4th Edition of the Clinical Case Contest related to the non-surgical clinical management of renal lithiasis.

Offical template

Title: Chemolysis of uric acid lithiasis in a patient with crossed renal ectopia.

Keywords: ectopia; chemolysis; urinary pH; uric acid; alkalinization.

1. Abstract

We present the case of a male with crossed and fused renal ectopia, who following an episode of abdominal pain, was diagnosed with high volume renal lithiasis disease associated with pyelocaliceal dilatation. Given the characteristics of the patient and the lithiasis, urinary alkalinization by means of pharmacotherapy and dietary recommendations was decided. The patient presented a clear decrease in the lithiasis load and pyelocaliceal dilatation, with no side effects. Currently, he remains with the same treatment and is being followed up in outpatient clinics.

2. Introduction

Among renal congenital anomalies, renal ectopia is an uncommon alteration. Also, within this picture, crossed and fused renal ectopia is an extremely rare subtype of renal ectopia. This is an anomaly in which both kidneys are fused and on one side of the midline of the body, maintaining two renal pelvises and two ureters that will end one on each side of the bladder. Fused crossed renal ectopia is usually asymptomatic. The diagnosis is usually incidental when imaging tests are ordered for other reasons.

The following is the case of a 68-year-old male with fused crossed renal ectopia and high lithiasic volume. Different treatment options were considered, finally opting for chemolysis. There are few cases reported in the literature on the management of lithiasis in crossed and fused renal ectopia and its pharmacological treatment.

3. Description of the clinical case

a. Relevant background

A 68-year-old male with incidental diagnosis, in 2013, of crossed and fused renal ectopia. Renal function is preserved. History of renal lithiasis that did not require treatment.

b. Diagnosis support studies and results

Blood test on arrival at the Emergency Department:

- Blood test: L 14,500; Hb 16.1 g/dL; platelets 223,000.

- Biochemistry: Cr 1.46 mg/dL; 49.10 mL/min/1.73 m2 Urine systemic: pH 5.5

Blood analysis at discharge from the emergency department:

- Hemogram: L 8 600; Hb 14.3 g/dL; platelets 168 000

- Biochemistry: Cr 1.27 mg/dL; GFR 58.11 mL/min/1.73 m2

Abdominal X-ray (Figure 1): no lithiasis was observed.

CT abdomen pelvis without and with contrast (figures 2 and 3):

Single left kidney (crossed and fused renal ectopia) of pelvic location, normal cortical thickness and adequate contrast uptake; with complete double excretory system already known. Multiple high density images in the interior of the upper and middle caliceal groups in relation to lithiasis, some of them of great size and more numerous in the current study especially in the superolateral extrasinusal pyelonephrosis. No lithiasis are identified in the course of both ureters or inside the bladder. Double pyelocaliceal dilatation (up to 42 mm in superolateral extrasinusal pyelonephrosis).

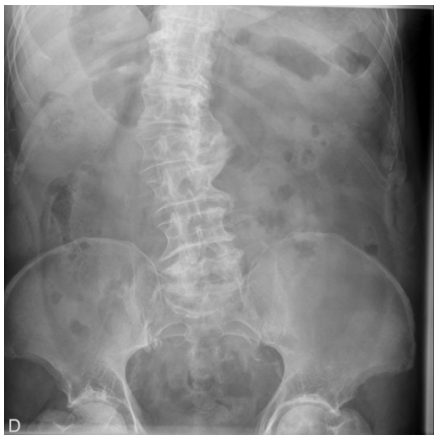

Figure 1

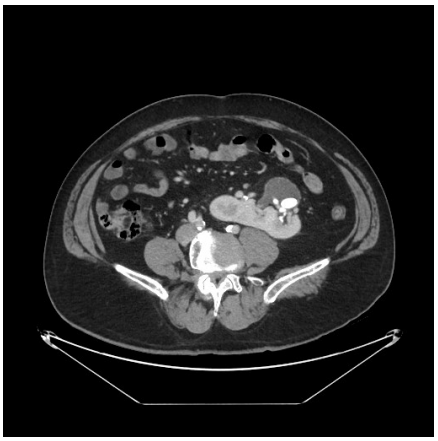

Figure 2

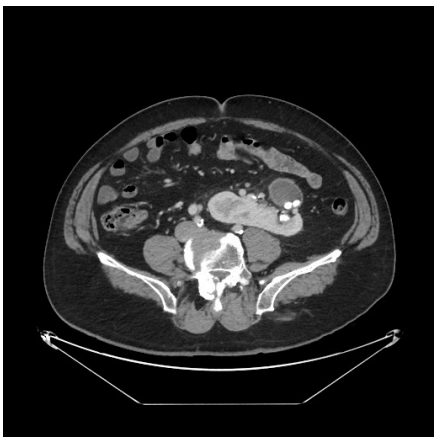

Figure 3

c. Diagnosis

Patient with the previously described history was evaluated in the Emergency Department for hypogastric abdominal pain of 3 days of evolution, with associated dysuria and pollakiuria. No associated gastrointestinal symptoms or fever.

