue to the diagnosis in asymptomatic cases also [1]. Excretory urography in children and adults with renal lymphangiomatosis shows bilateral nephromegaly with caliceal distortion caused by multiple large parapelvic cysts [10]. Ultrasonography reveals normal-sized or enlarged kidneys with or without increased parenchymal echotexture and loss of corticomedullary differentiation. Multiseptated thin-walled fluid collections are seen in perinephric or peri-pelvic regions. On CT-scan, the appearance of well-defined fluid attenuation multiseptated collections in perinephric or peri-pelvic regions with normal renal parenchymal enhancement is seen. The presence of fluid or fluid-filled spaces in the retro-peritoneum in close vicinity of great vessels, as well as, crossing the midline at the level of renal vessels is a diagnostic sign of renal lymphangiectasia.

The diagnosis of renal lymphangiectasia can be confirmed with needle aspiration of chylous fluid from the perinephric fluid collection. Renal lymphatic aspirates may come as a surprise because the fluid is not milky or "chylous" like that of the thoracic lymphatic duct. This is due to the fact that renal lymphatic ducts are outside the mesenteric drainage pathway. Renal lymphatics contain only "sporadic" cells (mostly lymphocytes) and small amounts of fat and protein material [7,9].

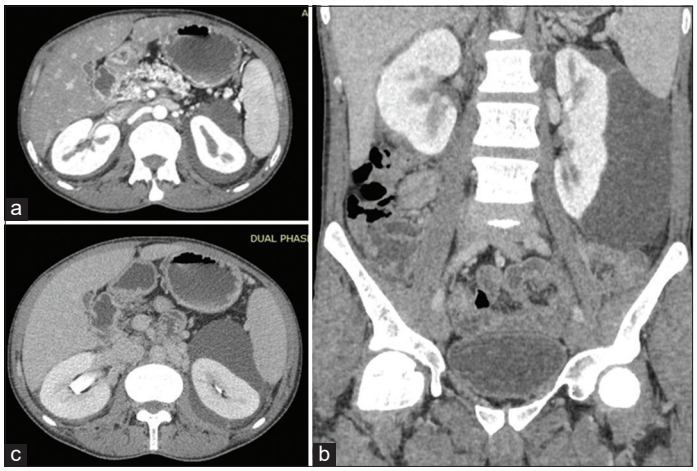

Figure 1: (a) Bilateral perirenal collections with Computed Tomographic value of 0–10 Hounsfield's Unit-corticomedullary phase in axial reformat; (b) nephrographic phase-septum is seen at upper pole on left side with bilateral perirenal collections in coronal section; (c) Excretory phase-bilateral kidneys showing normal excretion in axial image

Figure 2: Aspirated fluid

Table 1: Imaging characteristics of the disease

Peripelvic cysts	• Lymphatics distributed in the renal sinus are involved • Usually a cystic dilatation of simple characteristics within and clearly defined walls • Some show evidence of thin septations
Perirenal fluid collections	• Altered retroperitoneal lymphatic pressure balance that prevents the fluid from being appropriately reabsorbed • Continuous generation of fluid by the peri-renal lymphatic system • Capsular lymphatic dilatation • Lobular perinephric accumulations with multiple septations and fluid attenuation "enveloping" or surrounding the kidney
Retroperitoneal fluid collections	• Presumably dilated lymphatic vessels • Rare entity • In some cases, multiple tortuous structures form a linear pattern in the retroperitoneum seen distributed around the great vessels suggestive of lymph channel ectasia also known as cisterna chyli

Several differential diagnostic possibilities are to be considered for each presentation. In case of bilateral nephromegaly with pelvicalyceal splaying and distortion, adult polycystic kidney disease, lymphoma, nephroblastomatosis, and other causes of multiple renal masses, such as von Hippel-Lindau disease and tuberous sclerosis are the differential diagnosis considerations [6]. While perinephric fluid collections may mimic perinephric hematoma, urinoma, perinephric abscess, multilocular cystic nephroma if not correlated clinically. Complications of the disease include hemorrhage, rupture, ascites, and hypertension secondary to perirenal fluid collection. Renal vein thrombosis is a less commonly reported complication [4].

Treatment options for renal lymphangiectasia vary according to the symptoms, complications, and site of involvement. Treatment includes percutaneous drainage, marsupialization, nephrostomy, or sclerosing therapy for symptomatic patients with large collections. Nephrectomy can be opted for severe cases [3].

CONCLUSION

Bilateral renal lymphangiectasis is one of the rare entities of renal lymphatics that we may come across while routine imaging. This can be confused with a variety of other kidney conditions. Imaging plays a key role in differentiating this entity from such conditions, thus leading to proper management.

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