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Peri-Renal Lymphangiomatosis: About A Case

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Abstract **Case Report**

Renal lymphangiectasia is a rare disorder of the lymphatic system responsible for cystic infiltration of the bilateral perirenal and parapyelic space. The clinical presentation is nonspecific, hence the role of multimodal imaging which can show characteristic lesions allowing a positive diagnosis. We report a case of bilateral lymphangiectasia in a 60-year-old woman presenting with chronic bilateral low back pain with glomerular syndrome, and in whom the diagnosis was made using ultrasound and cross-sectional imaging, computed tomography.

Keywords: Renal lymphangiectasia, Bilateral perirenal and parapyelic space, Glomerular syndrome, Ultrasound, Computed tomography.

INTRODUCTION

Renal lymphangiectasia is a congenital malformation of the renal lymphatic system leading to obstruction and accumulation of lymph with cystic infiltration of the perirenal and bilateral parapyelic space, rarely described in the medical literature. The clinical presentation is non-specific, hence the role of multimodal imaging which allows to show cystic lesions in the perirenal space, sinus and hilum and especially the connection of these lesions with a dilatation of the lymphatic system of the retroperitoneum [1].

OBSERVATION

We report the case of a 60-year-old adult woman, referred for the exploration of a glomerular syndrome associated with chronic bilateral lower back pain. The patient's history was high blood pressure since 2009, which was under treatment. With a cervical tumor, with in particular an absence of nephrotoxic drugs or medicinal plants. The clinical examination found a conscious patient, without edema of the lower limbs and on the urine strip two protein crosses and zero blood crosses. Regarding the biological assessment, it objectified the presence of a glomerular syndrome made up of proteinuria at 3.3 g/24h without impact on the proteinogram with a creatinine level of 6.7 mg/l, i.e. a GFR of 101 ml/min/1.73m² "MDRD".

We completed an ultrasound scan in the context of an emergency of renal colic and for the study of the renal parenchyma objectifying a cystic dilatation with finely echogenic content in the perirenal and parasinus connecting to the level of the sinus associated with nephromegaly and kidneys with echogenic cortex with moderate dilatation of the pyelocaliceal cavities and the subpyelic ureter following tumor infiltration of the outlet of the two ureters (Figure 1)

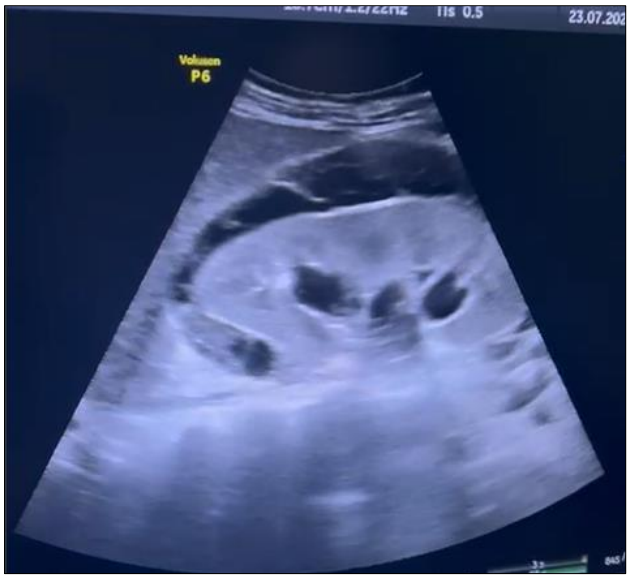

Figure 1

By completing with a uroscanner (arterial, portal, late phase) (Figure 2), which objectified the presence of a multilocular cystic infiltration in the bilateral and symmetrical perirenal crown of liquid density, without enhancement after injection of contrast product.

