



RENAL LYMPHANGIECTASIA. MDCT AND MRI FINDINGS

LINFANGIECTASIA RENAL. HALLAZGOS POR TCMD Y RM

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KEY WORDS (MeSH)

Tomography
 Magnetic resonance
 imaging
 Lymphangiectasis
 Kidney

PALABRAS CLAVE (DeCS)

Tomografía
 Imagen por resonancia
 magnética
 Linfangiectasia
 Riñón

SUMMARY

Renal lymphangiectasia is a rare entity of the renal lymphatics that occurs in both children and adults. It may be unilateral or bilateral and has no gender predilection. It has been known by other names such as renal lymphangiomatosis, renal lymphangioma, parapelvic lymphangiectasia, polycystic renal sinus, and renal hygroma, among others. Being a rare entity, radiologists usually mistake it for peripelvic cysts, renal cysts or hydronephrosis, hence the importance of becoming familiar with its imaging features.

RESUMEN

La linfangiectasia renal es una entidad rara de los linfáticos renales que se presenta tanto en niños como en adultos, puede ser unilateral o bilateral y no tiene predilección por sexo. Se ha conocido con otros nombres, como *linfangiomatosis renal*, *linfangioma renal*, *linfangiectasia peripelvica*, *enfermedad poliquistica del seno renal* e *higroma renal*. Esta entidad es poco frecuente, por lo cual es común que sea erróneamente interpretada en las imágenes como quistes parapiélicos, quistes renales o hidronefrosis. Por lo tanto es importante que el radiólogo esté familiarizado con sus características imaginológicas.

Introduction

Lymphangiectasia is a condition characterized by different degrees of dilatation of the lymphatic ducts. It is caused by an abnormal development of the lymphatic structures, with single or multilocular fluid-filled cavities, usually found in the neck (75%-80%) and axillary (20%) regions. Lymphangiectasia develops in rare occasions in the kidney, where it is believed to be due to an abnormal communication between the renal lymphatics and the larger retroperitoneal lymphatics. In this case, it has been known by other names like *renal lymphangiomatosis*, *renal lymphangioma*, *peripelvic lymphangiectasia*, *renal sinus polycystic disease*, and *renal hygroma*, among others (1).

Renal lymphangiectasia is a benign condition that has been described in children as well as in

adults. However, although its clinical characteristics and radiological findings are well defined, its pathophysiology is still unknown. The course of the disease varies, and several treatment options have been proposed, ranging from symptomatic management and percutaneous drainage to laparoscopic drainage and nephrectomy in the most severe cases.

Pathophysiology

The pathophysiology of this disease has not been elucidated to date. 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 inter-aorto-caval lymph nodes. There may be a developmental malformation and

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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 (2).

Clinical findings

Renal lymphangiectasia usually occurs without symptoms and is an incidental finding on imaging studies. When it is symptomatic, it is usually associated with abdominal pain (42%), bloating (21%), fatigue, weight loss and hematuria. In the most severe cases, blood hypertension, ascitis and renal function decline have been described (1,3-5). In children, it may also manifest with a palpable abdominal mass, kidney enlargement or pyelonephritis (4,6,7).

Cystic lesion patterns in the renal sinus

There are two different patterns of cystic lesions in the renal sinus. The first pattern manifests with multiple, confluent small cysts originating in the renal sinus that are benign, intraparenchymal lesions, usually bilateral. These cysts are called *peripelvic*, and lymphatic duct obstruction has been proposed as the main mechanism of origin, giving rise to renal sinus lymphangiectasia (3,8). The main differential diagnosis is hydronephrosis; however, in contrast studies, these cystic dilatations, unlike hydronephrosis, do not fill with the excreted contrast medium and often displace the collecting system as a result of extrinsic compression (Figure 1).



Figure 1. Bilateral renal lymphangiectasia. Urotomogram in excretory phase; axial sections (a and b) and coronal PMRI (c), showing bilateral cystic lesions of the renal sinus. This causes extrinsic compression and displacement of the adjacent collecting cavities. There is minimal dilatation of the proximal caliceal system.

