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A Rare Case of Ureteric valve with associated vesico-ureteric reflux on contra lateral side: A Case report

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Abstract: Congenital ureteric obstruction caused by ureteric valves with associated vesico-ureteric reflux on contra lateral side is an extremely rare entity. Our patient, a two and a half-year-old male, had undergone evaluation for recurrent pain in the abdomen and was diagnosed as a case of left hydronephrosis with hydro ureter on ultrasound abdomen. CT urography was suggestive of lower ureteric obstruction on left side with hydronephrosis and hydroureter. Voiding cystourethrogram was suggestive of Grade II vesico ureteric reflux on right side with no reflux on left side. Cystoscopy followed by excision of lower third of left ureter along with valve and Cohen's uretero neo cystostomy was planned. A high index of suspicion was required to make a correct preoperative diagnosis.
Keywords: Ureteric stricture, ureteric valve, Megaureter, vesico ureteric junction obstruction.

INTRODUCTION

Congenital ureteric valves with associated vesico-ureteric reflux on contra lateral site are very rare and have not been seen in literature. Children with this disorder are often misdiagnosed as having either uretero vesical junction obstruction or primary mega ureter. We report a case of congenital ureteric valve in lower ureter on left side along with Grade II vesi coureteric reflux on right side with a review of the literature.

CASE REPORT

A two and half-year-old male child presented with recurrent abdominal pain in the left lumbar region since two years. A renal ultrasound revealed left hydro-uretero-nephrosis. Voiding cystourethrogram was suggestive of Grade II vesico ureteric reflux on right side with no reflux on left side. A DTPA Renal scan showed a normally functioning left kidney with split renal function of 25% and an obstructive pattern on left side. A nephrostomy drain was put on left side. The right kidney was normal. CT urography showed lower ureteric obstruction on left side with left hydronephrosis and hydroureter. During cystoscopy, a 3 Fr ureteric catheter could not be negotiated beyond 4-5 cm from the vesico ureteric junction. Patient was posted for Cohen's uretero neo cystostomy. On intravesical approach and extra vesical mobilization of the left ureter, normal calibre lower ureter upto5 cm from the bladder wall with gross proximal dilatation of the upper two-thirds of the ureter. A longitudinal incision given over lower third of the dilated ureter .Ureterotomy revealed a membranous web which was excised. Free flow of urine into the urinary bladder checked. Ureterotomy closed over DJ Stent. Patient had uneventful recovery. Post operative radiological investigations showed improvement in morphology and function of left kidney with relief of obstruction

DISCUSSION

Congenital ureteric valves are a rare cause of ureteric obstruction. Only 60 cases of ureteric valves have been reported since Wolfler described his first case in 1877 [1]. For diagnosis of 'ureteric valves', the following criteria should be present:

(1) Presence of transverse folds of the ureteric mucosa containing bundles of smooth muscle fiber on histologic examination,

(2) Signs of obstructive disease above the valve with a normal ureter below it, and

(3) No other evidence of mechanical or functional obstruction [2].

According to Rabinowitz [3], ureteric valves can be classified as Type I or Type II, with Type I having smooth muscle present within the leaflet and Type II having smooth muscle at the base only. On the basis of previously reported cases ureteric valves can be classified morphologically into both cusp-like (leaflet) and diaphragmatic or annular type [3]. Our case is a cusp-like (leaflet) Type II true ureteric valve. The embryogenesis of ureteric valves remains unclear. Two major theories presently exist: the persistence of Chwalle's membrane and physiological folds. The persistence of Chwalle's membrane might explain the presence of lower ureteric valves [2]. Chwalle's membrane is an epithelial membrane in the lower

portion of the ureteric lumen and is a normal feature of ureteric development at six weeks of gestation. During the eighth week of gestation the membrane ruptures under the pressure of urine excretion. Partial rupture of the membrane may result in a retained membrane that would constitute a ureteric valve. However, this theory does not explain multiple valves in one ureter or valves in the upper or mid-ureter [4].

