Staghorn stone in megapolycalicosis in a child: Still the case for open surgery? Case report

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Abstract

We here report a rare case of congenital megapolycalicosis in a 14-year-old girl complicated by a 24-mm staghorn stone and numerous calculi at the level of all caliceal groups that had become symptomatic in recent weeks with malaise, hematuria, and urinary tract infection. Among the various therapeutic options, we opted for open surgery. The staghorn stone was removed by pyelotomy, and washout of the caliceal cavities released numerous microcalculi of 1.5−9 mm in size that were then removed. To our knowledge, this is the first case of pediatric megacapolycalicosis complicated by staghorn stone, which presents complex problems for the diagnosis and therapy.

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Introduction

Congenital Megapolycalicosis (CMP) is a rare renal malformation with an incidence of approximately 1/1,000,000 (ORPHA-93109). It is characterized by nonobstructive dilation of the renal calyces as well as an increase in the number of calyces (10-20), with a normal renal pelvis, ureter, and bladder.[1-3] It is thought to occur as a result of abnormal development of the renal medulla, which leads to hypoplastic renal pyramids and blunted dilated calyces. It is a nonobstructive dilatation of the calyces that tends to result in Urinary Tract Infections (UTI), and it predisposes to stone formation due to stagnant urine.[4] CMP should always be considered in the differential diagnosis of congenital hydronephrosis, polycalycosis, and infundibular stenosis.[5] We here report a pediatric case of CMP that had a preliminary diagnosis of idiopathic renal urolithiasis.

Case Report

In January 2020, a 14-year-old girl was admitted to our clinic with a history of urinary febrile infection, hematuria, high inflammatory parameters, and malaise.

The patient was in good health until 10 years of age. At 10 years of age, following a febrile urinary infection with hematuria, an Ultrasound (US) was performed, revealing mild dilatation of the right pelvis with diffuse microlithiasis that was treated with potassium citrate and magnesium.

In the year preceding the admission discussed here, the patient had started to experience episodes of feverish urinary tract infections, and she was referred to our Institution. Upon admission, the patient had a urinary febrile infection, hematuria, high inflammatory parameters, and malaise. She underwent a renal ultrasound and an Intravenous Pyelogram (IVP). The US revealed a right renal length of 157 mm (109 mm on the left side) and significant calyceal dilation of the kidney that was out of proportion compared to the pelvic dilatation and occupied by a large stone and diffuse urolithiasis. The IVP revealed a 25-mm stone in the pelvis, as well as the presence of multiple diffuse stones on all the renal calyces, excluding Ureteropelvic Junction (UPJ) obstruction. At admission, we scheduled US and a standard abdominal radiograph, which confirmed the diagnosis (Figure 1). MRI demonstrated a condition of megapolycalicosis, showing approximately 13 calyces and a cluster of microcalculi stacked in the major renal calyxes behind the staghorn stone (Figure 2).

Voiding Cystourethrogram (VCUG) revealed an absence of Vesicoureteral Reflux (VUR). A 99mTc-labeled Mercaptoacetyl-

triglycine (MAG3) renal scan augmented with furosemide revealed a differential function of 47% for the right kidney and 53% for the left, without obstruction. Assessment of the urinary electrolytes did not indicate hypercalciuria, hyperuricemia, or hyperoxaluria.

Basing on the clinical condition and the multitude of stones stacked behind the staghorn stone, with the parent's consent, we opted for open surgery.

A preliminary evaluation by cystoscopy was performed with a right retrograde pyelography and the application of a 4.8Ch, 22cm, double-J stent. The ureter appeared to have a normal caliber and shape, and insertion of a double-J stent was easy. The kidney was then exposed by a lumbotomy approach, the pelvis was opened with a transverse incision and the stone, which had adhered tightly to the walls of the pelvis, was extracted. It turned out to be an irregularly shaped staghorn stone of approximately 25x30 mm with squat offshoots extending towards the calices (Figure 3). The caliceal cavities were washed several time with saline solution, which resulted in the removal of more than 20 stones ranged from 1.5 to 9mm.

At the end of the procedure, the pelvis was reconstructed and a drain was placed in the retroperitoneum. Antimicrobial coverage of amoxicillin-clavulanate was used, while the drain was removed on third postoperative day. The patient was discharged on the fourth postoperative day, with hydroponic therapy and potassium citrate and magnesium therapy as well as a diet of low oxalate intake. The double-J stent was removed after three weeks. Upon examination, the stones were found to consist of calcium oxalate dihydrate.