The other renal sinus cystic pattern corresponds to large single cysts inside the sinus that originate in the medial renal parenchymal tissue from where they encroach into the renal sinus (Figure 2). These are called *parapelvic cysts* (8). On imaging, they have the same appearance as renal cortical cysts. In general, they are single or small in number, although sometimes, because of their size, they may be associated with greater symp-

tomatology and the development of hydronephrosis secondary to compression of the renal vessels and of the adjacent collecting system structures (8) (Figure 3). In the absence of accurate radiological and pathological correlation data, it is recommended that the generic term used to describe these entities be *cystic lesion of the renal sinus* (8).

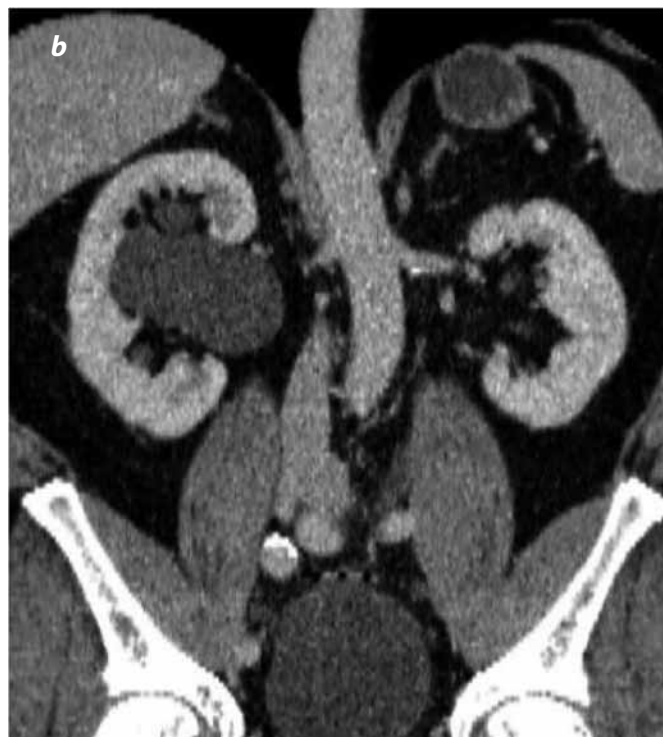


Figure 2. Parapelvic cyst. Axial view of excretory-phase urotomogram (a) and coronal nephrographic phase view (b), showing evidence of a cortical cyst originating in the middle third of the right kidney and extending into the renal sinus, thus causing a non-obstructive displacement of the adjacent collecting cavities.

Imaging characteristics of renal lymphangiectasia

The imaging characteristics depend on the site and extent of the lymphatic involvement. If only the *small intra-renal lymphatics* are compromised, the lesion may appear as a solid renal lesion or with slight diffuse kidney enlargement with no cystic space (9) (Figure 5).

When the *lymphatics distributed in the renal sinus* are involved, there is usually a cystic dilatation of simple characteristics, such as thin and clearly defined walls. Some show evidence of thin septations that are better characterized with ultrasound or T2 magnetic resonance (MR) sequences, and less reliably with computed tomography (CT) (Figures 4 and 5). Enhancement of these septations may also be seen in T1 uroresonance after gadolinium administration (Figure 5). Larger cystic dilatations may create obstructions of the collecting system, with proximal dilatation in some cases (Figures 1 and 5) (1,10).

A less frequent finding is the appearance of lobular perinephric accumulations with multiple septations and fluid attenuation “enveloping” or surrounding the kidney and slightly apparent

inside the renal sinus. This represents *capsular lymphatic* dilatation, consistent with peri-renal lymphangiectasia (2,3,11).

More recently, a perinephric fluid accumulation has been described in a patient with established renal lymphangiectasia. The collection is not surrounded by a wall and does not appear like a cystic lesion, as is usually the case in renal lymphangiectasia. This finding has been explained by the continuous generation of this fluid by the peri-renal lymphatic system, associated with altered retroperitoneal lymphatic pressure balance that prevents the fluid from being appropriately reabsorbed. In severe cases, this may result in failed percutaneous or surgical drainage that may even facilitate the formation of larger collections, leading eventually to the need for nephrectomy (12,13).

