Case Report

Congenital midureteral obstructions

Yogesh Kumar Sarin, Arvind Sinha, Sunita Ojha

Department of Pediatric Surgery, Maulana Azad Medical College and Associated Lok Nayak Hospital, New Delhi, India

For correspondence:
Arvind Sinha, Department of Pediatric Surgery, Advanced Pediatric Centre, Post Graduate Institute of Medical Education and Research (PGIMER), Chandigarh -160 012. E-mail: drsinha@vsnl.com

ABSTRACT

Congenital midureteral obstruction caused by a ureteral stricture, valve, or an adynamic segment, is an exceedingly rare entity. When encountered, it is generally misdiagnosed as megaureter or ureteropelvic junction obstruction. Two children with midureteral obstructions were seen over the last two years. One of these had an anatomical defect- a midureteral stricture; the other had a physiological abnormality- midureteral adynamic segment. In one of the cases, the midureteral obstruction was missed initially; the patient underwent ipsilateral ureteroneocystostomy for vesicoureteral reflux. Later, midureteral stricture was diagnosed; excision of the lesion with primary anastomosis was done, with successful salvage of the renal unit. The other patient was pre-operatively diagnosed to have midureteral stricture; intra-operatively an adynamic midureteral segment of 4 cms was found, which was resected, and ureteral anastomosis was done. Pathological examination revealed probe-patent ureter with muscular disarray, suggesting functional obstruction. Neither of the patients had contralateral renal abnormality. A high index of suspicion is required to make a correct pre-operative diagnosis. Antegrade or retrograde urography would clinch the diagnosis. Over-reliance on radionucleotide scans to diagnosis the level of ureteral obstructions could be misleading occasionally, as happened in one of our cases.

Key words: Ureteral obstruction, ureteral stricture, functional obstruction

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Congenital midureteral obstructions because of either valve or stricture, are extremely rare. Children with this disorder are often misdiagnosed as having either ureteropelvic junction obstruction, or primary megaureter. An awareness of this entity, combined with a systematic radiological evaluation of children with hydronephrosis, will allow the correct preoperative diagnosis to be made, and thus a more appropriate surgical management. We have had two cases of midureteral obstruction. The clinical findings, evaluation, and the treatment of these children are described.

CASE REPORTS

Case 1

HK was the product of an uncomplicated 38 weeks gestation. A maternal ultrasound at 28 weeks had revealed an enlarged right kidney, compatible with hydronephrosis. At birth, she weighed 3020 gms, and there were no obvious abnormalities on physical examination. A renal ultrasound revealed a right hydronephrosis. A voiding cystouretherogram at nine weeks of age was normal, and DTPA scan showed a normally functioning right kidney with a partially obstructive hydroureteronephrosis. The left kidney was normal. A follow up renogram at 11 months of age revealed mild dilatation of the right renal pelvis with a grossly enlarged ureter. Direct radionucleotide cystouretherography performed at this stage suggested a grade V vesicoureteric reflux. So, right Cohen's ureteroneocystostomy was done. During the surgery, a number 3 ureteric catheter could be passed upto the renal pelvis, but an infant feeding tube was getting stuck after a distance of 4-5 cms. But presence of any ureteral obstruction was discounted for, and this was thought to be due to the tortuosity and kinking of the ureter. Postoperatively however, the infant feeding tube which was left as a ureteric stent, drained only 10-12 ml urine a day. On the third postoperative day, an ultrasound-

Source of Support: Nil.

guided percutaneous nephrostomy was performed. A nephrostogram performed through the same showed a mid-ureteric block. On the 7th postoperative day, a local exploration was done, which revealed a "true" midureteral valve with an eccentric pinpoint orifice. The obstructed segment of the ureter was excised, and end-to-end ureteroureterostomy was performed. The child has been well since the operation. A follow up renogram twenty-one months after surgery, revealed residual ureteral dilatation but no obstruction, and a well-preserved ipsilateral kidney.

Pathologic examination revealed a ureteral valve lined by transitional epithelium overlying a stroma containing smooth muscle fibres and fibrous tissue.

Case 2

A three-year old girl, presented with pain on the left side of the abdomen. She was diagnosed to have hypertension, and was being managed on oral nifedepine therapy. An ultrasound examination of the abdomen revealed a hydronephrotic left kidney with a dilated left upper ureter. The right kidney was normal. Intravenous pyelography revealed a hydronephrotic left kidney, with the ureter dilated till the left sacroiliac joint [Figure 1]. The micturating cystouretherography (MCU) revealed no vesicoureteral reflux. The preoperative renal scan showed a dilated left kidney and ureter. The differential function of the left kidney was 25%. A pre-operative diagnosis of mid-ureteral obstruction was made. Intra-operatively, an adynamic midureteral segment of 4cms was found which was resected, and ureteral anastomosis was done. Pathological examination revealed probe-patent ureter with muscular disarray, suggesting functional obstruction. The patient continued to have hypertension in the postoperative period. A postoperative renal scan was performed, which showed a grossly hydronephrotic left kidney with functioning parenchyma at the upper pole, with severely impaired function. The differential left renal function was 11%. In view of the poor function and persistent renal hypertension, the patient was taken up for left nephrectomy. The patient is normotensive on follow up, and is doing well.

