Case Report

A ruptured renal lymphangiectasia in the retroperitoneum: A rare complication of a rare condition

Nizar EL Bouardi, MD\textsuperscript{a,b,*}, Meriam Haloua, MD\textsuperscript{a,b}, Badreddine Alami, MD\textsuperscript{a,b}, Alaoui Lamrani Youssef, MD\textsuperscript{a,b}, Meriem Boubou, MD\textsuperscript{a,b}, Mustapha Maaroufi, MD\textsuperscript{a,b}

\textsuperscript{a}Radiology Department, Hassan II university hospital, 69 Quartier Riad, Avenue nassr; Agdal, Fez, Morocco
\textsuperscript{b}Faculty of Medicine and Pharmacy, Sidi Mohammed Ben Abdellah University, Boite Postale 1893 - KM 2.200 Route Sidi Harazem Fès, 30070, Fez, Morocco

**A B S T R A C T**

Renal lymphangiectasia is a rare and benign renal lymphatics disorder. It is usually asymptomatic, and rarely revealed by lumbar pain, arterial hypertension or retroperitoneal fluid collection. Radiological diagnostic is challenging and can be done if radiologists are aware of the imaging findings. Here in, we report a case of a young woman admitted for the management of a blunt abdominal trauma. Computed tomography showed a huge right retroperitoneal fluid collection, communicating with a perirenal cyst. This perirenal collection was bilateral and symmetrical. The diagnostic of a ruptured perirenal lymphangiectasia was suspected and confirmed by needle puncture. We managed the case using percutaneous drainage. Reassessment at subsequent follow-up visits showed a total regression of the collection improvement in the patient’s clinical.

© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

Cystic renal lesions are frequently seen in abdominal imaging. Most of them are benign simple cysts. Multifocal cystic renal disease is less common with a vast number of differentials. One of them is renal lymphangiectasia. Radiologists should be aware of the imaging feature of this disease so this allows the physician to offer the appropriate treatment. We report a case of a ruptured perirenal lymphangiectasia in the retroperitoneum, and we will also discuss its pathophysiology, clinical presentation, imaging appearances, treatment, complications and differentials.

Case presentation

A 24-year-old woman, with unremarkable medical history, presented to the emergency department for a blunt abdominal trauma. At her admission, the patient was alert and well orientated. Her blood arterial tension (130/75 mmHg) and heart
Fig. 1 – Coronal-enhanced CT in portal (A) and delayed phase (B) showing pelvic renal (white arrow) and perirenal cysts communicating with a huge right retroperitoneal collection (red arrow). These cysts were not filled with the contrast agent in the delayed phase excluding a hydronephrosis as a differential.

rate (82 pulses per minute) were correct. Physical examination showed a tumefaction of the right flank, that was sensible in the deep palpation, associated to a dullness in the percussion. There was no hematuria. The rest of the abdominal palpation was painless. Thoracic and extremities examination showed no anomalies. Neurological examination was normal. Thus, the patient underwent an abdominal triphasic computed tomography scanner in the arterial, portal and delayed phases (15 minutes) that showed a huge retroperitoneal fluid collection measuring 130×80mm, having a liquid density (8 Hounsfield units). This collection was communicating with a perirenal cyst. Cysts were however, bilateral and symmetrical. They were also peripelvic cysts (Fig. 1). There were no parenchymal or vascular injuries or blood extravasation. The diagnostic of a ruptured perirenal lymphangiectasia was suspected. A full abdominal magnetic resonance imaging was performed in the aim to assess the diagnostic and look for other associated lymphatic disorders. Both perirenal and retroperitoneal collections were hyperintense in T2 sequences and hypointense in T1 sequences (Fig. 2). There were no other lymphatic disorders, especially the enlargement of the cisterna chyli. The patient underwent a needle puncture of the retroperitoneal collection and laboratory analysis was carried out, which revealed the presence of proteins and fat.

Fig. 2 – Magnetic resonance imaging in T2 (A and B) and T1 showing bilateral peripelvic and perirenal cysts hyperintense in T2, hypointense in T1. Notice the huge retroperitoneal collection sharing the same signal as the cysts.
globules attesting the lymphatic nature of the liquid. Thereafter, the diagnosis of renal lymphangiectasia was confirmed. We managed the case using percutaneous drainage. Subsequent follow-up visits and computed tomography (Fig. 3) and sonography (Fig. 4) examinations showed improvement in the patient’s clinical status and a total regression of the retroperitoneal collection.

Discussion

Only very few cases of renal lymphangiectasia have been described in the literature. It represents only 1% of all lymphatic disorders [1]. It has an equal incidence in males and females, and can be present in both children and adults.

Lymphatic dilatations are seen in perirenal, peripelvic, and intrarenal locations. Thus, the new term “renal lymphangiectasia” is more descriptive of the nature of the disease and replaced older terms of the disease (renal lymphangiomatosi, renal lymphangioma) [2].

The pathophysiology of this disease remains unclear. As known, the lymphatic drainage of the renal capsule and the perirenal tissues is interconnected through several large lymphatic trunks located inside the renal sinus that drain into the para-aortic, para-caval and inter-aorto-caval lymph nodes [3]. The hypothesis of a developmental malformation of the drainage of these lymphatic trunks, leading to their dilatation and the creation of cystic voids adjacent to the renal sinus in some cases, in the perinephric space in the other cases, is the most plausible [3].

