

The Diagnosis and Treatment for Primary Obstructive Megaureter in Adult and Children- Three Cases Experience and Review of the Literatures

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Megaureter is a disease including variable anomalies. It may be caused by either a primary cause or secondary to specific pathologic origin. Primary obstructive megaureter is a subgroup of megaureter and an uncommon disease. Its etiology is unclear and the management remains controversial. Our report includes management of three cases in the most recent four years. The follow up period of these cases ranged from 10 months to 4 years. Their prognosis is good. (JTUA 20:173-7, 2009)

Key words: Megaureter.

INTRODUCTION

Megaureter is a term implying a group of anomalies associated with increased ureteral diameter over 5mm in size. According to the international classification at '70s, Smith and Stephens presented that megaureter may be obstructed, refluxing, both refluxing and obstructed, or unobstructed and not refluxing.¹ It is either from a primary ureteral intrinsic factor or secondary to specific pathologic processes such as bladder outlet obstruction, neurogenic bladder, polyuria, or infection. The megaureter is a common cause of obstructive uropathy among neonates and young children. Up to 23% of patients with upper urinary tract dilatation could be diagnosed by prenatal ultrasound. They are more common in boys than girls, a little more for the left side, and 25% in bilateral.²

Primary obstructive megaureter is an uncommon disease. The result of lower ureteral obstruction causes dilatation of the upper tract and impaired renal function. In adults, it is easily mistaken as lower third ureteral stone. In children, the management of this disease remains controversial. We collected three cases including one adult and two children in recent four years. One female adult with right megaureter and the two baby boys with left megaureter all received excisional tailoring and reimplantation of ureter.

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CASES REPORT

A 49 years old female had received right ureteroscopy and double J stenting in 2001 due to right renal colic, fever, and right hydronephrosis. Recurrent symptoms and signs were noted again in 2003. The other two baby boys who are 6 and 20 months old presented with UTI, fever and gastrointestinal symptoms. Pre-operative serum creatinine levels of the three cases were normal. Abdominal sonography, intravenous pyelography, and retrograde pyelography (Fig.1) disclosed right hydroureteronephrosis in the adult patient and left hydroureteronephrosis in the two baby boys. All the cases had examined by voiding cystourethrogram and they did not show any vesico-ureteral reflux.

Endoscopic examinations did not show any bladder outlet problem and the ureteral orifices of the hydro-nephrotic side were located at normal position with normal appearance. Diuretic renogram of the two baby boys were done and showed obstruction of urinary drainage at left side. Moderate parenchymal damage was noted in the 6 months old baby and there was no obvious parenchymal damage of the 20 months old baby boy. Under the impression of primary obstructive megaureter, the excisional tailoring of the distal ureter and reimplantation with Politano-Leadbetter method of these cases was performed. (Fig.2,3,4)

The pathological findings disclosed fibrosis and increased collagen deposition in all the cases. Intravenous pyelography followed within 6 months all showed mild to moderate residual hydronephrosis but the hydroureter is obviously improved. The post-operative serum crea-



Fig. 1. RP of 49y/o female before OP showed right hydronephrosis even right D-J stent had been.



Fig. 3. Excisional tailoring of the dilated ureter after the resection of the adynamic segment distal.



Fig. 2. Left megaureter of the 20m/o baby boy with an adynamic segment distal ureter of small.



Fig. 4. After the excisional tailoring, the ureter was ready for reimplantation.



Fig. 5. IVP of 49 y/o female one year after reimplantation showed resolved.

tinine level was normal. The adult case has been followed for four years and her intravenous pyelography (Fig.5) after one year follow up was unremarkable. The other two baby boys have been followed for nearly one year, and there is no more symptom and sign noted again. The outcomes of all our three cases are good.

DISCUSSION

In our cases, patients suffered from fever, flank pain, urinary tract infection, and gastrointestinal symptoms. The symptoms and signs were not specific except that gastrointestinal symptoms were only noted in the two baby boys. Gastrointestinal symptoms were also frequently noted in our pediatric patient of vesico-ureteral reflux and ureteropelvic obstruction even without any urinary symptoms. So that, physicians should bear in mind that obstructive uropathy maybe noted in pediatric patients with gastrointestinal symptoms or even failure to thrive accompany with mild urinary symptoms. Renal sonography was the best way for screening these patients.

Renal sonographies were first done for primary evaluation in these three cases and disclosed hydroureteronephrosis. The same findings were disclosed by intravenous pyelography and retrograde pyelography but the dilated ureters comparing with the renal pelvis and calyx were disproportional larger than other pictures of distal ureteral obstruction caused by stones, tumor, or any secondary cause. In addition to this, there was a segment of small caliber ureter distal to the dilated one and the diameter of different portions of megaureter were nearly the same. Therefore, primary obstructive megaureter was impressed. No vesico-ureteral reflux was found by voiding cystourethrogram. There was no obvious difference of imaging finding between the adult and pediatric patients. Finally, primary obstructive megaureter were strongly suspected, and all these cases underwent tailoring and reimplantation.

