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Case Report
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UNILATERAL MEDULLARY SPONGE KIDNEY

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ABSTRACT

A 40-year-old female with an allergy to medium contrast, presented with bilateral loin pain and renal colic type radiation. Image study with non-contrast computerized tomography and magnetic resonance imaging revealed multiple nephrolithiasis and a complex cyst at lower pole of the right kidney and no findings in the left one. The patient was treated with laser lithotripsy by flexible ureterorenoscopy revealing multiple lithiasis within cystic dilatations of the urothelium. The complex cyst was submitted to partial nephrectomy. Finally, biopsy revealed renal parenchyma with polycystic changes in the external corticomedullary area. All findings were compatible with unilateral medullary sponge kidney diagnosis.

CLINICAL CASE

A 40-year-old female patient presented with a history of insulin resistance (treated with metformin), allergy to contrast medium, and maternal urolithiasis. The patient consulted for bilateral loin pain and right renal colic pain. Non-contrast computerized tomography (NCCT) (Figures 1 and 2) demonstrated multiple right nephrolithiasis and a 3-cm hypodense right lower pole lesion with some calcifications. This was diagnosed as a complex cyst. Related to this cyst, a magnetic resonance imaging (MRI) of the abdomen was requested which showed a 3.5 cm Bosniak 3 right renal cyst.

Resolution of the lithiasis was decided by means of flexible ureterorenoscopy (fURS) with laser lithotripsy. During nephroscopy, multiple lithiasis were found within cystic dilatations of the urothelium (Figure 3). Using laser, some of these dilatations were opened exposing multiple lithiasis of 2 to 3 millimeters. In addition, by means of radioscopy other calculi that were not visualized in the nephroscopy were detected. The patient progressed favourably, without incident.

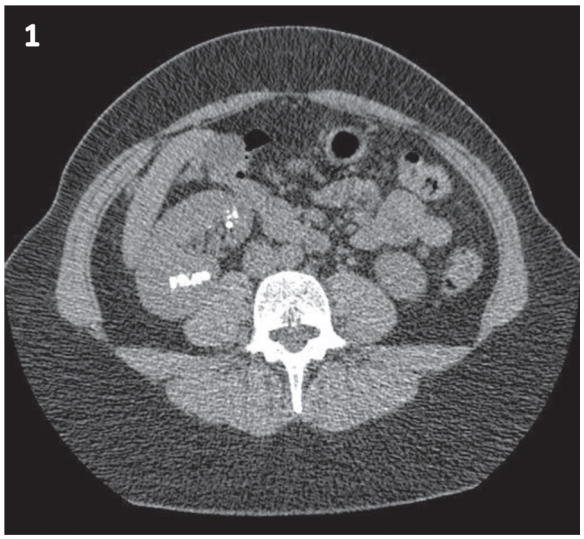

FIG. 1 Multiple right nephrolithiasis.

In relation to the Bosniak 3 renal cyst, the patient underwent a laparoscopic partial nephrectomy without incident. Biopsy showed "Fragment of kidney with polycystic changes in the external corticomedullary area, with renal dysplasia component." This was

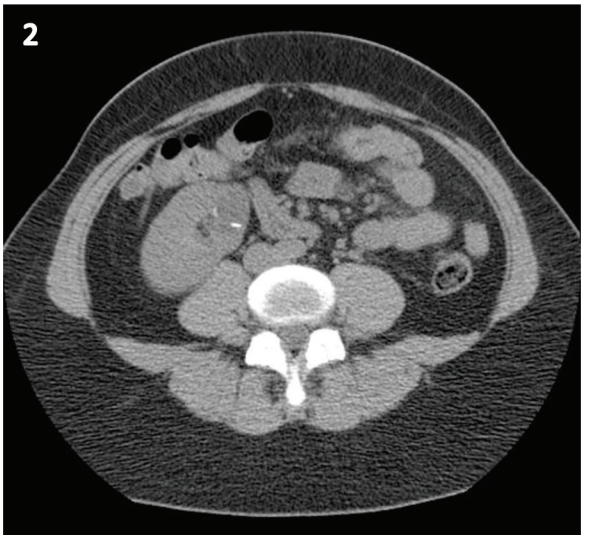

FIG. 2 Hypodense 3-cm lesion with a cystic appearance in the lower pole of the right renal cortex with some peripheral calcifications.

compatible with a renal polycystic disease although is not pathognomonic for unilateral medullary sponge kidney (MSK).

