Congenital midureteric stricture: A rare entity in pediatric patient

Prashant Sadashiv Patil, Gupta Abhaya, Kothari Paras L, Kekre Geeta, Dikshit Vishesh K, Shahaji Deshmukh, Apoorva Kulkarni

Department of Pediatric Surgery, L.T.M.G. Hospital, Sion, Mumbai, Maharashtra, India

Abstract

Congenital midureteric obstruction is a rare entity which can be caused by either ureteric valves, strictures or an adynamic segment. When encountered, it is generally misdiagnosed as megaureter or ureteropelvic junction obstruction. A high index of suspicion is required to make a correct pre-operative diagnosis. Antegrade or retrograde urography would clinch the diagnosis. Our patient presented with a history of left loin pain. Investigations suggested mid-ureteric stricture. Resection of stricture segment and primary ureteroureteric anastomosis was done which relieved the symptoms. Histopathological examination showed muscle fibrosis.

Keywords

Midureteric stenosis; midureteric stricture; hydronephrosis; ureteroureteric anastomosis.

Copyright © 2016 pediatricurologycasereports.com.

Corresponding Author: Dr Prashant Patil,

Ward 1A, Pediatric Surgery Ward, L.T.M.G.

Hospital, Sion, Mumbai, India.

E-mail: docprashant2010@gmail.com

Accepted for publication: 30 September 2016

Introduction

Congenital midureteric strictures (CMUS) are extremely rare cause of obstructive uropathy. Children with this disorder are often misdiagnosed as having either pelviureteric junction (PUJ) obstruction or primary megaureter. Diagnosis is suspected after computed tomography (CT) urography.

Retrograde pyelography (RGP) and/or intravenous urography (IVU) may be helpful for anatomic localization of stenosis/stricture and the course of ureter, as shown in our cases. The final diagnosis is confirmed during surgical exploration [1,2]. We report here our experience in clinical findings, preoperative investigations, and operative treatment in 4 year old boy diagnosed as midureteric stricture.

Case Report

A four and half years old male child complained of intermittent colicky pain in left flank since one year. Patient had no other urinary complaints. His bowel habits were normal. Physical examination revealed tenderness in left flank. Rest of abdomen was normal. His vital parameters were stable. Routine complete hemogram, renal function tests, urine analysis were normal. Kidney ultrasonography (USG) showed left hydronephrosis, Antero-posterior diameter of pelvis was 3.5 cm. Renal parenchymal thickness of left kidney was normal compared to right kidney. Left upper ureter showed mild dilatation.

Renal scintigraphy was suggestive of left pelvi-ureteric junction obstruction. Both kidneys had normal function. There was no scarring [Fig. 1].

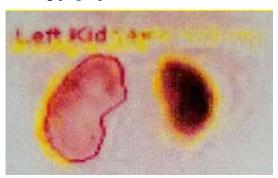


Fig. 1. Renal scan showing hydronephrotic left kidney.

CT urography revealed smooth tapering at the junction of upper and mid ureter at the level of L4 with poor opacification of distal ureter on prone scan, possibility of valve or membrane in ureter [Fig. 2]. Retrograde pyelography was done on table. It showed

non-visualisation of upper and mid-ureter [Fig. 3].



Fig. 2. CT urography showing smooth tapering at the junction of upper and mid ureter at the level of L4 with poor opacification of distal ureter on prone scan.



Fig. 3. Retrograde pyelography showing non visualization of upper and mid ureter.

Patient was explored through flank incision. Intraoperatively a stricture was found at the junction of upper and mid ureter with dilated upper ureter [Fig. 4].



Fig. 4. Intra-op photo showing upper ureter dilatation and stricture segment.

The stricture was excised and ureteroureteric anastomosis was done over a DJ stent. DJ stent was removed after 6 weeks. Histopathological examination of excised segment revealed normal urothelial lining, wall showing thickening with muscle fibrosis. Patient is asymptomatic since 1 year on follow-up. Left renal hydronephrosis resolved.

