A case of Undetected Congenital Primary Obstructed Megaureter

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Abstract

Primary obstructed Megaureter (POM) defined as dilatation of ureter bigger than the normal size with or without renal pelvis dilatation. POM was first described in 1923 by Clark [1]. This is the second most common aetiology causing hydronephrosis in infant and it more prevalence in boys compared with girls. Early detection of POM is important to guide us in the long management of the patient and better outcome. Surgical intervention still mainstay treatment for POM in failure of conservative management.

Keywords: Primary obstructed megaureter, hydronephrosis, surgery.

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Case:

A 20 years old female patient presented to us with complaints of right iliac fossa pain for one week. She described the pain was constant and pricking in nature, not relieved or aggravated by food intake and non-radiating. Patient also had nausea and vomiting during the pain attack, otherwise no fever, haematuria or sandy urine. On clinical examinations noted mild tenderness over right iliac fossa, no palpable mass and her urogenital examination was normal. Renal function test and x-ray KUB showed no significant findings. Patient underwent ultrasonography of the urinary tract and showed left gross hydronephrosis (13.6cm with parenchymal thickness of 1cm) with no evidence of distal obstruction. In intravenous urethrogram (IVU) noted gross hydronephrosis and hydroureter at left kidney. Left kidney showed 47% in her DTPA(Diethylenetriaminepentaacetaetate) scan compared with right side 53%. This patient underwent retrograde pyelogram on table to look for patency of ureter and noted tortuous dilated left megaureter from distal ureter up to renal calyx system (figure 1-2). During the surgery noted dilated ureter with thickened wall after the skeletonization of left ureter from urethral orifice extending proximally. Politano leadbetter ureteroneocystostomy procedure was performed for this patient in view of age and better outcome after the surgery (figure3).

Discussions:

Primary obstructed Megaureter (POM) defined as dilatation of ureter bigger than the normal size with or without renal pelvis dilatation. POM was first described in 1923 by Clark[11]. This is the second most common aetiology causing hydronephrosis in infant after ureteropelvic obstruction and it more common in boys compared with girls. This pathology prevalence in the left site compared to right site[2,3]. Delay in the development of muscle or abnormality in the ureter during the gestation cause primary megaureter in the newborn. POM most often detected in children and usually by prenatal or postnatal detail scans. Prenatal scan usually will be done at the gestation age of 18-20weeks. Baby with positive findings in scan such as hydroureter or hydronephrosis will be closely followed up after the delivery. High risk infant or positive antenatal findings in the newborn will undergo further imaging for the early detection of POM. Newborn with hydroureter or hydronephrosis in the postnatal scan are very prone to get urinary tract infection due the anomaly in urinary tract. This type of patient will be started with antibiotic for at least for six months. Song et al.[13]showed that the risk of UTI is higher with VUJ than pelviureteric junction (PUJ) obstruction, and UTIs in this group tend to occur within the first 6 months of life.

First Investigation of choice to detect the POM in newborn or children will be ultrasonography. This imaging modality will be useful in detecting the width of ureter, kidney size and length. The grading of kidney dilatation will scored according to Hofmann[5] with help of ultrasound. Width of ureter for POM children will be focus at the distal part of ureter or bladder level as commonest etiology was VUJ obstruction. Advancement of imaging at the current era lead to more detection of POM cases in the newborn. Studies shows that POM almost caused 23% in the upper urinary tract dilatation and ranked third position after vesicoureteral reflux and pelviureteric junction stenosis[6]. Next favourable imaging modality is diuretic isotopic renogram which will be essential together with ultrasonography in detecting POM. This diuretic isotopic renogramcan performed either with 99mTc MAC-3 or DTPA in detecting degree of renal function and any obstruction in the urinary tract. Voiding cystourethrogramraphy (VCU) is helpful in excluding vesicoureteral reflux and bladder outlet obstruction in POM patient prior to any surgical intervention.

POM patient divided in to asymptomatic and symptomatic. For the asymptomatic patients mostly they will diagnose antenatally using imaging modalitities and proper follow up after birth. However symptomatic patient will be presented with recurrent urinary tract infection (UTI), loin pain, stone formation and deterioration of renal function. These category patients are usually neglected antenatal scan and seek medical aid in third or fourth decade of their life.