Clinical manifestations are usually absent and renal lymphangiectasia is an incidental finding in abdominal imaging. When it is symptomatic, it is usually associated to abdominal pain (42%), abdominal distension, (21%), fatigue, weight loss and hematuria [4]. In the most extreme cases, blood hypertension due to the compression of renal arteries, ascites and renal function decline have been described [4]. Some authors report the aggravation of symptoms during pregnancy [5].

In children, it may also manifest with a palpable abdominal mass, kidney enlargement or pyelonephritis.

Sonography frequently demonstrates anechoic lesions with increased through transmission and sharply defined thin wall. These cystic spaces extend along and into the peripelvic region as the larger lymphatic channels drain through the renal pedicle.

Computed tomography examination reveals peripelvic or perirenal multiloculated cystic lesions with fluid attenuation
ranging from 0 to 10 Hounsfield units. Higher attenuations are secondary to intracystic hemorrhage. The walls and septations are usually thin and regular, but may also be thick and irregular. Rarely, retroperitoneal fluid collections may be present. Free intraperitoneal fluid may be seen in severe cases [6].

The typical appearance on MRI of lymphangiectasia consists of cystic lesions that are hypointense on T1 and hyperintense on T2. Also, the involved kidney may be enlarged with increased cortical intensity and decreased medullary intensity in T2. In some cases, retroperitoneal perivascular thin lymphatic channels can be seen especially a tortuous dilatation of cisterna chyli and thoracic duct. More recently, lymphoscintigraphy has been used to detect abnormal lymphatic flows associated with lymphangiectasia.

Needle puncture of the cyst is the key to the diagnosis, allowing to distinguish renal lymphangiectasia cysts from other renal cysts. The presence of proteins and fat globules confirms the lymphatic origin of the cysts. Laboratory features of these renal fluid collections in distinguishable from normal lymph by the presence only of sporadic cells, small amounts of fat globules, proteins and high renin levels in the fluid, suggesting the renal origin of the lymph [7].

Differentials of renal lymphangiectasia include hydronephrosis, polycystic diseases, nephroblastomatosis and multicellular cystic nephroma depending on the age and appearance of the disease. Other causes of perihepatic collections have also to be ruled out (urinoma or abscess) [8].

Complications include hemorrhage, ascites, and hypertension secondary to perirenal fluid collection. Less commonly reported complication is venous thrombosis and rupture [9].

Percutaneous drainage can be useful in the conservative management of symptomatic patients, when collections are very large and causing pressure symptoms or in exacerbation during pregnancy. Marsupialization has been reported as a treatment option where a connection is made with the peritoneal cavity, draining the fluid into it. Some drugs can be used including diuretics to control the ascites, anti-hypertensive in the case of arterial hypertension. In severe uncontrollable cases, nephrectomy may be performed [10].

To conclude, radiologists play a crucial role in the diagnostic of this rare condition, allowing physicians a suitable and early management before complications.

---

**Patient consent**

Consent was obtained from the patient. The study was conducted anonymously.

**Availability of data and materials**

The data sets are generated on the data system of the university hospital of Fez.

**References**

[1] Hauser H, Mischinger HJ, Beham A, Berger A, Cerwenka H, Razmara J, et al. Cystic retroperitoneal lymphangiomas in adults. Eur J Surg Oncol 1997;23(4):322–6.

[2] Schwarz A, Lenz T, Klaen R, Offermann G, Ulrich F, Nussberger J. Hygroma renale: pararenal lymphatic cysts associated with renin dependent hypertension (page kid-ney). Case report on bilateral cysts and successful therapy by marsupialization. J Urol 1993;150:953–7.

[3] Gupta R, Sharma R, Gamanagatti S, Dogra PN, Kumar A. Unilateral renal lymphangiectasia: imaging appearance on sonography, CT and MRI. Int Urol Nephrol 2007;39:361–4.

[4] Davidson AJ, Hartman DS. Lymphangioma of the retro-peritoneum: CT and sonographic characteristics. Radiology 1990;175:507–10.

[5] Meredith WT, Ahlstrom NG, Levine E, Grantham JJ. Exacerbation of familial renal lymphangiomatosis during pregnancy. Am J Roentgenol 1988;151:965–6.

[6] Ramseyer LT. Case 34: renal lymphangiectasia. Radiology 2001;219:442–4.

[7] Pandya VK, Shah MK, Gandhi SP, Patel HV. Bilateral renal lymphangiectasia. J Clin Diagn Res 2016;10(9):TD01–2. doi:10.7860/JCDR/2016/19475.8409.

[8] Umapathy S, Alavandar E, Renganathan R, Thambidurai S, Kasi Arunachalam V. Renal lymphangiectasia: an unusual mimicker of cystic renal disease – a case series and literature review. Cureus 2020;12(10):e10849. doi:10.7759/cureus.10849.

[9] Riehl J, Schmitt H, Schafer L, Schneider B, Sieberth G. Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis. Nephrol Dial Transplant 1997;12:1701–3.

[10] Ashraf K, Raza SS, Ashraf O, Memon W, Memon A, Zubairi TA. Renal lymphangiectasia. Br J Radiol 2007;80(954):e117–18.