In some cases, multiple tortuous structures that form a linear pattern in the retroperitoneum may also be seen distributed around the great vessels. They suggest the presence of lymph channel ectasis (2). Consequently, it may be stated that the distribution of renal lymphangiectasia varies according to the location of the lymphatics that are predominantly affected. The dilation of the lymphatic system may be intra-renal, peri-renal,



Figure 3. Parapelvic cyst. Coronal view of an excretory-phase urotomogram (a) and coronal MIP reconstruction (b) showing a parapelvic cyst originating in the middle lower third of the left kidney and encroaching on the renal sinus. This reduces the diameter of the collecting system and creates a proximal dilatation.

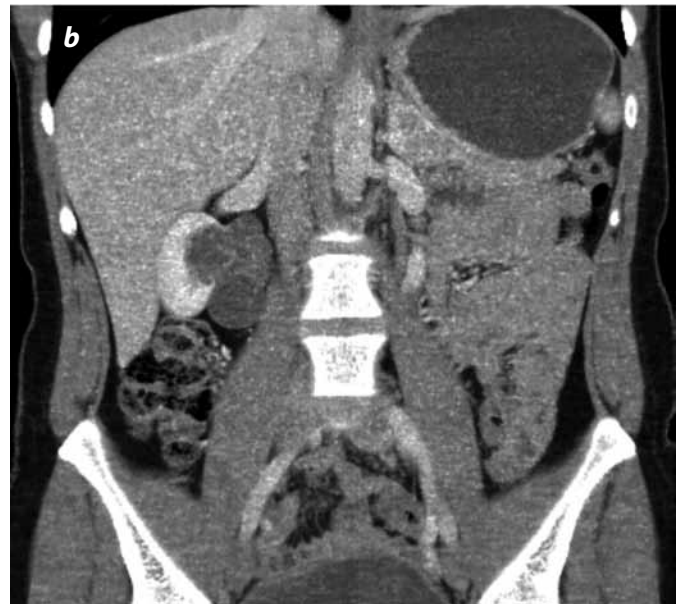
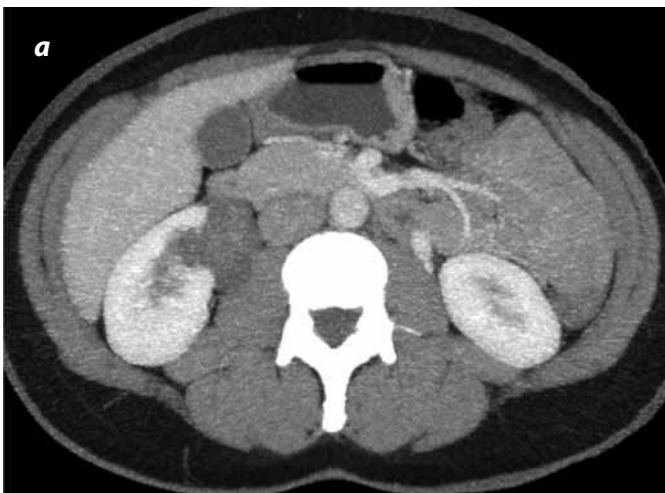


Figure 4. Right renal lymphangiectasia. Axial (a) and coronal (b) MIP views in the nephrographic phase, showing lymphangiectasia of the right renal sinus. There is suggestion of thin septations that are better seen in the coronal view (b).

