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Bilateral Primary Obstructive Megaureter with Concurrent Kidney and Ureteral Stones: A Rare Case

Dr Akash Kumar Gupta[1*], Dr Pramod Kr Sharma[2], Dr Soumendranath Mandal[3], Dr Anirban Bhunia[4]

[1]Post Doctoral Trainee, Department of Urology, Calcutta National Medical College & Hospital
[2]Associate Professor, Department of Urology, Calcutta National Medical College & Hospital
[3]Hod & Professor, Department of Urology, Calcutta National Medical College & Hospital
[4]Post Doctoral Trainee, Department of Urology, Calcutta National Medical College & Hospital

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***Corresponding author:** Dr Akash Kumar Gupta
Post Doctoral Trainee, Department of Urology, Calcutta National Medical College & Hospital

Abstract | **Case Report**

Primary obstructive megaureter (POM) is a rare congenital anomaly in adults, typically involving functional obstruction at the distal ureteral segment without an identifiable mechanical cause. Adult presentation is uncommon, and bilateral involvement with concurrent renal and ureteral stones is exceedingly rare. We report the case of a 33-year-old male who presented with bilateral flank pain, recurrent graveluria, and intermittent fever. Imaging revealed bilateral hydronephrosis, dilated distal ureters, and multiple calculi in both kidneys and ureters. Exclusion of reflux or bladder outlet obstruction confirmed the diagnosis of bilateral POM. The patient underwent staged open surgical interventions, including ureterolithotomy, distal ureteral excision, tapering, extravesical ureteral reimplantation using the modified Lich-Gregoir technique, and pyelolithotomy. A total of 133 smooth calculi were retrieved from both ureters and kidneys. Histopathological examination confirmed POM by demonstrating absence of smooth muscle in the resected ureteral segments. This case highlights the diagnostic and therapeutic challenges in managing adult bilateral POM with extensive stone burden. It underscores the importance of considering congenital anomalies in adults with unexplained ureteral dilation and recurrent urolithiasis, and emphasizes the need for timely surgical intervention to preserve renal function and prevent recurrence.

Keywords: Primary obstructive megaureter, urolithiasis, nephrolithiasis, ureterolithiasis, adult congenital anomaly.

INTRODUCTION

Primary obstructive megaureter was described for the first time by Caulk.[1] It is common in children but its presentation as a primary anomaly is rare in adults.[2] The cause of primary obstructed megaureter is the aperistaltic, narrowed prevesical portion of the ureter. Megaureter may be classified as: obstructive, refluxing, non-obstructing & non refluxing and obstructing & refluxing. It can be primary or secondary. They may be asymptomatic or present with flank pain, recurrent urinary tract infection (UTI), and hematuria in symptomatic situations. We present a very rare case report of a patient with bilateral megaureter with concurrent kidney and ureteral stone.

CASE PRESENTATION

33-year-old male presented to us with bilateral flank pain, more pronounced on the right side, for 9 months. His symptoms included recurrent episodes of graveluria and intermittent fever for which he was treated at local hospital. He had no history of other urinary or bowel symptoms, and no comorbidities or surgeries. Physical examination was normal with body mass index of 23 kg/m2. Investigations like CBC, renal function test, electrolytes, coagulation, and LFT were normal. Urine analysis showed 03 RBC, 22 WBC and no growth on urine culture. USG findings revealed bilateral renal and ureteral stones, with gross HDUN bilaterally. CECT KUB showed B/L HDN with multiple calculi in both kidneys with RK – largest 2.1*1.7 cm in pelvis. LK- multiple calculi in clusters in lower calyx. B/L ureters were dilated, most prominently affecting distal ureters with multiple calculi in both distal ureters (figure 2). Dig MCU- no reflux was noted (figure 3). Uroflowmetry was normal with max flow rate of 18ml/sec. The patient underwent cystoscopy that revealed normal bladder with no features suggestive of bladder outlet obstruction.

