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Case Report

A rare occurrence of primary obstructive megaureter with secondary ureteric stones: A case report

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ABSTRACT

Background: Primary obstructed megaureter (POM) is a congenital functional obstruction caused by an adynamic distal ureteric segment. Although it commonly presents with urinary tract infection, flank pain, or impaired renal function, its association with urolithiasis is rare in children and may cause diagnostic difficulty.

Case Presentation: We report a 10-year-old boy who presented with intermittent left flank pain for one year. Imaging revealed a unilateral left obstructed megaureter with multiple mobile ureteric stones and a renal calculus, along with a contralateral renal duplex system. Ultrasonography demonstrated hydroureteronephrosis, while micturating cystourethrogram excluded vesicoureteric reflux. MAG-3 renography showed impaired drainage and reduced differential renal function. Intravenous urography confirmed distal functional obstruction consistent with POM rather than obstruction secondary to stones. Surgical management included excision of the adynamic distal ureteric segment, removal of all stones, and ureteric reimplantation using an extravesical (modified Lich–Gregoir) technique. Histopathology confirmed the diagnosis. The postoperative course was uneventful.

Conclusion: POM complicated by urolithiasis in children is rare and can be misdiagnosed as secondary hydroureteronephrosis. A high index of suspicion and detailed evaluation are required. IVP and CT urography are crucial for diagnosis, and surgical management is essential to prevent recurrent infection and renal impairment.

Keywords: Primary obstructed megaureter, Urolithiasis, Pediatric ureteric obstruction, Intravenous urography.

INTRODUCTION

Primary obstructed megaureter (POM) results from an aperistaltic segment with narrowing of the distal ureter [1]. It occurs predominantly in male children and is usually unilateral; however, 15%–25% of cases are bilateral. Ultrasound of the kidneys, ureters, and bladder (KUB) is the initial investigation to detect ureteral dilatation, while intravenous urography (IVU), CT urography, and MAG-3 diuretic renography are definitive diagnostic modalities. A micturating cystourethrogram (MCUG) is always performed initially to exclude vesicoureteral reflux (VUR). Surgical intervention is indicated in complicated cases such as urolithiasis, renal insufficiency, recurrent urinary tract infections, and in symptomatic patients [2,3]. We present a rare case of left primary obstructed megaureter with multiple secondary mobile ureteric stones. The adynamic ureteral segment was resected, all stones were removed, and the ureter was reimplanted using an extravesical technique.

CASE REPORT

A 10-year-old boy presented with intermittent left flank pain for the past one year, which was relieved by paracetamol. There was no history of voiding difficulty or fever. He had no relevant medical or surgical history. Physical examination was unremarkable, with no palpable abdominal mass. He was afebrile, with a pulse of 85/min, blood pressure of 120/80 mmHg, and weight of 35 kg.

Laboratory investigations revealed hemoglobin of 12.4 g/dL, white blood cell count of 7,500/µL, C-reactive

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protein (CRP) of 10 mg/L, urea 26 mg/L, creatinine 0.5 mg/L, sodium 141 mEq/L, and potassium 4.02 mEq/L. Urine complete examination showed pH 5, glucose negative, protein negative, leukocytes 320,000/mL, red blood cells (RBCs) 10,000/mL, and no crystals. Urine culture showed no growth.

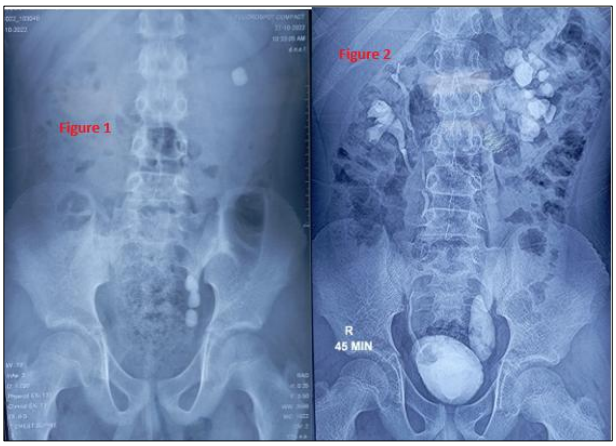

Figure 1: X-ray KUB showed three rounded calculi in line of lower ureter and one calculus at left renal area.
Figure 2: The right kidney showed normal uptake and excretion with a duplex system.