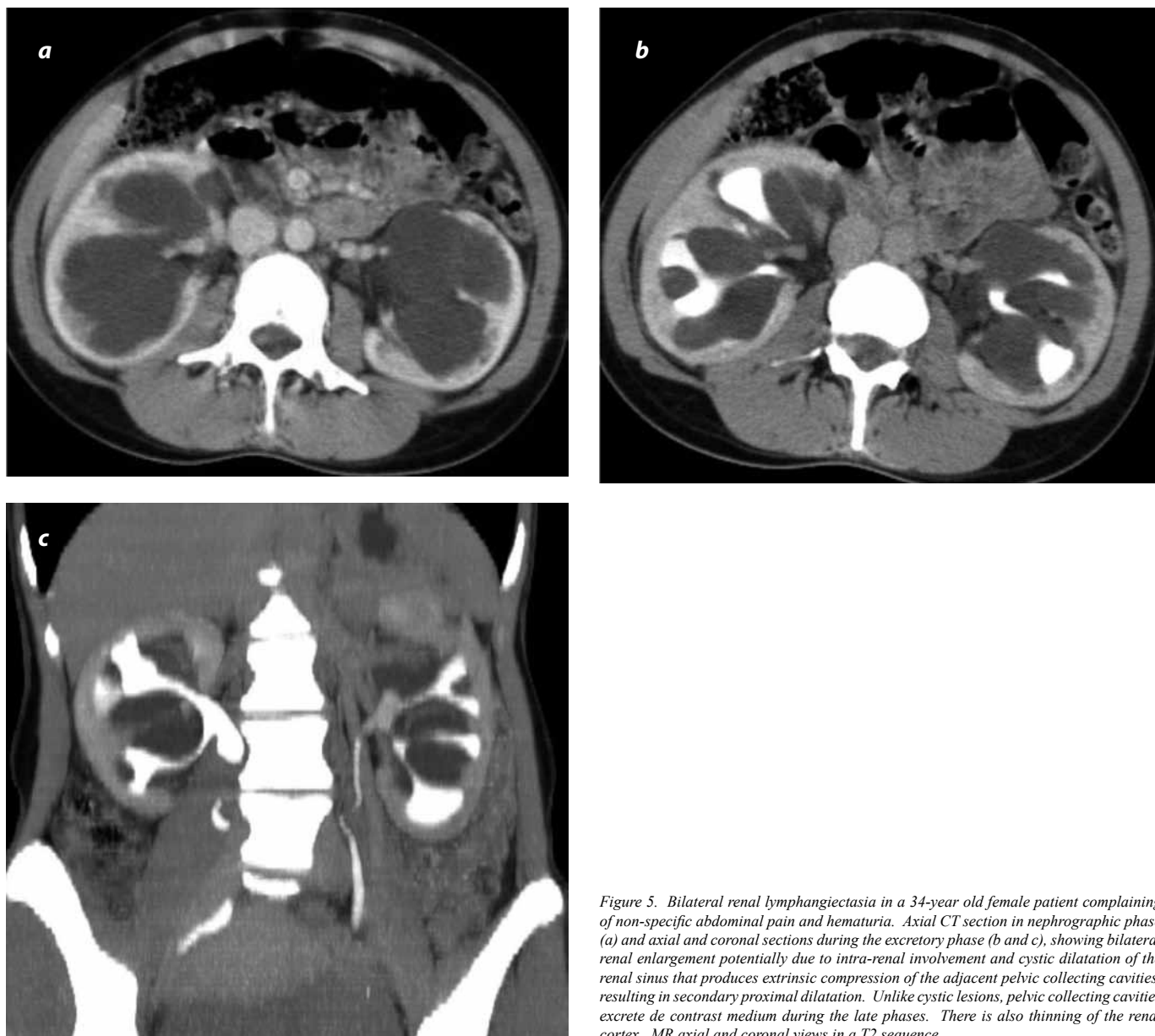


Figure 5. Bilateral renal lymphangiectasia in a 34-year old female patient complaining of non-specific abdominal pain and hematuria. Axial CT section in nephrographic phase (a) and axial and coronal sections during the excretory phase (b and c), showing bilateral renal enlargement potentially due to intra-renal involvement and cystic dilatation of the renal sinus that produces extrinsic compression of the adjacent pelvic collecting cavities, resulting in secondary proximal dilatation. Unlike cystic lesions, pelvic collecting cavities excrete de contrast medium during the late phases. There is also thinning of the renal cortex. MR axial and coronal views in a T2 sequence.