Laboratory tests and abdominal X-ray (Figure 1) were requested from the Emergency Department, with no clear findings suggestive of lithiasis. When reviewing previous imaging tests, a previous CT scan from 2018 was found, describing crossed and fused renal ectopia as well as the presence of renal

lithiasis. Given the moderate deterioration of the patient's renal function, it was decided to request a CT scan of the abdomen pelvis (Figures 2 and 3), which showed multiple renal lithiasis and pyelocaliceal dilatation.

At that time the patient did not present criteria for urgent urinary tract referral, so after the cessation of pain and improvement of renal function, he was discharged with a diagnosis of renoureteral colic. Subsequently, an appointment was made in the outpatient urology department for review.

In Urology consultations, complementary tests were reviewed and it was found that the patient had lithiasis with 400-500 Hounsefield units, which were radiolucent in the abdominal X-ray. These findings, together with acid urine pH, suggest uric acid lithiasis.

d. Treatment

During his stay in the emergency department, the patient received analgesic treatment avoiding NSAIDs (due to renal function impairment) as well as fluid therapy. Analgesia was also prescribed at discharge.

As for the management of the lithiasis, initially a surgical approach by ureterorenoscopy was proposed. Percutaneous nephrolitectomy was ruled out given the renal location. Prior to the procedure, an attempt was made to place a double J catheter. However, it was impossible due to impassable stenosis at the anterior urethra. The case was presented in a clinical session. The anatomical characteristics of the patient (medial location of ectopia, tortuous ureteral tract, urethral stricture), the high volume of lithiasis and the high suspicion of uric acid lithiasis, led to modify the management of the case. It was decided to opt for pharmacological treatment and if the lithiasis was not resolved, to consider surgical treatment again. Thus, the patient started treatment with potassium and magnesium citrate (Lit-Control® pH Up) at a dose of 1 capsule every 8 hours, associating hygienic-dietary measures (exercise, adequate water intake and dietary control). For better control and follow-up, the possibility of using the myLit-Control® App was offered, which measures water intake, urinary pH and the intake of food supplements.

e. Evolution and follow-up

After initiating pharmacological treatment, it was agreed to follow up every 3 months with imaging tests (alternating ultrasound and CT) and systematic urine analysis. At the last review, the patient had been on pharmacological treatment for 9 months. Systematic urine analysis was performed, showing an adequate range of chemolysis (pH around 6.9) as well as control CT (Figures 4 and 5) which showed a significant decrease in the volume of lithiasis as well as previous pyelocaliceal dilatation.

The patient has had no complications during follow-up, reporting correct tolerance to treatment and denying any side effects. He is currently being followed up in the urology outpatient clinic, maintaining the same treatment with the aim of eliminating the remaining lithiasis.

f. Clinical results

Urine systemic: urinary pH 7.1
CT abdomen pelvis without contrast (figures 4 and 5).

Single left kidney (crossed and fused renal ectopia) of pelvic location, normal cortical thickness and adequate contrast uptake; with complete double excretory system already known. Multiple images of lithiasis are identified in upper superior and middle caliceal groups that have decreased in number. Comparatively, a decrease in the pyelocaliceal dilatation of the upper and extrasinusal pyelonephrosis is identified, not identifying the lithiasis images that were seen in the pelvis in the previous study. No lithiasis images are demonstrated in the ureteral tract or bladder. At the present time no pyelocaliceal dilatation of the lower pyelon.

