



Congenital mid ureteric valve: A case report with review of literature

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ABSTRACT

Ureteral anomalies are difficult to diagnose on routine investigations in the background of a megaureter. Ureteric valves are a rare cause of congenital obstructive uropathy. Mid ureteric valves are rarer than proximal or distal ureteric valves, and most patients are misdiagnosed preoperatively. Therefore, intraoperative identification is critical. We present a case of mid ureteric valve which presented as unilateral hydroureteronephrosis.

Key Words: Mid ureteric valve, ureteric valve, hydroureteronephrosis, child.

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Received: 2020-09-22 / Revisions: 2020-10-03

Accepted: 2020-10-31 / Publication Date: 2020-11-01

Introduction

Hydroureteronephrosis in the pediatric age group is usually secondary to vesicoureteric reflux (VUR) and vesicoureteric junction obstruction (VUJO) [1-4]. Obstructive ureteric anomalies are difficult to diagnose in the background of a megaureter. Ureteric valves may be confused with a pelviureteric junction or vesicoureteric junction obstruction, depending on its level [1-6]. Mid-ureteral obstruction is less frequent (17%) than proximal (50%) or distal ureteric obstruction (33%) [1]. Very few cases of mid ureteric valves have been published till date. Here, a 1.5-year-old girl with congenital mid ureteric

valve stenosis is presented with all the features, including clinical findings, radiological and histological examination and surgical management.

Case report

A 1.5 year girl presented with intermittent flank pain and recurrent urinary infection since birth. Ultrasound revealed left moderate hydronephrosis [anterior-posterior diameter (APD) 18mm] with massive proximal and mid ureteric dilatation (diameter 12.5mm). The right kidney was normal. Blood parameters and urinalysis were normal.

Micturating cysto-urethrogram (MCU) showed no evidence of reflux. Conventional intravenous pyelography (IVP) was suggestive of moderate to gross left sided hydroureteronephrosis with delayed concentration and excretion of dye, but the level of the obstruction was not well delineated. Computerized tomography

intravenous pyelogram (CT/IVP) revealed left sided hydronephrosis (APD: 16mm) and a dilated and tortuous left ureter (11mm) with sudden narrowing at the L4-5 level (Fig. 1).

The distal ureter was normal.

The nuclear renal scan showed a suboptimally functioning obstructed left kidney with delayed T1/2 for drainage (Fig. 2).



Fig. 1. CT/IVP showing left hydronephrosis up to mid ureter.

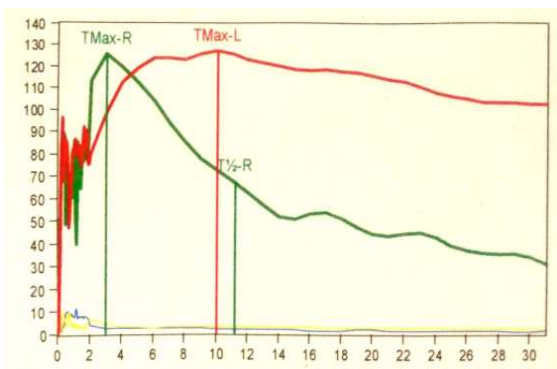


Fig. 2. Delayed drainage of the left kidney system on a nuclear kidney scan.

Cystoscopy with retrograde urography demonstrated a left dilated pelvis and upper ureter suggestive of mid ureteric obstruction (Fig. 3).



Fig. 3. Left dilated pelvis and upper ureter on retrograde urography.

Exploration revealed a dilated left ureter with sudden cut off at mid ureteric level and a normal calibre ureter distally. After Ureterotomy, a cusp-shaped valve obstructing the lumen was visualised (Fig. 4). Local ureterectomy with primary ureteral reconstruction was done over a DJ stent. Histopathology suggested a Type II ureteral valve.



Fig. 4. Intraoperative findings.

Postoperatively at 6 months, sonographic improvement was seen with left renal APD of 8.8mm and upper ureteric diameter of 4.3mm. EC renal scan showed improvement of left renal function and improved drainage (Fig. 5).

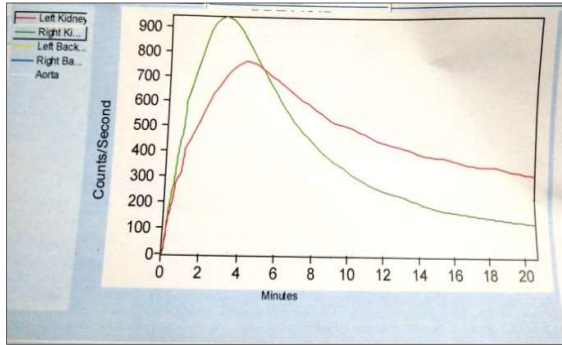


Fig. 5. Improvement of drainage in the post-operative nuclear renal scan.