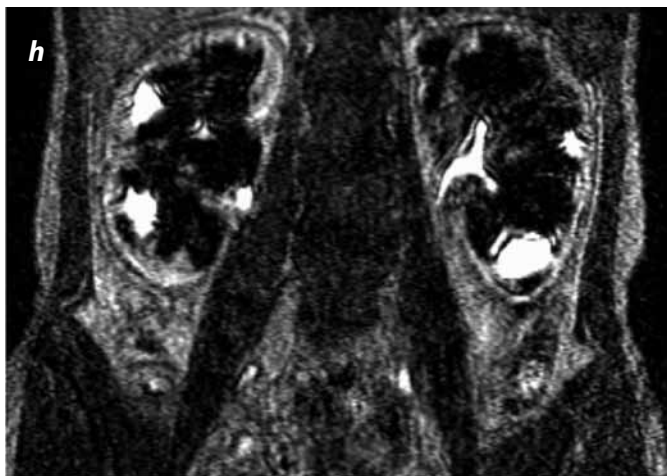
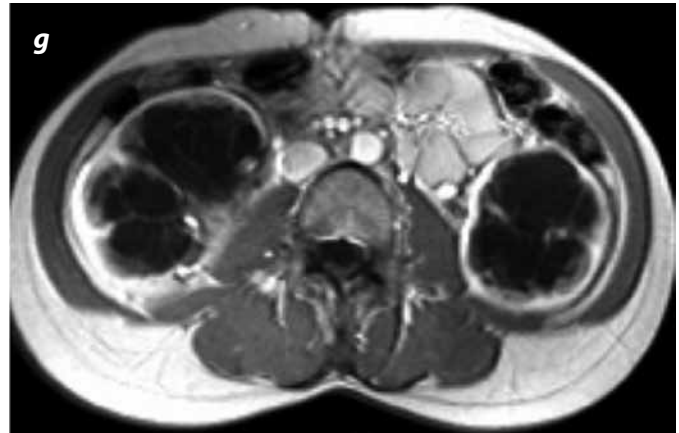
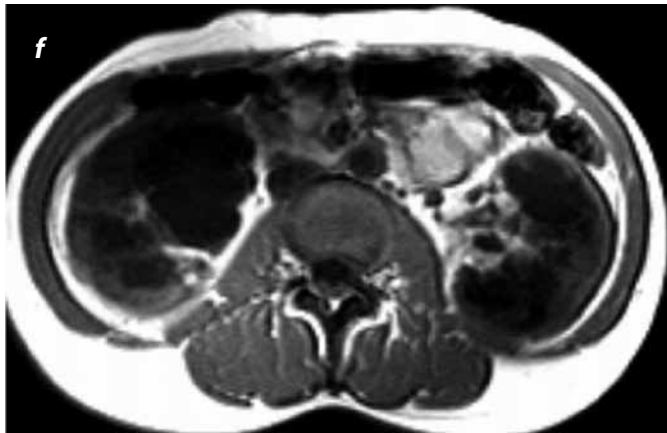
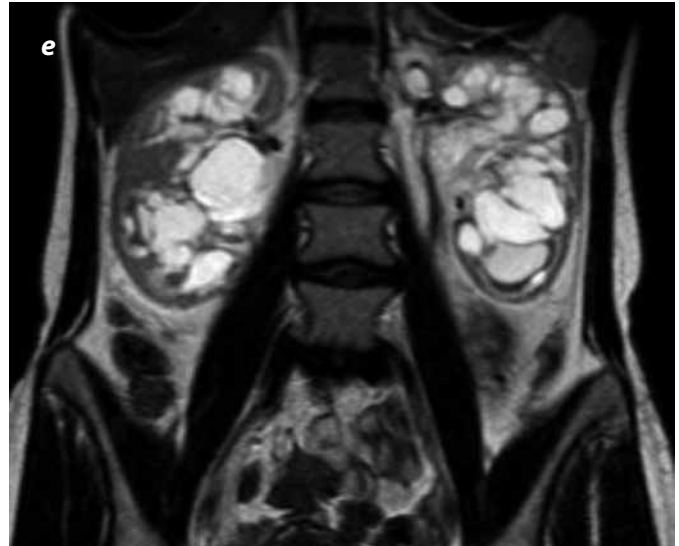
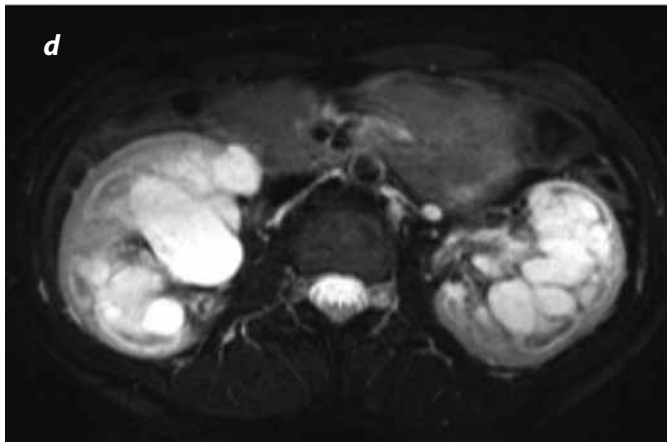


Figure 5. Bilateral renal lymphangiectasia in a 34-year old female patient who complains of non-specific abdominal pain and hematuria. Axial CT view in the nephrographic phase (d and e). There is clear evidence of thin septations forming cystic lesions that cannot be seen on CT. In this sequence it is not possible to differentiate cystic lesions from the collecting cavities. Non-contrast T1 sequences (f) obtained one minute after gadolinium administration (g), showing enhancement of the thin septations in renal lymphangiectasia. Late-phase uroresonance (h) showing gadolinium excretion through the dilated collecting cavities.

peripelvic, or involve several of these components, and the radiological manifestations will be consistent with those locations.

