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Case Report

Congenital Megaureter in an Adult: A Rare Presentation

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ABSTRACT

Introduction: Congenital megaureter is a condition usually diagnosed in neonates and children; its primary presentation in adults is rare. Spontaneous resolution occurs in over half of all affected children, and this is hypothesized to occur as a result of the ureterovesical junction (UVJ) maturation and growth. When this condition does present in adults, it typically does so during the third or fourth decade, and unilateral disease, most often left-sided is more common than bilateral disease.
Presentation of Case: A forty year old male patient presented with recurrent attacks of left ureteric colic since 6 months. On examination, abdomen was soft and non-tender. An ultrasound of the abdomen revealed grossly dilated left ureter with 5 mobile calculi each of approximately 1.5 centimetre size and left hydronephrosis. Plain radiograph of the abdomen revealed multiple calculi in the lower end of the left ureter. Intravenous pyelography study demonstrated dilated left ureter abruptly ending just proximal to the ureterovesical junction with 5 mobile calculi and a hydronephrotic left kidney. Patient was posted for surgery and a reduction left ureteroplasty with removal of the calculi and re-implantation of the ureter by Politano-Leadbettter technique with left sided DJ stenting was done. Postoperative period was uneventful and the patient was discharged in 7 days.
Conclusion: Primary presentation of congenital megaureter in adults is rare. It is expected that leaving the underlying abnormality intact may pose the patient to the recurrent stone formation and persistence of symptoms.

Keywords: Megaureter, Ureteroplasty, Politano-Leadbetter technique.

INTRODUCTION

Initially described by Caulk in 1923, congenital megaureter is a condition usually diagnosed in neonates and children; its primary presentation in adults is rare. [1] Spontaneous resolution occurs in over half of all affected children, and this is hypothesized to occur as a result of the ureterovesical junction (UVJ) maturation and growth. [2–4] Diagnostic criteria include: dilated ureter, absence of vesicoureteral reflux, absence of infravesical obstruction and absence of distal ureteral obstruction. [5] When this condition does present in adults, it typically does so during the third or fourth decade, and unilateral disease, most often left-sided is more common than bilateral disease. [6,7]

PRESENTATION OF CASE

A forty year old male patient presented with recurrent attacks of left sided flank pain since 6 months, colicky type and radiating to the groin. He had no complaints of fever, haematuria or burning micturition. He was a known hypertensive on medication for the same. On examination, abdomen was soft and non-tender. Urine examination revealed 3-4 RBCs and 2-3 WBCs /hpf. An ultrasound of the abdomen revealed grossly dilated left ureter with 5 mobile calculi each of approximately 1.5 centimetre (cm) in size and left hydronephrosis. Plain radiograph of the abdomen revealed multiple calculi in the lower end of the left ureter (Figure 1). Intravenous pyelography (IVP) study demonstrated dilated left ureter abruptly ending just proximal to the ureterovesical junction with 5 mobile calculi and a hydronephrotic left kidney (Figure 2). Patient was posted for surgery and a reduction left ureteroplasty with removal of the calculi and re-implantation of the ureter by Politano-Leadbettter technique with left sided DJ stenting was done. Postoperative period was uneventful and the patient was discharged in 7 days. An IVP done 9 months postoperatively revealed a normal sized left ureter with no calculi, mild residual left hydronephrosis and no reflux on straining (Figure 3).

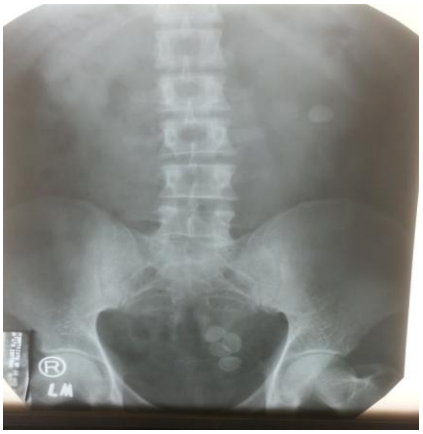

Figure 1: Plain radiograph of KUB region showing left lower ureteric calculi.