In subsequent follow-up of her cystic pathology and considering that it is not possible to perform an Intravenous Pyelogram (IVP) due to the allergy to the contrast medium, she had an Uro-MRI that shows persistent right nephrolithiasis, in context of MSK. However, after fURS, the patient remains asymptomatic.

DISCUSSION

MSK is a rare renal congenital malformation characterized by a tubular dilatation of the distal portion of the collecting ducts, with numerous cysts and diverticula strictly confined to the medullary pyramids.[1] Generally it occurs bilaterally, being rare the unilateral form. This malformation causes nephrocalcinosis and recurrent calcium lithiasis secondary to the presence of hypercalciuria, hypocitraturia and distal renal tubular acidosis (dRTA) as long as there is urinary stasis in the dilated ducts. It is considered an uncommon pathology, with a prevalence in the general population of 0.5–1%, being much more frequent in calcium stone formers.[2]

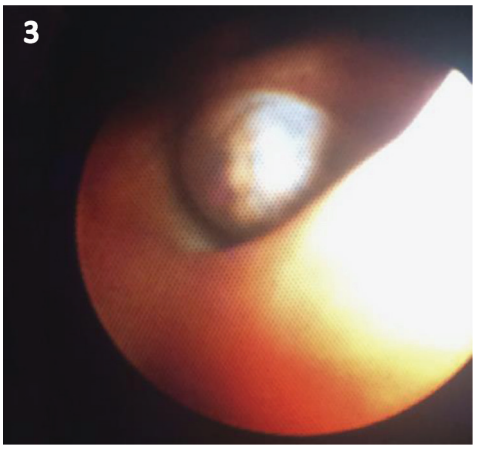

FIG. 3 Cystic dilatations for the urothelium.

It occurs equally in both genders, usually in young adults. The most common clinical manifestation is urolithiasis, frequently recurrent and usually accompanied by nephrocalcinosis, followed by pyelonephritis. Some patients may present only pain without stone passage or infection.[3] Furthermore, it has been associated with other diseases such as Wilms tumor, polycystic disease, horseshoe kidney, hyperparathyroidism, congenital hepatic fibrosis, Caroli syndrome, Ehlers-Danlos syndrome, pyloric stenosis, among others.[4]

Currently, the Gold Standard for diagnosis is IVP. Diagnostic findings include papillary blush in mild cases and linear striations known as "paint brush" and papillary "bouquets" in the most severe cases.[5]

The widespread use of CT scan has reduced the use IVP, since it is the most used method in the context of renal colic; however, given that papillary dilatations are very thin (1 mm or less), a high spatial resolution is required for its detection, which cannot be achieved by CT scan (4 mm thick axial slices). Another method is MRI; however, it is not widely available, and it is not a routine examination in the study of patients with nephrolithiasis because of its limited ability for detecting urinary tract stones.[6,7]

Biopsy of the papillae can document a pathognomonic triad for MSK: undifferentiated interstitial cells, abnormal multilayered inner medullary collecting ducts (IMCD) epithelium, and IMCD dilation.[8]

Within the therapeutic options, the conservative treatment is recommended in patients with stone risk factors (hypercalciuria, hypocitraturia, hyperuricosuria, and hyperoxaluria). This is mainly based on improving the metabolic conditions that favours the formation of calculi, usually using potassium citrate, along with clinic recommendations concerning diet and the increase in water intake. These recommendations seem to be effective in preventing the formation of calculi and its complications.[9] In the case of symptomatic recurrences, the treatment of lithiasis can be performed by laser papillotomy using ureteroscopy, which is considered a safe and effective method.[10]

In our patient, all the clinical and radiological findings point to a unilateral MSK diagnosis despite the fact that it is not possible to perform an IVP, which is considered the Gold Standard in these cases. We are facing a rare disease, with a high rate of recurrence and a pathophysiology that is not yet completely understood.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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