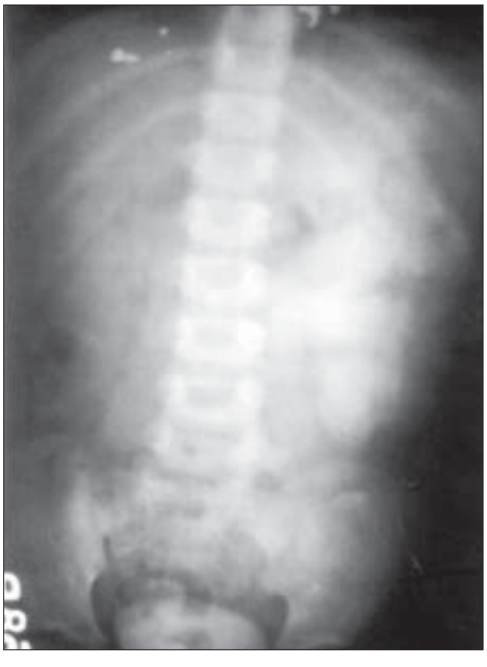

Figure 1: IVP of the case 2 showing left hydronephrosis with the left ureter dilated till the left sacroiliac joint

DISCUSSION

Congenital anatomical or functional obstructions of the mid-ureter are exceedingly rare. There have been only 18 previously cases of well-documented ureteral valves in pediatric literature.[1] It is important not to confuse the 'true' ureteral valves with non-obstructive ureteral fetal folds. At times, eccentric, cusp-like flaps or folds can be obstructing, but these may be secondary to the elongation and tortuosity seen in megaureters having and underlying anatomic or functional obstructions, but to authenticate 'ureteral valves', the following criteria should be present.[2]

1. Anatomically demonstrable transverse folds of ureteral mucosa containing smooth muscle fibres.
2. Obstructive changes above but not below the valve.
3. No other evidence of obstruction.

However, some authors dispute the criterion that smooth muscle should be present within the body of the valve.[1] In our first case, the ureteral valve was lined by transitional epithelium overlying a stroma containing smooth muscle fibres.

The exact embryogenesis of the ureteral valve is unknown,[3] but has been variously explained on the basis of the fetal fold theory[4] and Chawalle's theory.[5]

Functional obstruction at the ureteropelvic junction or the ureterovesical junction is a commonly encountered problem. Congenital functional obstruction of the midureter however, is a much rarer condition, with only 17 reported cases.[6] In our second case, there was no intrinsic obstruction present, such as a congenital ureteral valve. Histopathologically, the ureter had shown muscular disarray, suggestive of a functional obstruction. Allen described the pathological appearance of a case of midureteral obstruction;[7] in which the segments were narrowed, but the muscle fibres did not appear to be abnormal, suggestive of a functional obstruction. The term "Midureteral adynamic segment" has been in vogue for the last few years only.[6] The etiology of congenital ureteral obstruction secondary to adynamic segments regardless of location, is unclear. One theory suggests that compression by iliac vessels may be the cause of midureteral obstructive lesions. Others suggest a failure

of recanalization of the ureter,[8] or acquired fetal inflammation of the ureter similar to biliary atresia.[9]

In one of our cases, the diagnosis of the midureteral obstruction was missed on preoperative imaging studies. The diagnosis was subsequently confirmed by an antegrade dye study of the ureter through a nephrostomy. In the second case, the diagnosis could be made preoperatively, because the intravenous pyelography delineated the site of obstruction. We drew a few lessons from our short experience. Although ultrasonography could provide anatomic details, it may not be able to delineate the exact underlying pathology. On the other hand, overuse of nuclear uroradiology misled us to a wrong pre-operative diagnosis of vesicoureteral reflux in one of the cases. It has to be most emphatically stressed here, that probably intravenous pyelography (IVP) is the gold standard in the diagnosis of such unusual lesions. IVP should always supplement the nuclear imaging studies which detail the functional information, but lack in providing accurate anatomic details.

A systematic progression of imaging studies in the evaluation of hydronephrosis, such as ultrasonography, voiding cystouretherography, intravenous pyelography, and especially if the diagnosis is in question, percutaneous antegrade pyelography or retrograde ureteropyelography will allow a preoperative diagnosis in the majority of cases of congenital midureteral obstruction. The resulting increase in diagnostic accuracy will allow a specific surgical intervention.

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