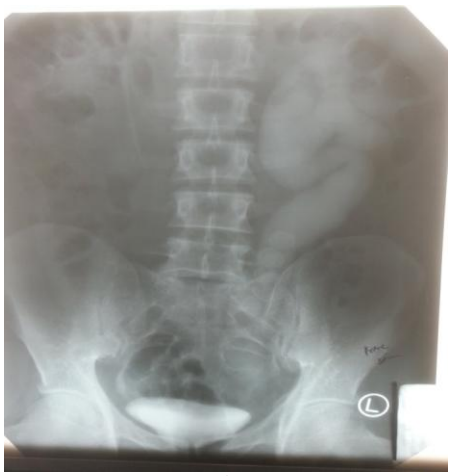

Figure 2: IVP showing left dilated ureter with left hydronephrosis and multiple left lower ureteric calculi.

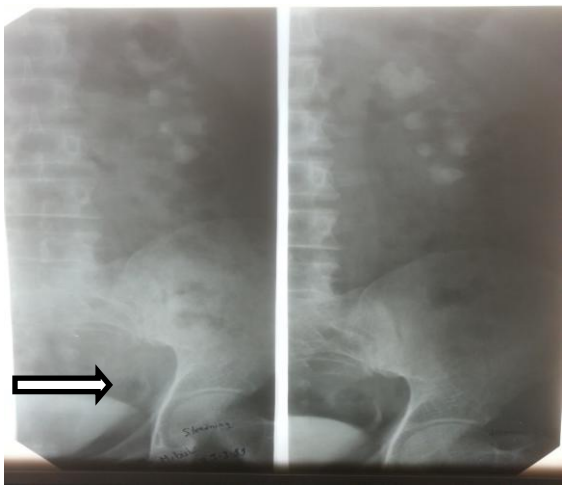

Figure 3: Postoperative IVP showing normal size of the ureter after reduction ureteroplasty with mild residual hydronephrosis and no reflux on straining (as shown by the arrow).

DISCUSSION

The current belief is that primary obstructive megaureter (POM) presents primarily in adults when the congenital abnormality does not cause symptoms or illness and is not seen via an imaging study performed in children. Spontaneous regression fails to occur, yet patients remain asymptomatic through childhood and into their adult years. Eventual symptoms that may occur include: urinary tract infections, renal parenchymal damage and recurrent stone formation.

They may be asymptomatic or present with flank pain, recurrent urinary tract infection (UTI), and hematuria in the symptomatic situations. Pain is the most common presenting symptom. [8] In mild cases, there is a 2–3 cm fusiform lower ureteral dilation just proximal to the tapered extravesical distal segment. [9] With increasing severity, the ureter dilates more proximally and involves the collecting system. In extreme cases, there is significant hydronephrosis, and loss of the renal parenchyma leading to the impaired renal function. The characteristic criteria for the diagnosis of POM are; absence of vesicoureteral reflux (VUR), absence of obstruction in the infra-vesical area, and absence of secondary causes of lower ureteral obstruction. [10,11] The adult presentation of POM is in the third or fourth decades of life, and unilateral involvement is more common than bilateral disease. The condition is usually seen in the left side with the male predominance. [12,13] In a large reported series of 55 adults with symptomatic POM, Hemal et al identified 20 patients (36%) to have urinary tract stones. [14] The rate of synchronous renal–ureteral unit stones was 5.5%. [14] Associated anomalies can be found with POM such as ureteropelvic junction obstruction, horseshoe kidney, megacalicosis, megalourethra, megacystis, and contralateral renal atrophy. From these anomalies, contralateral renal atrophy was the most common, and was found in 9% of cases. [14] Most of the stones were located in the ureter; only 3 of the 55 patients (5%) had isolated renal calculi. Large stones can develop in the dilated portion of the ureter due to urinary stasis. Delakas et al. [15] described an adult patient with POM who developed a 12 cm isolated ureteral stone within the dilated portion of involved ureter. Extracorporeal Shock Wave Lithotripsy (ESWL), ureteroscopy, ureteral meatotomy and stenting, and percutaneous nephrostomy are non-invasive or less invasive techniques that sometimes are needed in the management of POM or POM related stones. [14]

Rosenblatt et al. [16] reported two cases of adult POM that presented with urolithiasis. Due to small stone sizes, they just treated the stones without surgical repair of the underlying megaureter.

CONCLUSION

Based on the best of our knowledge primary presentation of congenital megaureter in adults is a rare occurrence. Our case presented with multiple mobile calculi with a dilated left ureter and gross left hydronephrosis for which surgical management was a must. It is expected that leaving the underlying abnormality intact may pose the patient to the recurrent stone formation and persistence of symptoms.

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