Discussion

Intrinsic ureteral obstruction between the pelviureteric and ureterovesical junction in children is caused by primary or secondary strictures of the ureter, ureteric polyps, ureteric valves (upper, mid, or lower ureter), stenosis, or diverticula. A high level of suspicion and detailed investigations are necessary for diagnoses [2].

Congenital ureteric valves are a rare cause of ureteric obstruction and have been rarely reported since it was described by Wolfler in 1877 [2]. Various theories of embryogenesis of ureteric valves have been put forward.

- Recanalization starts at the midpoint and progresses in both directions.
- Persistent Chwalle's membrane where the ureter joins the urogenital sinus.
- Abnormal differential growth of ureteric bud with respect to metanephric blastema [3, 5].
- Ischemic injury occurring during the elongation of the ureteral bud [4].

- Persistent ureteric mucosal folds.
- Intrauterine ureteritis [4].

The persistence of Chwalle's membrane might explain the presence of lower ureteric valves but aberrant growth of ureteric bud seems more compatible with valves in upper and mid-ureter and also multiple valves in one ureter [3, 5].

Valves are usually reported in the upper ureter (50%), followed by distal ureter (33%) and least commonly in the mid-ureter (17%). Bilateral involvement is exceedingly rare and presents early with anuria [1].

Morphologically, ureteric valves are classified as cusp-like (leaflet), diaphragmatic or annular type [3]. Histologically, Rabinowitz classified valves as Type I where smooth muscle was present in the leaflet and Type II wherein smooth muscle was present only at the base [3].

Usual presentation of these valves is fever, recurrent urinary tract infection, recurrent abdominal or flank pain or incidentally diagnosed hydroureteronephrosis [3]. Sometimes, as reported by Ormond, a congenitally dilated ureter may contain a calculus. In the pediatric patient presenting with a calculus, it is essential to rule out distal obstruction. Due to the rarity of mid ureteric valves, the chances of delayed presentation as well as delayed diagnosis are high. Sometimes such valves may even be found during a ureteric reimplantation for vesicoureteric junction obstruction, where a normal lower ureter may be seen which, on dissection, may reveal the dilated segment with a cut off further upstream [6].

Diagnosis needs a high degree of suspicion. Delay leads to significant deterioration of function of the affected renal system which may become unsalvageable [7].

Diagnosis of ureteric valves in a patient presenting with hydroureteronephrosis can be made if the following criteria are met:

1. Presence of transverse folds of the ureteric mucosa containing bundles of smooth muscle fibres on histological examination,
2. Signs of obstructive uropathy above the valve with a normal ureter below
3. No other evidence of functional or mechanical obstruction [4].

Association with urinary anomalies namely ureteric duplication, reflux, ectopic ureter, and contralateral hypoplastic kidney or renal agenesis is present in more than half the cases [4].

Antenatal diagnosis of ureteric valves is difficult but has been reported [8]. Ultrasonography and Micturating cystourethrogram (MCU) are the initial investigations guiding the diagnosis of hydroureteronephrosis, followed by a functional urography to confirm an obstructive pathology.

CT IVP has lower rates of prediction in lesser dilated systems [9]. Magnetic resonance urography (MRU) can visualize the entire course of the ureter, identify ectopic insertions, allow for the diagnosis of UPJ obstruction, identification of any extrinsic cause of the ureteral obstruction (e.g. crossing vessel) and has the ability to detect asymmetric split renal function and parenchymal alterations [10]. Arlen et al. showed that MRI led to a definite diagnosis in all children of mid ureteral pathologies [11]. A cystoscopy with retrograde urogram will demonstrate both the level and degree of obstruction. However in the case of an eccentric orifice, a ureteric catheter may be sometimes difficult to negotiate [6].

The differential diagnosis is congenital ureteric stricture, which is often revealed by

hydronephrosis on antenatal ultrasound [1]. The stricture corresponds to segmental ureteric fibrosis with smooth muscle hypoplasia [1]. Surgery is the mainstay of treatment of ureteric valves. Excision with primary ureteral reconstruction can be performed laparoscopically as well as an open procedure [12]. Sometimes ureteric valves closer to the vesicoureteric junction may be identified during a transvesical ureteric reimplantation if the level is close to the bladder and may be managed transvesically. However, in the case of a massively dilated ureter, difficulties may be faced during reimplantation [6]. Minimally invasive techniques including holmium laser ablation or removal of the valve with biopsy pens are also conducive [13].

Conclusion

Mid ureteric valves are rare causes of unilateral hydroureteronephrosis, and most patients are misdiagnosed preoperatively. Therefore, despite advances in diagnostic radiological modalities, a high degree of suspicion is critical to diagnosing the ureteral valve as a cause of unilateral hydronephrosis and hydroureter.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Patient confidentiality is maintained and written consent for the publication of patient details and clinical pictures in this journal has been obtained from the patient's parents or closest relative and can be given as required.

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