An X-ray KUB demonstrated three rounded calculi aligned in the lower ureter and one calculus in the left renal area (Fig. 1). Ultrasonography of the KUB revealed a normal right kidney measuring 9.7 cm and a left kidney measuring 10.2 cm with moderate hydronephrosis and a dilated ureter up to the distal end (diameter 2.0 cm). Three calculi were identified in the left lower ureter measuring 1.0 cm, 1.2 cm, and 1.7 cm, along with a 1.3 cm calculus in the mid-pole of the left kidney.

Figure 3: Narrowed segment measuring approximately 1.5–2.0 cm was excised.

The micturating cystourethrogram showed a normal urethra with no evidence of vesicoureteral reflux (VUR) on either side. A MAG-3 scan revealed left renal function of 41% with delayed and obstructed drainage, while the right kidney showed 59% function with normal drainage. Intravenous urography (IVU) demonstrated normal uptake and excretion of contrast from the left kidney, with a dilated and tortuous ureter containing three stones in its lower third. The right kidney showed normal uptake and excretion with a duplex system (Fig. 2).

A diagnosis of primary obstructed megaureter (POM) with multiple secondary ureteric stones was made. Surgical intervention was indicated due to symptoms, associated complications (secondary stones), and decreasing differential renal function. Under general anesthesia with endotracheal intubation, the patient was positioned supine and draped. Foley catheterization was performed, with catheter control maintained in the surgical field to allow bladder filling later.

Figure 4: Four stones were milked distally and removed.

The ureter was approached through an extraperitoneal Gibson incision, and proximal control was achieved using a sling to prevent upward migration of stones. The renal stone was mobile and was found in the lower ureter at the time of surgery. The ureter was transected close to the vesicoureteric junction (VUJ), and a narrowed segment measuring approximately 1.5–2.0 cm was excised (Fig. 3). Histopathological examination of the specimen confirmed the diagnosis of POM.

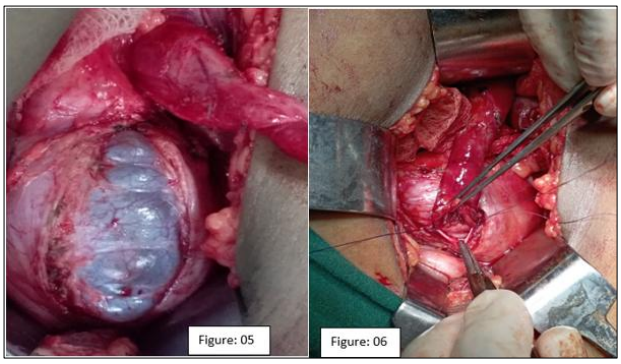

Figure 5: The detrusor muscle was incised over a length of 6–7 cm while preserving the mucosa.
Figure 6: The bladder mucosa and ureter were anastomosed using 5-0 polyglactin sutures.

The remaining distal ureter was dilated (approximately 2.0 cm in diameter); therefore, all four stones were milked distally and removed (Fig. 4). The ureter was reimplanted using an extravesical technique (modified Lich–Gregoir) over a double-J stent. The bladder was filled, and the detrusor muscle was incised over a length of 6–7 cm while preserving the mucosa (Fig. 5). The

bladder mucosa and ureter were anastomosed using 5-0 polyglactin sutures (Fig. 6). The detrusor muscle was then closed over the ureter, burying a ureteric length of 6–7 cm to create an adequate submucosal tunnel (Figs. 7 and 8).

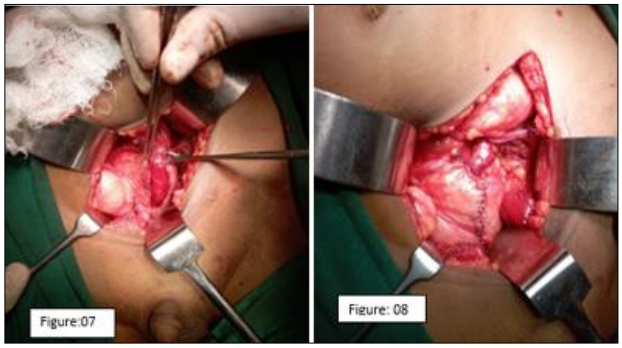

Figure 7 & 8: The detrusor muscle was then closed over the ureter, burying a ureteric length of 6–7 cm to create an adequate submucosal tunnel.