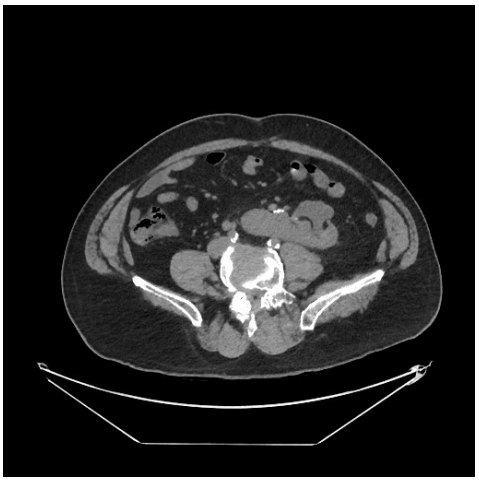

Figure 4

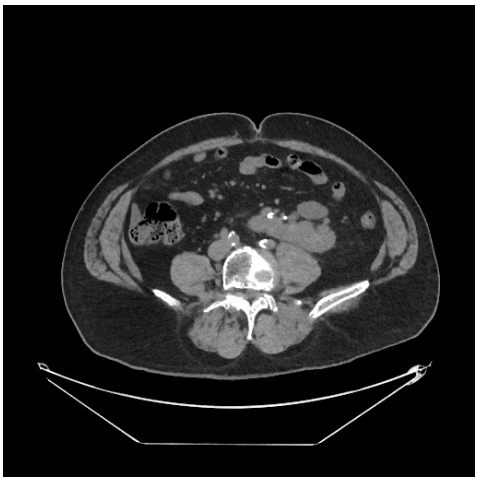

Figure 5

4. Discussion

Fused crossed renal ectopia is a rare congenital anomaly in which one kidney crosses the midline and sits next to the orthotopic kidney. The ureters, on the other hand, remain in position, inserting on either side of the trigonal bar. This anomaly has been described more frequently in males than in females, in a 3:2 ratio. Likewise, left renal ectopia (both kidneys are on the right side) is more common than right renal ectopia.

Depending on the location, the degree of rotation of the fused portion as well as the extent of the fusion, we can find different types of fused renal ectopia (unilateral ectopia with fusion of the upper or lower pole; sigmoid or "S" kidney; pancake kidney; "L" kidney and discoid kidney).

To understand the origin of this anomaly we must briefly review the formation of the genitourinary system during embryonic development. The formation of the metanephros or definitive kidney begins in the fifth week of intrauterine life and culminates in the ninth week. Its development depends on the interaction of the ureteral bud that gives rise to the ureter, pelvis, calyces and collecting tubules and the nephrogenic mesenchymal blastema from which the nephrons derive. The renal blastema originates at the level of the upper sacral segments. Its ascent to its final position at the level of the lumbar vertebrae is attributed to four mechanisms (caudal growth of the spine, elongation of the ureter, modeling or rotation of the renal parenchyma, and fixation of the kidney to the retroperitoneum). Factors that prevent this normal renal ascent will cause renal ectopia such as abnormal development of the ureteral bud, defective metanephric tissue, and genetic abnormalities. There are several theories that have attempted to explain the origin of this anomaly (mechanical, ureteral, induced migration, teratogenic theory). However, the origin of this anomaly is currently unknown.

Most patients with this anatomical alteration are asymptomatic and the finding is incidental when performing imaging tests for other reasons. However, the presence of urinary tract infections and lithiasis is not uncommon given the morphology of the urinary tract. Both conditions usually manifest as diffuse abdominal pain, hematuria or voiding syndrome.

As for imaging tests, there are several that can help in the diagnosis (ultrasound, intravenous urography, CT, magnetic resonance imaging). Generally, the one that will provide us with the most information (anatomical characteristics of the ectopia, its relationship with surrounding structures) is CT. Especially with a view to planning a surgical intervention.

Regarding the management of lithiasis in patients with this type of congenital anomaly, there is currently no clear expert consensus. There are cases described in the literature in which extracorporeal shock wave lithotripsy is used, while in other cases ureterorenoscopy or percutaneous nephrolitectomy are used. Regarding the use of pharmacological chemolysis, no case of these characteristics has been described in the literature.

Percutaneous nephrolitectomy is considered the "gold standard" procedure for the treatment of high volume renal lithiasis (20 mm and larger). Some authors advise the use of this technique for the treatment of lithiasis in patients with crossed and fused renal ectopia. However, it is not free of complications. The location of the kidney increases the risk of damage to the renal pedicle as well as to adjacent organs. That is why in our case this type of intervention was discouraged.

Ureterorenoscopy has gained much importance in recent years. Especially since the use of flexible ureterorenoscopes. It is currently indicated for the management of ureteral lithiasis and renal lithiasis up to 20 mm (in larger lithiasis, percutaneous surgery should be considered as the first option, unless this cannot be an option). In the case of our patient, this technique was initially chosen. However, the patient's right ureteral tortuosity and impassable urethral stricture led us to consider other therapeutic options.

Pharmacological chemolysis is nowadays an alternative for the management of uric acid lithiasis recognized by European guidelines. Although they do not establish clear indications as to when we should opt for this treatment, they do establish clear indications as to when we should opt for this treatment.

should opt for this treatment, they do recommend maintaining the patient's urinary pH at values of around 7 - 7.2 for it to be as effective as possible. They also emphasize close monitoring of urinary pH and modification of the treatment regimen according to these values. In the case of our patient, oral chemolysis was chosen, obtaining after 9 months an almost complete elimination of renal lithiasis, avoiding the need for invasive measures such as ureterorenoscopy.

5. Conclusions and recommendations

Anatomical renal anomalies are generally infrequent. Nevertheless, we should be familiar with them as well as with the possible treatment options in the context of ureteral or renal lithiasis. Surgical procedures can be difficult, with higher risks and with a lower lithiasis-free rate and a higher rate of reoperation. This is why we should consider the possibility of offering medical management (chemolysis) to this type of patient and especially to those with a high volume of lithiasis, either as definitive treatment of their lithiasis or as a step prior to surgery.

6. Bibliographical references (*of special interest, **of extraordinary interest)

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