The follow-up was set up as follows: hydroponic therapy with pyridoxine support, six-monthly ultrasound and clinical controls of renal function with six-monthly urine culture, and annual MAG3 scintigraphy. At present, the patient is asymptomatic and postoperative Mag3 scintigraphy showed no obstruction.[6]

Discussion

CMP was first described by Puigvert in 1963.[7] Approximately 100 cases with unilateral or bilateral megacalyces have been described in the literature to date.[8] It is primarily a disease of the renal calyces, and it is usually diagnosed because of its complications, such as calculi or infections of the urinary tract. CMP can be also diagnosed prenatally. In absence of complications, the disease is usually discovered incidentally during the urologic examination undertaken for different reasons. The anomaly is found predomi-

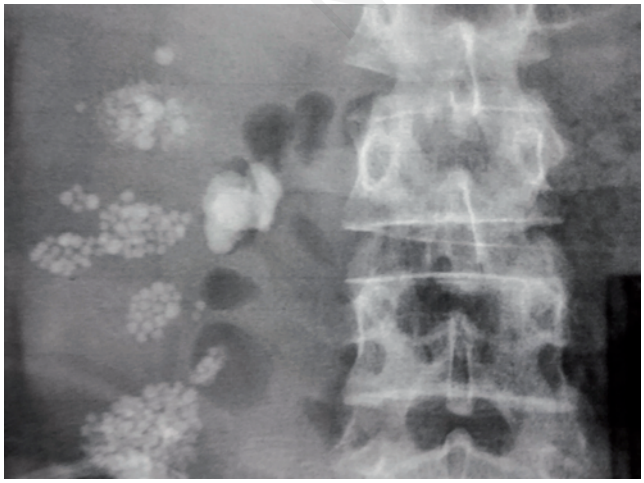

Figure 1. RX abdominal plane. A staghorn stone and a multitude of calculi occupying the entire surface of the kidney.

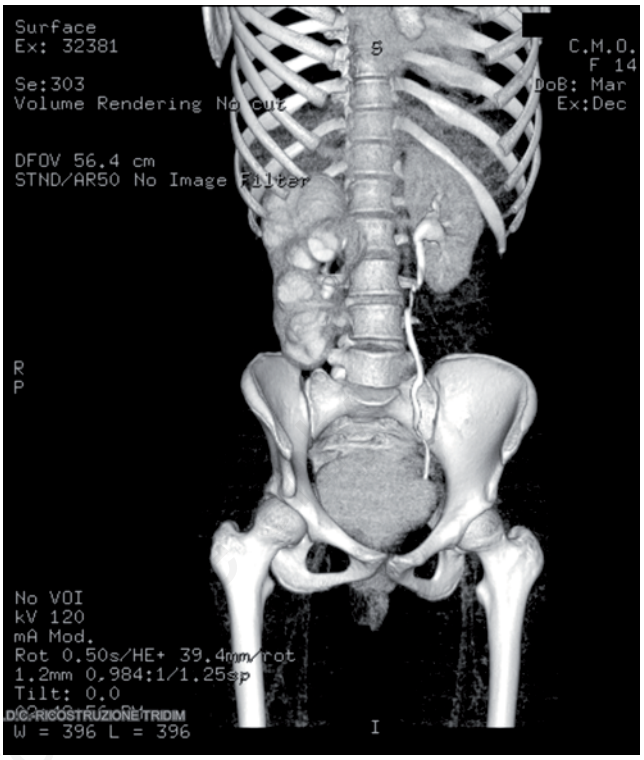

Figure 2. MRI, showing gross dilatation of all right renal calyces, faceting, an increase in the number of calyces, and a normal renal pelvis.