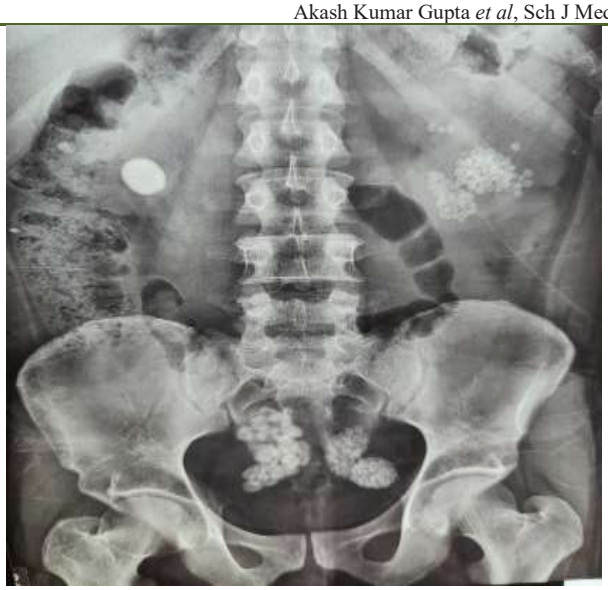

Figure 1: DIG XRAY showing B/L renal and ureteric stones

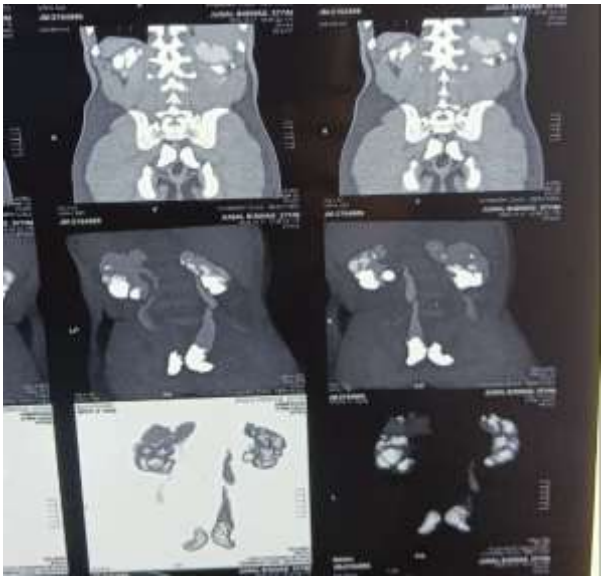

Figure 2: CT UROGRAM showing B/L HDUN with calculus in both kidneys and ureter

Figure 3: DIG MCU showing no reflux

MANAGEMENT

Patient underwent right open ureterolithotomy + excision of stenosed segment + distal ureteral tapering with extravesical reimplantation (modified lich-Gregor) over a DJ stent first (figure 4). On the right side rigid Dresdan 20.8 Fr nephroscope was introduced through the ureter, which revealed multiple small stones in pelvis which was removed with forceps. The large 2cm stone could not be retrieved as it was not amenable to take it out via PUJ (figure 5). Total of 67 small, round and smooth stones were removed from right ureter and kidney (figure 7). On the left side similar procedure was done with an additional pyelolithotomy was done for stone clearance as stone was mobile and not seen in the ureter during the procedure (figure 6). Flexible Uretero renoscope showed the stones in kidney. Total of 66 small, round, smooth surface stones were removed from left kidney (figure 8). Right DJ stent was removed after 4 weeks. Post op recovery was uneventful for both the procedures. At present patient is having right renal calculus which is planned for right PCNL at a later date. HPE of excised distal ureteric segment was suggestive of primary megaureter as there was absence of smooth muscle.

