CASE REPORT

Complicated Megaureter with Aganglionosis in An Adult Lady

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ABSTRACT

Dilated ureters are often due to mechanical obstruction. This case report describes a young adult lady with primary megaureter due to aganglionosis finally managed with extravesical Leech-Gregor Uretroneocystostomy.

Key words: *Megaureter. Aganglionosis. Uretroneocystostomy. Urotithiasis.*

INTRODUCTION

Common application of maternal-fetal ultrasound allows the diagnosis of many urological anomalies before birth including megaureter.[1] Usually it is a silent condition but may manifest clinically in infancy with infection, lumbar pain or hematuria. Rarely it present in adults with complications like recurrent infections, nephrolithiasis (46%) and renal failure (13.5%).[2] Here we report a case of primary megaureter with bilateral renal stones and renal failure in adult age group.

CASE REPORT

A 22 years old married female presented to us with bilateral lumber pain, fever and anorexia for last one year. Her past history revealed bilateral renal stones, operated few years back in a community hospital. Laboratory reports showed impaired renal functions with serum creatinine of 4.6 mg/dl. Ultrasound showed hydroureteronephrosis with residual stones in both kidneys. Renal cortex on right side was completely lost while left renal cortex was preserved. X-ray KUB confirmed residual stones on either side. Urine culture was positive for *Pseudomonas*, sensitive to *Quinolone* and *Nitofurantoin*.

Her cystoscopy and retrograde pyelography (RPG) was preformed which showed normal ureteral orifices. RPG showed grossly dilated and tortuous left ureter with pin-hole ureteral stricture about 2 cm above UV junction (Figure 1). Bilateral DJ stents were placed with much difficulty. Follow-up radiograph showed migrated DJ on left side with its both ends coiled in left lower ureter. Another DJ stent was placed with the help of ureterorenoscope. After placement of stents her renal functions normalized in 2 weeks. A MAG3 renogram after 4 weeks showed 80% contribution to renal function by the left kidney and markedly reduced function of the right kidney.

On the basis of radio-isotope scan and RPG findings, resection of left lower ureter and uretero-neocystostomy was planned. A transperitoneal lower midline approach showed massively dilated left ureter whose diameter was greater than that of the small bowel. There was ureteral narrowing and obstruction few centimeters above UV junction, from which double J stent was merely passing (Figure 2). Lower end of ureter was excised with cuff of healthy tissue (Figure 3). An excisional tapering technique was performed in about 5-6 cm of lower ureter and extravesical Leech-Gregoer Ureteroneocystostomy was performed over DJ stent.

Her postoperative recovery was uneventful. Drain was removed after 24 hours and urethral catheter was removed on 4th day. She was discharged on the 6th postoperative day. Biopsy of the lower ureter showed agangilionic segment with normal muscles fibers. After 3 weeks, shock wave lithotripsy was done for the left sided residual renal stones. Left double J stent was removed after 5 weeks when she was stone free on that side.

Her follow-up ultrasound at the 6th week and 3 month showed residual pelvicaliceal dilatation with normal renal functions.

Figure 1: Rerograe pyelography showing dilated left ureter with pinhole distal ureteral stricture.

Figure 2: Peroperative view of the dilated ureter.

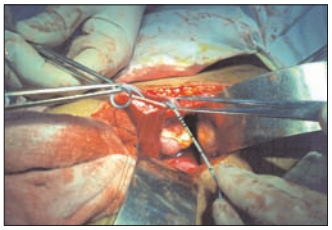

Figure 3: Excised uretral tissue.

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Received May 09, 2009; accepted March 25, 2010.

DISCUSSION

Megaureter is a non-specific term that simply means dilated ureter. It does not explain the underlying pathology.[1] Anomalous Wolffian duct and ureteral buds have credit for this anomaly.[3] With the advent of antenatal ultrasound an increased number of cases are identified prior to the onset of symptoms. In adults, treatment guidelines are not very clear as small number of cases are documented in literature.[4] Both conservative and surgical treatment options are practiced. Surgical treatment of adult megaureter is a better option when it presents with complications.[5] Since long its treatment algorithm includes excision of lower ureter and ureteroneocystostomy.[6] However, surgical approach, re-sizing of ureteral diameter and method of ureteral re-implantation vary and depend upon patient's age, the cause of megaureter and the skill of the surgeon.

In this case, extravesical approach was used along with excisional tapering,[6] of the hugely dilated left lower ureter. Extravesical ureteroneocystostomy[7] by Leech-Gregoer method was preferred over intravesical approach because it is simple, does not need excessive mobilization of ureter as well as excludes wide open cystostomy. Therefore, after extravesical approach urethral catheter can be removed with in few days. On the other hand longer length of ureter is needed in intravesical implant; bladder needs to be widely opened[3] and its repair compromises bladder capacity. Patient also needs prolonged indwelling urethral catheter that has its own complications.

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