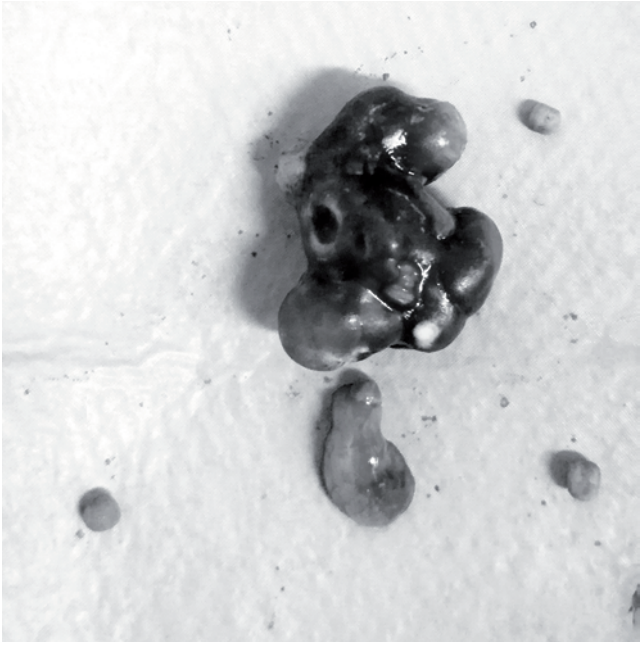

Figure 3. The stone with its stubby calyceal offshoots and numerous calculi of 1.2-1.3 mm in size.

nantly in male patients, with a male to female ratio of 6:1, primarily in Caucasians, and it is rarely found in association with an ipsilateral primary megaureter, which usually occurs independently. To date, only twelve cases have been described, with a male to female ratio of 5:1 and a left- to right-sided ratio of 3:1.[9] CMP has been also reported in African-American women, Egyptian children, and adult Chinese women.[10-13]

Clinically, CMP is characterized by enlarged kidneys and uniform dilatation of all the calyces. The infundibula, pelvis, and ureter are normal, and there is no obstruction in the collecting system. In addition to dilatation, there is also an increase in the number of calyces (polycalycosis) compared to the normal condition, from typically 7–9 till to 20–25.[14] There is a greater lithogenic predisposition in these patients. There is no apparent progression of the anatomical anomaly or a decrease in the function of the affected kidney over time.[15,16] Abdominal radiography combined with ultrasonography may be able to diagnose most clinically significant stones, and MRI can be considered the gold standard for the diagnosis.[17,18] The peculiarity of our case lies in the fact that, despite presenting with a large number of stones, the patient remained asymptomatic, only exhibiting significant clinical symptoms in the last year with microscopic/macroscopic hematuria, urinary infection, and malaise.

Surgery is not necessary for the treatment of a primary anomalous kidney with CMP, although the presence of a large stone or infection mandates appropriate therapy.[19] The treatment options can range from simple follow-up to Extracorporeal Shockwave Lithotripsy (ESWL), Percutaneous Nephrolithotomy (PCNL), minimally invasive (laparoscopy/robot-assisted) or open surgery.[20] The choice of best practice for removing kidney stones is still a matter of debate in children. Sultan *et al.* recently tried to standardize the approach to pediatric urolithiasis.[21] They highlighted the advantages and the disadvantages of various minimally invasive surgical options such as ESWL, Retrograde Intrarenal Surgery (RIRS), PCNL, laparoscopy and robotic surgery. In their algorithm, the authors recommend the open technique only in selected cases. Alivizatos and Skolarikos state that open stone surgery should be avoided in most cases but should be considered for patients for whom a reasonable number of less invasive procedures would not be useful.[22] Indeed, we excluded ESWL since it is not indicated with stones >2 cm, in case of complex stones, or with a staghorn stone, and we also ruled out PCNL due to the frequent need for double access and a higher risk of bleeding. The presence of numerous calculi stacked behind the main stone was decisive in the choice of open surgery instead of laparoscopic treatment to avoid the risk of loose stones in the abdomen.

Thus, our choice of open surgery was based more on the clinical situation than on well-defined guidelines and, in our opinion, proper selection of these patients is crucial in order to obtain the most favorable surgical outcomes.[23]

Conclusions

Congenital megapolycalicosis is a rare developmental anomaly of the kidney characterized by nonobstructive dilatation of the renal calyces. This nonobstructive dilatation of the calyces can cause urinary tract infection and stone formation due to stagnant urine. Only rarely does CMP occur in the pediatric population along with staghorn stone. The choice of the most effective treatment for such stones is still a matter of debate, although the use of minimally invasive techniques appears to be generally effective. Open surgery should be reserved for selected cases only.

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