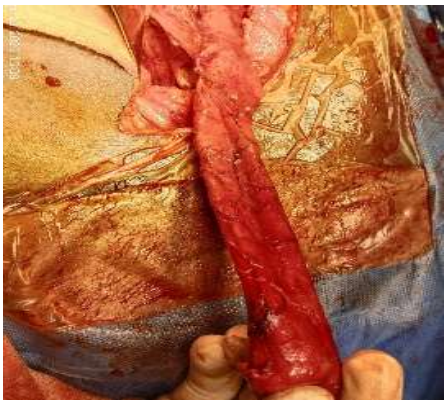

Figure 4: Intra op finding of RT megaureter after dismembering

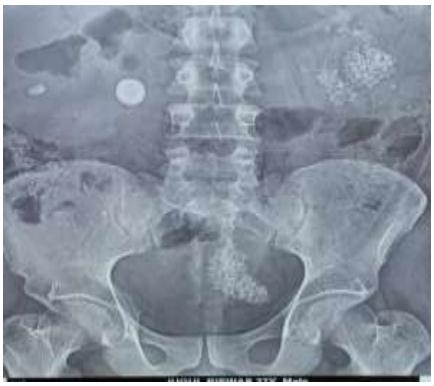

Figure 5: Post op DIG XRAY KUB showing residual calculus in RT kidney

Figure 6: Intra op finding of LT megaureter

Figure 7: Stone retrieved from both left and right ureter and kidney

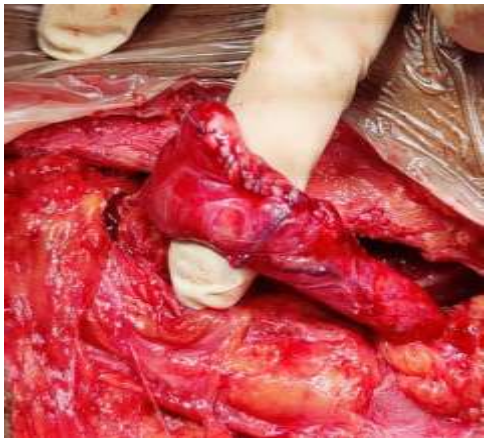

Figure 8: Intra op picture Post left reimplantation

DISCUSSION

The normal diameter of the ureter is typically less than 5 mm. When the ureteral width exceeds 7 mm, it is generally classified as a megaureter. Among the various subtypes, primary obstructive megaureter (POM) represents a congenital functional obstruction, most often located at the distal ureter, without any evident mechanical cause. While POM is usually identified during infancy or early childhood, some cases remain undiagnosed until adulthood. This delay in detection occurs because the congenital obstruction may not produce significant symptoms during the early years of life. Although spontaneous resolution may not occur in these cases, affected individuals often remain clinically asymptomatic throughout childhood and adolescence. In adulthood, POM can become symptomatic, typically presenting with urinary tract infections (UTIs), renal functional impairment, or recurrent urolithiasis. Stone formation is often attributed to urinary stasis in the dilated ureter, which creates an environment conducive to crystal aggregation. In the most extensive published series to date involving 55 adolescents and adults with symptomatic POM, Hemal *et al*. reported that 36% (20 patients) had associated urinary tract calculi. Most of these were located within the ureter, while only three patients (approximately 5%) had renal calculi without concomitant distal ureteral calcifications.[3] This finding emphasizes the role of the anatomical abnormality in stone pathogenesis. Remarkably, in rare instances, massive stone burden can occur within the ureter itself. A case documented by Delakas *et al*. described an adult with POM who developed a 12-cm stone confined to the dilated ureter, highlighting the extent to which urinary stasis can contribute to extreme calcification.[4] Beyond stone formation, giant forms of megaureter may lead to gross abdominal distension, occasionally mimicking a cystic mass on physical examination or imaging.[5] Additionally, POM may coexist with other congenital anomalies such as:

- Unilateral renal agenesis
- Duplicated collecting systems
- Ectopic kidneys
- Contralateral multicystic or dysplastic kidneys
- Horseshoe kidneys
- Hirschsprung disease. [6]

Although rare, complications such as renal rupture due to backpressure from longstanding obstruction have also been reported in the context of undiagnosed or neglected POM [7].

CONCLUSION

Congenital megaureter should remain an important diagnostic consideration for urologists when evaluating cases of distal ureteral dilation, especially in the absence of a clear etiology. In adults, undiagnosed primary obstructive megaureter (POM) may lead to

progressive complications and often requires definitive surgical management to prevent recurrence of symptoms and long-term morbidity. In this case report, we present a highly unusual instance of bilateral concurrent renal and ureteral calculi in an adult patient with POM—an exceedingly rare clinical scenario. We detail the diagnostic approach, intraoperative findings, and our experience with the surgical management of this complex and challenging condition.

Ethical Approval: N/A

Consent For Publication: Appropriate informed consent was taken from patient as per our institutional protocol.

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