Original Research Article

A study of primary megaureter – Our experience

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Abstract

Amongst the congenital anomalies of the Ureter, Megaureter is one of the commonest, next only to the various types of duplications. Though every dilated and large ureter can be described as Megaureter, Primary obstructive megaureter is the entity wherein the dilated ureter is associated with a short, juxtavesical, narrow, adynamic segment and is not associated with reflux. This is a study of 8 patients who presented with primary obstructive megaureter in the department of urology, Osmania General Hospital, Afzalgunj during 2013-2015. Apart from the clinical presentation of this disorder, the modalities of arriving at the Diagnosis, the associated complications and the management are discussed.

Key words

Megaureter, Congenital anomalies, Dilated ureter.

Introduction

Dilatation of the upper urinary tract is not an uncommon urological finding and results from varied aetiological factors. Amongst these, an almost total ureteral dilatation associated with an abrupt narrow calibre, juxtavesical ureteral segment constitutes the entity of primary obstructive megaureter.

Classification of megaureter

- Reflux Megaureter: Primary and Secondary
- Obstructed Megaureter: Primary and Secondary
- Nonrefluxing, Nonobstructive Megaureter: Primary and Secondary

Etiology

The presence of a narrowed juxtavesical, adynamic ureteral segment is the most important cause of primary obstructive megaureter. The

narrow segment measures from 0.5 to 4 cm in length. It is now widely accepted that the obstruction is more functional than mechanical [1].

It is 3.5 - 5 times more common in males. The left ureter is involved 1.6 - 4.5 times more often than right. Megaureter is bilateral in about 25% of patients. In 9% of the cases, there is contralateral renal agenesis. Rarely megaureter coexists with PUJ obstruction. The condition is not known to be hereditary but families with more than one member with megaureter have been described.

Pathology

In many cases, the area of abrupt transition between dilated and normal sized ureter shows faulty muscular development with a segment partially or completely deficient in muscle. An excessive amount of collagen occurs within the undilated segment of ureter and in its adventitia.

Ultrastructural studies show an increase in collagen between muscle bundles of the obstructing segment and between individual muscle cells.

Mode of presentation

Urinary tract infection and flank pain are the usual modes of presentation. Some children may present with signs and symptoms of renal insufficiency like failure to thrive, uremia, anaemia and renal rickets. On occasion the megaureter is an incidental finding during surgery for an unrelated problem like appendicitis.

Diagnosis

The diagnosis is initially established with ultrasound or intravenous urography [2]. Ultrasound examination shows dilatation of the ureter and hydronephrotic changes in the kidney. I.V.U. is the most useful investigation in the diagnosis of obstructive megaureter. The ureter is dilated which progressively widens distally. The most distal portion has a marked fusiform or bulbous dilatation which abruptly changes into a short, undilated segment about 0.5 - 4 cm in length, which enters the bladder. Opacification of the atonic segment may be seen on oblique films or postvoid film. Dilatation of the distal ureter is more pronounced than that of proximal ureter. Despite ureteral dilatation the calyces are normally cupped and pelvis is normal or somewhat plump. Renal function usually remains near normal.

Fluoroscopy shows disturbed peristalsis with failure of at least the more distal portion of the dilated segment to coapt during contraction. This results in regurgitation of the bolus into the upper ureter. Retrograde catheterisation of the undilated segment is usually unimpeded.

If untreated the condition progresses with development of hydronephrosis and increasing ureteral dilatation and tortuosity.

A Retrograde pyelogram is generally not required. If the anatomy of the distal ureter is not well defined with intravenous or ante grade pyelography, a retrograde study can be performed just prior to surgical intervention.

A voiding cystourethrogram is always obtained to rule out reflux [3].

Diuretic renal scan and whittaker tests are useful diagnostic adjuncts when an obstructive lesion is suspected but the findings on I.V.U are equivocal [8]. The diuretic renal scan does not provide anatomical detail but the excretion and drainage of the radionuclide can be quantified and the presence or absence of obstruction can be determined with a high degree of certainity. A half time clearance of radioactivity requiring longer than 20 minutes is indicative of obstruction [6-9].

If the diuretic renogram is equivocal further tests are done.

- Cystoscopy with ureteral catheterisation to see if any hydronephrotic drip is present.

- Whittaker test: A pressure gradient of greater than 20 cm of H_2O indicates obstruction.