Discussion

Congenital midureteric strictures (CMUS) are a rare entity, which may narrow of ureteral lumen diameter by 60% [3]. The pathophysiology and etiology of CMUS is not certain, but has been attributed to various factors such as abnormal ureteric

embryogenesis, intrauterine ureteritis, extrinsic compression by blood vessels and some derangement during recannulization of the ureter during fetal life [4]. In severe strictures, the smooth muscle layer is replaced by fibrous tissue and associated with other abnormalities of the urinary tract [5,6]. The differential diagnosis must additionally include ureteral valves and fibro epithelial polyps [7,8].

For a good treatment planning, it is important to obtain the exact localization of the ureteral tract obstruction. Hamid et al [9] recommended that the RGP should be performed in this cases where information provided by USG, IVU, and diuretic renal is scintigraphy equivocal. Otherwise, magnetic resonance urography has the potential to give accurate anatomical, functional details of ureterorenal system and, arguably, the location of the stricture. Additionally, CT urography may be helpful, as in our case.

Definitive management includes open or laparoscopic resection of the stenosed segment with uretero-ureterostomy [8,10]. Endourological dilatation or endoscopic incision of the ureteral stricture may be considered, but these techniques have a lower chance of success than an ureteroureterostomy or ureteral reimplant [11]. In the current case, the stricture was

excised and uretero-ureteral anastomosis over a DJ stent was done.

As a results, CMUS is a rare but important differential diagnosis of hydronephrosis. There should be a systematic sequence of imaging studies for the evaluation of hydronephrosis. However, RGP still have an important roles for the accurate diagnosis of

the stenosis. Tension-free primary ureteroureterostomy is the most favorable treatment option.

Acknowledgements

The author(s) declare that they have no competing interests and financial support.

References

- Chandrasekharam VV. Laparoscopic repair of congenital midureteric strictures in infants and children. J Pediatr Surg. 2015;50(11):1909-13.
- Docimo SG, Lebowitz RL, Retik AB, Colodny AH, Bauer SB, Mandell J. Congenital midureteral obstruction. Urol Radiol. 1989;11(3):156-60.
- Brugnara M, Cecchetto M, Manfredi R, Zuffante M, Fanos V, Pietrobelli A, Zaffanello M. Prenatal diagnosis of a rare form of congenital mid-ureteral stricture: a case report and literature revisited. BMC Urol. 2007;8;7:8.
- Singh SJ, Watson AR, Somers J, Broderick N, Rance CH. Congenital midureteric stricture. Saudi J Kidney Dis Transpl. 2001;12(1):9-13.
- Smith BG, Metwalli AR, Leach J, Cheng EY, Kropp BP. Congenital mid-ureteral stricture in children diagnosed with

- antenatal hydronephrosis. Urology. 2004; 64(59:1014-9.
- Cauchi JA, Chandran H. Congenital ureter strictures: an uncommon cause of antenatal detected hydronephrosis. Pediatr Surg Int. 2005; 21(7):566-8.
- 7. Tartari F, Bezhani E, Rota T, et al. Ureteral valves associated with ureteral stricture. Report of 4 cases. Eur Urol. 1998; 33(4):412-3.
- 8. Harshawardhan Vedpalsingh T, Avinash Vijay J, Vinayak Gorakhnath W, Manoj Vilas B, Sudarshan Omprakash D. Congenital Bilateral Mid Ureteral Stenosis: A Rare Finding. J Clin Diagn Res. 2015;9(5):PD03-4.
- 9. Hamid R, Bhat NA, Rashid KA.

 Congenital midureteric stricture:
 challenges in diagnosis and management.

 Case Rep Urol. 2015;2015:969246.
- Bhandarkar DS, Lalmalani JG, Shah VJ.
 Laparoscopic resection and

ureteroureterostomy for congenital midureteral stricture. J Endourol. 2005;19(2):140-42.

11. Smith BG, Metwalli AR, Leach J, Cheng EY, Kropp BP. Congenital midureteral stricture in children diagnosed with antenatal hydronephrosis. Urology. 2004;64(5):1014-9.



Pediatric Urology Case Reports is an open access journal. Articles published in this journal are licensed under the Creative Commons Attribution 4.0 International License (see http://creativecommons.org/ and http://creativecommons.org/licenses/by/4.0/).