Plain X-rays will show soft-tissue mass appearance, due to increased renal size. Therefore, depending on their size, they may displace the adjacent structures, for example, the intestinal loops (1). On CT, one or several lesions with fluid attenuation may be found in the renal sinus; however, unlike ultrasound or MR, the multilocular appearance of the lesion may not be so evident. Characteristically, these lesions do not invade the adjacent structures, although they may cause displacement (Figures 1 and 4) (1).

On MR, cystic lesions will appear hypointense in T1 sequences, and hyperintense in T2. The presence of fine thin septations, found in many renal lymphangiectasias, can be adequately characterized with this imaging technique. Increased renal size and alterations in corticomedullary differentiation suggest intrarenal lymphangiectasia. Additionally, the renal parenchyma may be compressed and appear thinner than usual (Figure 5) (11).

Urotomography and uroresonance will be of great use in differentiating cystic lesions of the renal sinus from hydronephrosis, a finding which is often difficult to interpret in peripelvic lesions,

in particular on ultrasound, due to the fact that cystic lymphatic dilatations are distributed around the renal pelvis and calices (Figure 6). Additionally, they may reliably represent the displacement and distortion of the pyelocaliceal structures resulting from extrinsic compression. Although cysts originating in the renal sinus may be very large on occasions, the development of hydronephrosis is quite rare, considering that they do not usually produce renal hilar obstruction (11,12).

Differential diagnosis

Aside from hydronephrosis (Figure 6), the differential diagnosis of renal lymphangiectasia includes conditions such as polycystic kidney disease, nephroblastomatosis and tumor lesions such as multilocular cystic nephroma or lymphoma. Other causes of perinephric accumulations, such as urinomas and abscesses, must also be considered (1,2). An adequate diagnosis may be made on the basis of the clinical information, together with the typical imaging characteristics, although it is not always easy. Adult polycystic kidneys show evidence of scattered parenchymal cysts that may vary in size and may or may not be associated with pancreatic or hepatic cysts. In cases of lymphoma, masses