Management of POM defer according to the age of patient, onset of detection, type of megaureter and complications arise from POM itself. If the POM detected in childhood, most of the urologist will opted to go with conservative treatment and follow up imaging.Keating et al. reported that when the decision to intervene was based on absolute renal function, up to 87% of patients could be followed up conservatively[7]. Children who treated conservatively for POM needed long term follow up as symptoms can develop after years of observation [8]. There are few parameters determine the spontaneous resolution of POM such as age of presentation (either prenatal or postnatal), type of megaureter, differential renal function (DRF) in isotope renogram MAC-3 (DFT <40%) and renal washout pattern.

Surgical intervention will be indicated in certain conditions such as failure of conservative treatment, massive hydroureteronephrosis with worsening of renal excretory function upon presentation, recurrent urosepsis, and onset of symptoms or obstruction with function deterioration. Majority of POM patient will undergo ureteric reimplantation with or without ureteric tapering. However the time for surgical intervention depends on the patient age that requires operative procedure for POM. In western countries due to challenging operation in newborn
Figure 1: Difficulty insertion of angiocatheter through the left vesicoureteric junction

Figure 2a: left distal ureter tortuous, engorged and dilated up to renal calyx system

Figure 2b: DTPA (Diethylenetriaminepentaacetate) scan: Good functioning kidney on the right side compared on the Left side

Figure 2c: Ultrasound KUB: Gross Hydronephrosis left kidney with thinning cortex
or child less than one year, practise of certain alternative procedure such as temporary double-J stenting, endoscopic balloon dilatation, cutaneous ureterostomy and endoureterotomy are performed. This less invasive procedure can be definitive or temporary treatment for the younger patient without ureteric reimplantation along with surveillance imaging and follow up.

Double-J stent insertion is preferred in newborn with POM and prevents ureteric reimplantation in infancy due small bladder and technical difficulty. This procedure will be done endoscopically and regular follow up is needed for change of stent and urinary tract system assessment whether can treat conservatively or early surgical intervention for the patient. If the patient presented with sepsicaemia secondary to urinary tract of obstruction then more invasive procedures such as percutaneous
nephrostomy or cutaneous ureterostomy will be indicated. This alternative temporary management will depend on patient condition on time of presentation and availability of services in the centre.

However, the ureteric implantation stills the mainstay treatment for POM patient in order to achieve long term good outcome from the surgical intervention. Although this procedure consider challenging in children less one year but early intervention improve the patient renal excretory function and prevent further damage to urinary tract. In ureteric implantation the aperistaltic segment or the narrowed part of the ureter will be excised and the remainder tunnelled into bladder in different ways to prevent reflux. According to a study by Paquin[9] in 1959, the mucosa tunnel should be at least five times than ureteral diameter to be effective, a ratio that may be used as a guide for effective reimplantation. Ureteric implantation can be done by extravesical and intravesical techniques. Extravesical technique by Lich and Gregoir was preferred initially as simple to perform with low morbidity. This procedure also reduced length of stay in the hospital and post-operative pain management compared with intravesical approach. Meanwhile Intravesical reimplantation (Cohen, and Politano-Leadbetter) techniques are more complex and have intra or post-operative complications such as bowel injury, leakage of urine, urethral kinking and stricture[10]. Post-operative endoscopical instrumentations for Cohen’s procedure patient will be difficult due to trigonal ureteric reimplantation thus Politano Leadbetter more preferable.

In recent era with advanced of laparoscopic surgery for ureteric reimplantation the outcome of minimally invasive surgery (MIS) similar with open surgery and better in reducing morbidity and complications.

Long term follows up needed for POM patient if treated conservatively. Reported late deterioration in a 14-yearold boy with bilateral primary megaureters who presented with increased dilatation and functional deterioration 6 years after he was discharged from conservative follow-up [11].

Conclusion:

Early detection of POM is very pertinent, hence the urologist can plan algorithm of management for better outcome. Long term regular follow up with imaging required for POM patient if planned for conservative management. Definitive treatment will be surgical intervention if conservative treatment failed or recurrent complications of POM arise during the observation period. Minimally invasive surgery will benefit the POM patient, however it depends to the availability of the centre and laparoscopic experience of Urologist.

References