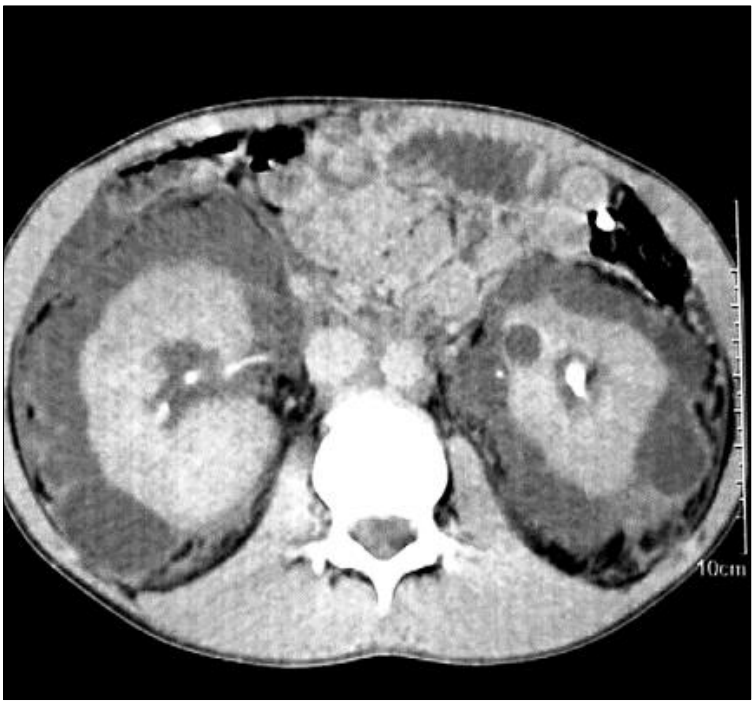

Figure 2

Abdominal MRI in T2 sequence in coronal and axial section, T1 sequence and diffusion in axial section shows the presence of a perirenal collar composed of small cysts, liquid signal, in T1 hypointense, T2 hypersignal and diffusion, without detectable contrast uptake with respect to the parenchyma.

The diagnosis of perirenal lymphangiectasia was retained. For the exploration of HTA, a thyroid assessment, a cardiac echo, a Doppler echo of the renal arteries and the fundus were normal. The patient was put on nephroprotective treatment with clinical and biological monitoring. The evolution was marked by the negativation of proteinuria.

DISCUSSION

Renal lymphangiectasia is a very rare benign disease of the renal lymphatics. Over the years, this disorder has had many names including renal lymphangiomtosis [2], peri-lymphangiectasia, renal hygroma and polycystic renal sinus disease [3]. The

etiopathogenesis of this disease remains poorly understood, it is generally considered to be a malformation of the renal lymphatic system preventing communication with the rest of the lymphatic trunks. The abnormal lymphatics dilate and form cystic spaces in the renal sinus and in the perirenal space [4]. Clinically, it is generally asymptomatic, but it can be revealed by several symptoms, most often these are abdominal pain such as renal colic (42%). Other symptoms are described in the literature as palpable masses (21%), hematuria, fatigue, weight loss, high blood pressure or occasional deterioration of renal function (most often reversible) [4, 5]. The imaging characteristics are simple to understand. Ultrasound most often reveals the presence of multiple symmetrical, perirenal, fluid-filled cystic lesions that can cause distortion of the kidney contours. Nephromegaly with loss of cortico-medullary differentiation has also been reported [6, 7]. Cystic lesions are well demarcated on CT scan. They appear as homogeneous, thin-walled collections with homogeneous fluid content, located perirenal, distorting the kidney contours [8, 9]. Diffuse renal hypertrophy may be observed [10]. MRI has allowed better characterization of cystic lesions and assessment of their true extent with better sensitivity and specificity. Another advantage of MRI is that it can perform lympho-MRI, thanks to the highly T2-weighted sequences, which allows the detection of dilatation of the retroperitoneal lymphatic vessels and prove communication with renal sinus lesions. This is a key specific sign of diagnosis. Dilation of the peri-, centro-renal and retroperitoneal lymphatics can cause compression of the renal parenchyma and excretory tracts with hydronephrosis that can lead to renal failure [11]. The evolution can be marked by a major increase in the volume of cystic formations with a risk of compression of the pyelocaliceal excretory tracts and the vascular pedicle [3]. Certain complications may appear such as hematuria, ascites, occasional deterioration of renal function and arterial hypertension by activation of the renin-angiotensin system [3-5]. Differential diagnoses of renal lymphangiectasia include polycystic kidney disease, nephroblastomatosis, lymphoma [4-6]. Other causes besides perinephric collections should also be excluded,such as urinomas and abscesses. Treatment of asymptomatic cases is not necessary. When collections are very large and cause compression of surrounding structures, or in case of exacerbation, such as that encountered during pregnancy, they may require percutaneous drainage [2]. Marsupialization is a treatment option when a connection is made with the peritoneal cavity [5]. Diuretics may be prescribed in case of ascites. Antihypertensive drugs are used to control hypertension. In severe and uncontrollable cases, nephrectomy may be performed.