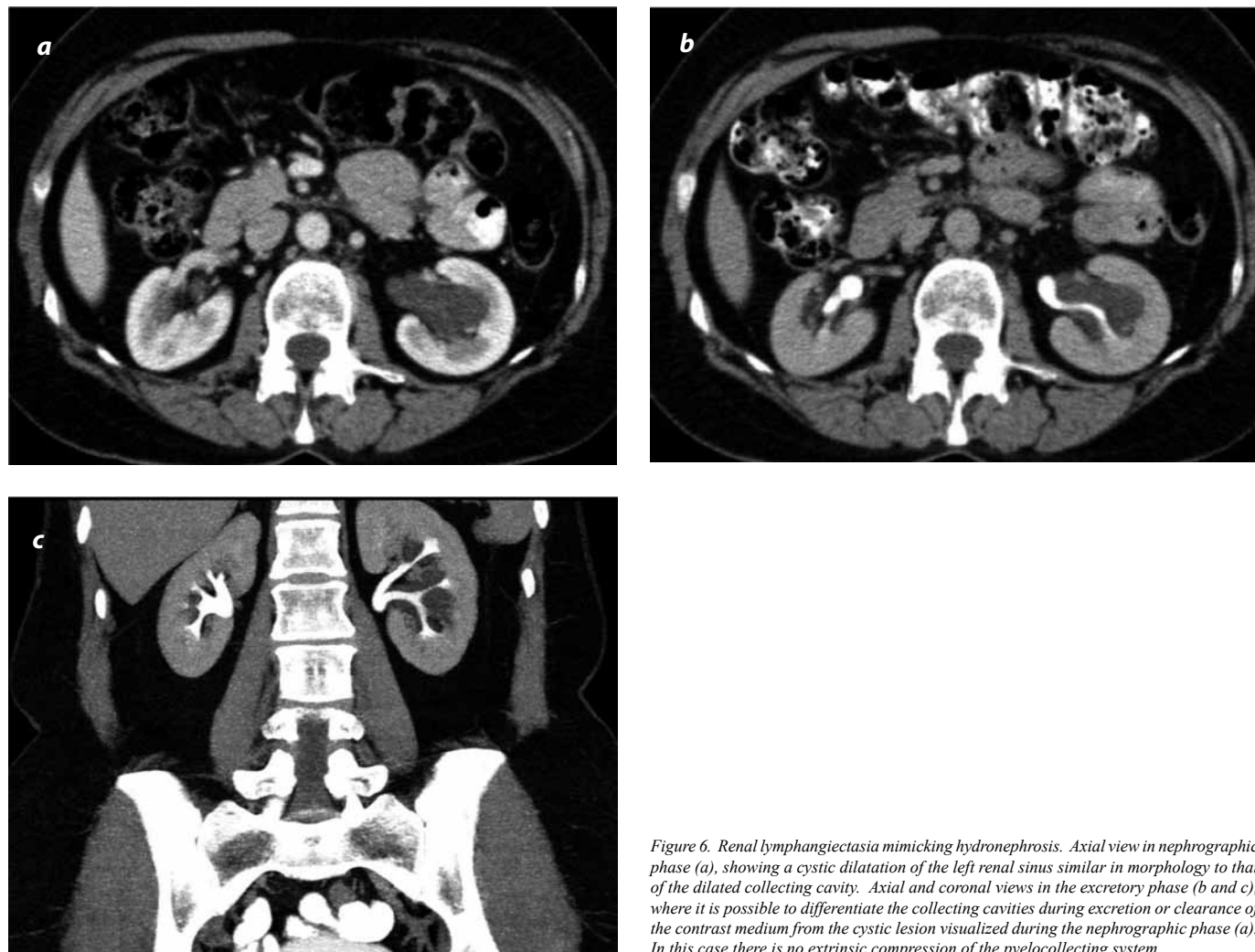


Figure 6. Renal lymphangiectasia mimicking hydronephrosis. Axial view in nephrographic phase (a), showing a cystic dilatation of the left renal sinus similar in morphology to that of the dilated collecting cavity. Axial and coronal views in the excretory phase (b and c), where it is possible to differentiate the collecting cavities during excretion or clearance of the contrast medium from the cystic lesion visualized during the nephrographic phase (a). In this case there is no extrinsic compression of the pyelocollecting system.

are observed with greater levels of attenuation than those found in renal lymphangiectasia. Nephroblastomatosis appears as a soft tissue mass in children, and causes kidney enlargement (11).

Complications

Complications include hematuria, ascitis, renal function decline and renin-dependent arterial hypertension (1). It has been found that pregnancy may exacerbate disease progression. In contrast, the disease has been found to be self-limiting in neonates (2,10).

Treatment

Treatment is not required in asymptomatic cases. In symptomatic cases or in pregnancy-associated exacerbations, percutaneous drainage may be performed (4). Percutaneous cyst aspiration is first-line treatment for renal cysts in many situations (2); however, it is important to remember—particularly in larger lesions—that multiseptation in renal lymphangiectasia renders aspiration unsuccessful in a large percentage of patients, resulting in high relapse rates. Recent studies have shown the usefulness of aspiration and sclerosis of cystic lesions of the renal sinus. However, sclerosing therapy is contraindicated for peripelvic cysts and for those that may be connected with the collecting system, due to the potential of producing stenosis from leakage of the sclerosing agent (2).

There are also options for symptomatic management such as diuretics for ascitis control, and antihypertensive medications for the treatment of arterial hypertension. Laparoscopic ablation and nephrectomy have been described for more severe cases (7,4,14).

Aspirate and laboratory findings

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 (10).

Conclusion

Renal lymphangiectasia is a rare entity; consequently, it is important for radiologists to be familiar with its characteristics and different forms of imaging appearance. In that way, we will be able to provide clinicians with the appropriate information required to determine the most adequate treatment for each individual patient.

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Received for evaluation: July 20, 2010

Accepted for publication: February 16, 2011