DISCUSSION

In our case, the child had a unilateral primary obstructed megaureter with multiple secondary ureteric stones and a contralateral renal duplex system. The British Association of Paediatric Urologists (BAPU) defines a ureteric diameter greater than 7 mm as abnormal [1]. Primary obstructed megaureter (POM) is characterized by a structural abnormality of the distal ureteric musculature, resulting in an adynamic ureteric segment and subsequent functional obstruction. POM associated with multiple secondary stones is uncommon in children and is extremely rare in the presence of a contralateral renal duplex system.

Children with POM usually present with recurrent urinary tract infections, flank pain, or impaired renal function, while presentation with urolithiasis is rare [3,4]. The diagnostic criteria for POM include a dilated ureter with smooth tapering of its distal end, absence of vesicoureteric reflux (VUR), absence of bladder outlet obstruction, and absence of secondary causes of lower ureteric obstruction such as stones or strictures at the vesicoureteric junction (VUJ) [4–5].

Stone formation in POM is mainly attributed to urinary stasis [6] and infection [7], while metabolic causes are rare [8]. The most common stone composition is calcium oxalate (59%), followed by calcium phosphate (10%); other types include uric acid, struvite, and cystine stones [9]. In patients presenting with multiple calculi in the distal ureter, it is often difficult to differentiate whether the megaureter is secondary to impacted ureteric calculi or represents primary obstructed megaureter. In such cases, the diagnosis of POM as the underlying cause of multiple ureteric calculi may be suggested by intravenous urography (IVU) and confirmed by histopathological examination of the surgical specimen.

In our patient, both ureteric and renal calculi were present. Initially, four stones were identified in the lower ureter; however, these stones were mobile and changed position over time, as demonstrated on serial X-ray KUB (Fig. 1) and ultrasonography. The unusual configuration, mobility of the stones, and the presence of a dilated, tortuous ureter raised suspicion of POM. An intravenous urogram was therefore performed, which established the diagnosis by demonstrating that the obstruction was distal to the stones rather than caused by the stones themselves. Typical IVU findings include partial or complete dilatation of the ureter with narrowing at its distal end near the ureterovesical junction. Stones may also be seen within the dilated ureter, renal pelvis, or calyces.

When the affected kidney is nonfunctional, IVU may fail to demonstrate the obstructed ureter due to non-opacification. In such situations, the diagnosis of POM may be missed preoperatively or suspected only during surgery when a narrowed distal ureteric segment is identified. CT urography is a more useful diagnostic modality in this scenario. Histopathological examination in our case revealed an adynamic, narrowed distal ureter. Microscopic analysis showed increased myocyte apoptosis, along with a reduced number of interstitial cells of Cajal-like cells and decreased smooth muscle content, confirming POM as the underlying pathology.

If POM is suspected on IVU or CT urography, a micturating cystourethrogram must be performed to exclude vesicoureteric reflux and bladder outlet obstruction [10,11]. Cystourethroscopy can also be helpful in evaluating the urethra and bladder and in excluding secondary causes of obstruction.

Kumar A. reported a case of a 14-year-old boy with bilateral primary obstructed megaureter associated with secondary stones [12]. Delakas D. reported a case of a giant ureteral stone with primary megaureter in a 20-year-old female who presented with an acute abdomen [13]. Only a few cases have been reported in children in the literature, with most cases of urolithiasis associated with primary obstructed megaureter reported in adults.

In children, primary obstructed megaureter complicated by urolithiasis can easily be misdiagnosed as hydroureteronephrosis secondary to ureteric calculi. Therefore, a high index of suspicion and detailed evaluation are required, as these conditions share similar clinical and radiological features. Although routine imaging may not reliably differentiate the two entities, intravenous pyelography (IVP) and CT urography are crucial diagnostic tools for identifying POM. Micturating cystourethrogram and cystourethroscopy are necessary to exclude bladder outlet obstruction and other secondary causes of obstruction. Urolithiasis associated with POM in children requires surgical management to prevent complications

such as recurrent urinary tract infections and progressive renal function impairment.

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