Complications
Recurrent urinary tract infections, Calculus formation, Renal failure [7]

Management
Operative intervention is usually required in patients with primary obstructive megaureter [4]. In some cases, when the condition is asymptomatic and does not impair renal function, observation alone is reasonable.

The ureter is remodelled by excisional tapering or and reimplanted with a conventional ureteroneocystostomy. Tapering [10] is done only that portion of wide ureter that would become the new intravesical ureter after reimplantation. The goal of remodelling is to reduce the diameter of the ureter to permit the development of submucosal tunnel that is about 5 times longer than the ureteric diameter [11].

Materials and methods

During the period 2013 -2015 (2 years), 8 cases of primary obstructive megaureter were admitted in the Urology department of Osmania General Hospital and were included in the study.

Results

Out of 8 cases, 5 were unilateral (4 on left side and 1 on right side) and 3 were bilateral. There were 6 male and 2 female patients (3:1). Age of patients ranged from 8 months to 50 years. The youngest patient was a 8 months old male child. The sister of the same child also presented with a left side megaureter later at the age of 2 years. Half of the patients were young individuals seen below 20 years of age.

Clinical presentation
Abdominal pain - 6
Recurrent UTI - 7
Lump abdomen - 1
Haematuria - 2
Past history of Surgery - 3

The most common presentation was pain abdomen and recurrent UTI. One patient had loin-mass. 3 patients gave H/o previous surgery. 2 patients underwent ureterolithotomy, while one patient was subjected for exploration of the lower ureter with a mistaken diagnosis of doubtful calculus.

The routine basic work up of those cases included urine analysis, urine for c/s, haematological and biochemical study and basic radiological investigations of chest X-ray and plain X-Ray KUB to rule out calculus disease.
Further urological work-up included U.S.G and I.V.U followed by M.C.U.G. U.S.G and I.V.U showed dilated pelvicalyceal system and dilatation of ureter in all cases. Pyelonephritic changes were seen in 3 cases. Voiding Cystourethrogram did not reveal any reflux. R.G.P was carried out in one case to demonstrate the dilated ureter with narrow distal segment.

In one case of bilateral megaureter, the right kidney was grossly hydronephrotic and the ureter was not visualised on I.V.U. Retrograde catheterisation was unsuccessful and antegrade pyelography was done. Renal scan was also done in this case to estimate the function of the Kidney and to determine the level of obstruction.

Complications
Calculus - 5
Pyelonephritis - 3

Poorly functioning kidney was seen in one patient. Calculus formation was the most common complication noted which was present in 5 cases. Renal calculi were seen in 2 cases, both renal and ureteric calculi in 2 cases and a lower ureteric calculus in one case. Pyelonephritic changes were seen in 3 patients. Gross hydronephrosis with poor function was noted on right side in one patient with bilateral megaureter.

Management

Tailoring and reimplantation - 3
Ureterolithotomy and reimplantation - 3
Pvelolithotomy and reimplantation - 2

Tailoring of the dilated lower ureter and reimplantation was done in 3 cases. Patients who presented with ureteric calculus as a complication of megaureter underwent ureterolithotomy followed by reimplantation of the dilated ureter. Pyelolithotomy and reimplantation was done in two cases who presented with renal calculi and megaureter.

Discussion

Dilatation of the Upper urinary tract is not an uncommon urological finding and results from varied etiological factors. Amongst these, an almost total ureteral dilatation associated with an abrupt narrow calibre juxtavesical ureteral segment, constitutes the interesting entity of primary megaureter. The silent and apparently stenotic supravesical ureteral segment is held responsible for the gross dilatation of the proximal ureter. This aroused immense interest in the past and was the subject of various hypotheses. However, it is widely accepted that the obstruction is more functional than mechanical and this interesting condition of primary megaureter has become an established entity.

During the period of the 2013-2015 (2 years) we have seen 8 such cases of primary megaureter. Most cases presented with vague symptoms like pain abdomen and recurrent UTI. 3 Patients gave history of previous surgery for calculus disease.

In our series the most common complication noted is calculus formation which is seen in 5 out of 10 patients. The calculus can be in the ureter and or Kidney and is disproportionate to the dilatation of the ureter. A high degree of clinical suspicion and adequate interpretation of routinely done investigations like KUB and IVU is required to clinch the diagnosis. Undiagnosed and late cases are associated with pyelonephritic changes. Ocassionally kidney may be grossly enlarged, mistaken for PUJ obstruction.

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