With the increasing prevalence of prenatal and neonatal ultrasound examination, many primary megaureter cases can be detected prenatally. When significant obstruction is present, there is a need for early surgical correction to preserve renal function.¹ However, the safety and efficacy of a non-surgical approach to asymptomatic patient has been confirmed by many authors in recent years. A study by Shukla et al found that complete resolution in 52.5% of asymptomatic patients at a mean of 2-9 years of follow up and improved or stable hydroureteronephrosis in 47.5% of non-operatively patients.³ Therefore, they suggested long term follow up of children with prenatally diagnosed primary megaureter with mild to moderate hydroureteronephrosis and they believed that ultrasound should be periodically continued until the child reaches adulthood at least in instances where complete resolution is not documented.

McLellan et al disclosed that retrovesical ureteral diameter less than 1.32 cm appears to be predictive of resolution.⁴ They also found that the presenting grade of hydronephrosis was an important predictor of the resolution rate. They concluded that increasing or severe hydronephrosis, decreasing renal function and retrovesical ureteral diameter greater than 1 cm may correlate with the need for surgical repair. In our cases, the prenatal data was not available and the patients had been suffered from different degree of symptoms. Grade 5 hydronephrosis and large retrovesical ureteral diameter (23mm in the female adult, 15mm in the 6 months old baby boy, and 16mm in the 20 months old boy) were noted. Thus we decided to have surgical repair in managing our cases.

Tubaro et al had reviewed 92 obstructive megaureter in children.⁵ They suggested that temporary urinary diversions may be successfully used in selected patients

with fever or bad kidney function. Moreover, ureteral tapering was usually unnecessary during subsequent reimplantation. Lee et al recommended two stages of reimplantation. The first stage ureteral reimplantation without anti-reflux procedure was done as a temporary treatment of obstructive megaureter in neonate and infant because there were technical limitations in the first year of life due to small bladder capacity.⁶ Their method relieved the obstruction while allowing the children to mature. By delaying the second stage definite surgical repair of the megaureter, there was potential for the ureter to decrease in diameter which made the subsequent reimplantation more feasible. In our experience, there is no difficulty in reimplantation of ureter, even in the first year of life.

In managing the ureteral tailoring of the distal ureter after resection of the small caliber adynamic segment, we chose the excisional tapering method. Fretz et al reported that Starr plication was reliable for primary obstructive megaureters in their study of 16 patients with average follow up for 6.2 years.⁷ It may not be suitable for a small bladder because of the increased bulk of the ureter with plication. The plication is preferred for a moderately dilated ureter, while excisional tapering is preferred for a more severely dilated or a markedly thickened one.²

The pathological examinations of our cases disclosed fibrosis and collagen deposition of the distal ureters and there was no difference between the adult and pediatric patients. There is general agreement that there is no true narrowing at the distal ureter, but a functional obstruction arising from an aperistaltic or an adynamic juxtavesical segment. Payabvash et al conducted a study which was the first showing increased myocyte apoptosis and decreased vascular and neural supply at the site of the congenital vesico-ureteral obstruction.⁸ In their series, the amount of collagen fibers was significantly increased after myocyte apoptosis in primary obstructive megaureter. In our cases, collagen deposition of the distal ureter were also noted.

Tc-99m MAG3 diuretic scintigraphy is recognized as a reliable method, even in children, in evaluating hydronephrosis. However, Belkis et al found that diuretic renography was unreliable in the neonatal period because of the low glomerular filtration level at that age, and a minimum age of 1 month was recommended for follow up by diuretic renography.⁹ In the study of long term outcome in 46 renal units, Link et al reported that male patients, those undergoing surgery at a younger age and patients with lower grades of preoperative hydronephrosis were most likely to exhibit improvement or resolution of hydronephrosis postoperatively.¹⁰

CONCLUSION

Primary obstructive megaureter is an uncommon disease. Most of the cases are now discovered prenatally and conservative management with long term follow up are appropriated. Indications for surgery included recurrent or breakthrough urinary tract infections, decreased renal functions, and increased degree of hydroureteronephrosis. The induction of apoptosis in normal functioning myocytes of distal ureter and substitution of fibrotic tissue may contribute to the pathogenesis of primary obstructive megaureter. In carefully selected patients, ureteral tailoring and reimplantation remains a highly successful treatment modality for primary obstructive megaureter.

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