CONCLUSION

Renal lymphangiectasia is a rare benign renal disease with characteristic imaging findings, the management of which is conservative in most cases. The prognosis depends on the compression of the urinary excretory tract, renal parenchyma and vascular pedicle, hence the need for regular follow-up.

REFERENCES

1. Ashraf, K., Raza, S. S., Ashraf, O., Memon, W., Memon, A., & Zubairi, T. A. (2007). Renal lymphangiectasia. *The British journal of radiology*, *80*(954), e117-e118.
2. Meredith, W. T., Levine, E., Ahlstrom, N. G., & Grantham, J. J. (1988). Exacerbation of familial renal lymphangiomatosis during pregnancy. *American Journal of Roentgenology*, *151*(5), 965-966.
3. Riehl, J., Schmitt, H., Schäfer, L., Schneider, B., & Sieberth, H. G. (1997). Retroperitoneal lymphangiectasia associated with bilateral renal vein thrombosis. *Nephrology, dialysis, transplantation: official publication of the European Dialysis and Transplant Association-European Renal Association*, *12*(8), 1701-1703.
4. Davidson, A. J., & Hartman, D. S. (1990). Lymphangioma of the retroperitoneum: CT and sonographic characteristic. *Radiology*, *175*(2), 507-510.
5. Schwarz, A., Lenz, T., Klaen, R., Offermann, G., Fiedler, U., & Nussberger, J. (1993). Hygroma renale: pararenal lymphatic cysts associated with renin-dependent hypertension (Page kidney). Case report on bilateral cysts and successful therapy by marsupialization. *The Journal of urology*, *150*(3), 953-957.
6. Blumhagen, J. D., Wood, B. J., & Rosenbaum, D. M. (1987). Sonographic evaluation of abdominal lymphangiomas in children. *Journal of ultrasound in medicine*, *6*(9), 487-495.
7. Pickering, S. P., Fletcher, B. D., Bryan, P. J., & Abramowsky, C. R. (1984). Renal lymphangioma: a cause of neonatal nephromegaly. *Pediatric radiology*, *14*, 445-448.
8. Sarikaya, B., Akturk, Y., Bekar, U., & Topaloglu, S. (2006). Bilateral renal lymphangiomatosis mimicking hydronephrosis: multidetector CT urographic findings. *Abdominal imaging*, *31*, 732-734.
9. Wadhwa, P., Kumar, A., Sharma, S., Dogra, P. N., & Hemal, A. K. (2007). Renal lymphangiomatosis: imaging and management of a rare renal anomaly. *International Urology and Nephrology*, *39*, 365-368.
10. Kim, J. K., Ahn, H. J., Kim, K. R., & Cho, K. S. (2002). Renal lymphangioma manifested as a solid mass on ultrasonography and computed tomography. *Journal of ultrasound in medicine*, *21*(2), 203-206.
11. Wani, N. A., Kosar, T., & Gojwari, T. (2010). Perinephric fluid collections due to renal ymphangiectasia. *Am J KidneyDis*, *56*(1), 181-4.