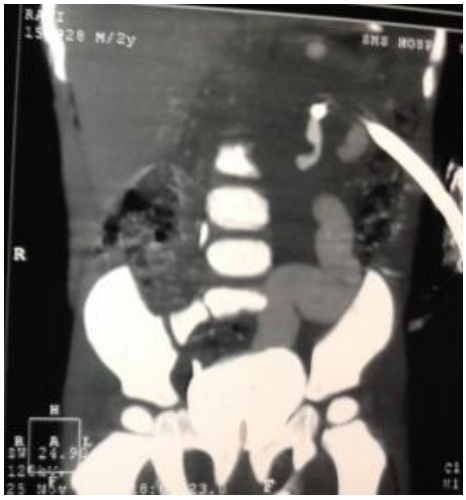

Fig 1: Grossly dilated pelvis

Fig 2: Intra operative

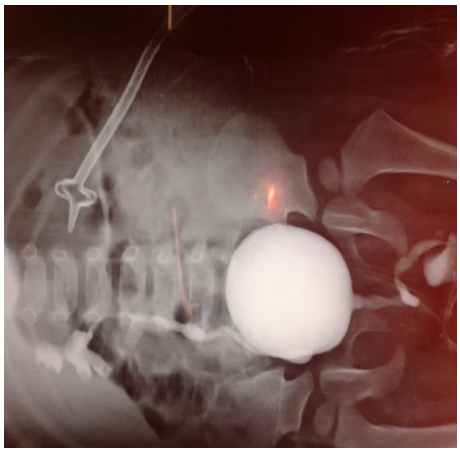

Fig 3: Nephrostomy

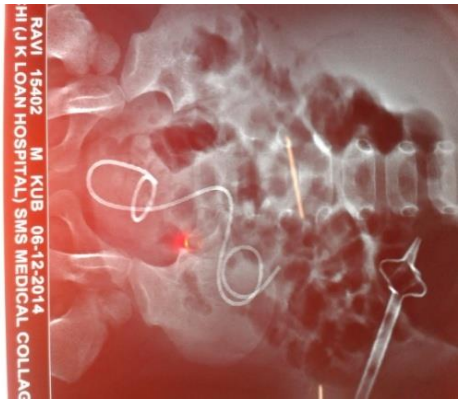

Fig 4: Post operative with stent.

At the fifth week of embryologic life, the ureteric bud originating from the mesonephric (Wolfian) duct grows towards the meta nephric blastema. On the other hand, the meta nephros undergoes an ascent towards its definitive location. If the ureteric growth is faster than the renal migration, then a "ureteric fold" would form. Ureteric valves are the persistence of exaggerated and obstructive fetal physiologic ureteric folds. This theory seems more compatible with multiple valves in one ureter [4.] The distribution of valves within the ureter is reported as 50% in the proximal ureter, 17% in mid-ureter, and 33% in the distal ureter. One case of multiple valves on the same side has also been described [1]. Bilateral involvement is exceedingly rare, and has been described only by Wall and Wachter in 1952 [2] and Paul Daher in 2007 [1].

A patient of ureteric valve can present as a lump in the abdomen due to hydronephrosis, abdominal pain or can be detected incidentally while evaluating suspected cases of mega ureters, uretero pelvic or uretero vesical junction obstruction.

More than 50% of cases with ureteric valves also have associated urinary anomalies, including ureteric duplication, reflux, ectopic ureter, and contra lateral hypoplastic kidney or renal agenesis [3]. Our patient had Grade II reflux on the right side.

The differential diagnosis isco genital mega ureter and congenital ureteric stricture, which is rare, usually found in early adulthood, but often revealed by hydronephrosis on antenatal ultrasound [1]. The stricture corresponds to segmental ureteric fibrosis, usually associated with smooth muscle hypo plasia. The ureteric lumen is usually narrow but sometimes can be of normal caliber. The stricture can be present anywhere in the ureter [1].

Treatment of ureteric valve depends on its location and the severity of renal damage and includes pyelo ureterostomy, primary uretero ureterostomy, or excision of leaflet and ureteric re implantation in cases of distal involvement [3]. Endoscopic incision is also

thought to be a useful treatment for the ureteric valves [1]. Recently, antenatal ablation of ureteric valve by Nd-YAG laser has also been reported [5].

In our case, on the basis of all radiological investigations, the initial provisional diagnosis was left uretero vesical junction obstruction. Despite the availability of advanced diagnostic radiological modalities, a high index of suspicion is needed to make the diagnosis of ureteric valve as a cause of unilateral hydronephrosis and hydroureter.

In our case we confirmed the presence of ureteric valve by performing ureterotomy at the junction of upper dilated and lower non-dilated ureter. We could not negotiate left ureteric orifice due to thickened and edematous urinary bladder wall.

We performed primary uretero ureterostomy over 4.5Fr DJ Stent. We suggest that the diagnosis of ureteric valve should be kept in mind and needs to be confirmed by cystoscopy followed by retrograde ureterography, while evaluating a case of hydronephrosis and hydroureter.

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