

Horseshoe Kidney (HSK) with Upper Ureteral Valve Masquerading as Pelvi-Ureteric Junction (PUJ) Obstruction. A Case Report and Review of Literature

Satish Kumar¹, Reshma Murali^{2*}, Srinivas³, Ravinder Reddy² and Asha Thomas⁴

¹Consultant Paediatric Surgeon, Department of Paediatric Surgery, Bangalore Baptist Hospital, Bangalore, India

²General Surgery Resident, Department of General Surgery, Bangalore Baptist Hospital, Bangalore, India

³Consultant Urologist, Department of Urology, Bangalore Baptist Hospital, Bangalore, India

⁴Consultant Radiologist, Department of Radiology, Bangalore Baptist Hospital, Bangalore, India

***Corresponding Author:** Reshma Murali, General Surgery Resident, Department of General Surgery, Bangalore Baptist Hospital, Bangalore, India.

Received: March 09, 2020; **Published:** July 29, 2020

Abstract

Horseshoe kidney (HSK) is most common type of renal fusion anomaly with high incidence of hydronephrosis due to pelviureteric junction obstruction (PUJO) and has been increasingly diagnosed antenatally. Upper ureteral valve obstruction presents like PUJO and progression of hydronephrosis can be rapid which may need early intervention. Here we have reported a rare case of upper ureteral valve in HSK whose antenatal ultrasonography showed evidence of HSK and right hydronephrosis and was postnatally diagnosed to have HSK with right sided hydronephrosis. RGP showed reduced differential renal function on right side (35%) with obstruction at PUJ. On pyelotomy, ureteral valve was confirmed causing obstruction to the proximal ureter. A standard pyeloplasty was performed with stent placement. Upper ureteral valve causing hydronephrosis, which was difficult to differentiate from PUJ obstruction on imaging. In our study we have are highlighting the importance of RGP in such uncommon presentations.

Keywords: *Horseshoe Kidney; PUJ Obstruction; Hydroureteronephrosis; Upper Ureteral Valve*

Introduction

Horseshoe kidney (HSK) is most common type of renal fusion anomaly with high pyelogram (RGP) and appropriate management is presented.

Incidence of hydronephrosis due to pelvi ureteric junction obstruction (PUJO) and has been increasingly diagnosed antenatally. The usual management is as for any other antenatally diagnosed PUJO. The presentation of upper ureteral valve obstruction is akin to PUJO and progression of hydronephrosis can be rapid which may need early intervention. A case of HSK with hydronephrosis due to upper ureteral valve obstruction, highlighting the diagnostic difficulties, timely benefit of RGP.

Case Report

A healthy baby girl with antenatal evidence of HSK and right hydronephrosis was delivered at term. Postnatal ultrasound confirmed HSK with right sided hydronephrosis and the baby was well with no dysmorphism, had normal renal parameters and cardiac echo. Follow

Citation: Reshma Murali, *et al.* "Horseshoe Kidney (HSK) with Upper Ureteral Valve Masquerading as Pelvi-Ureteric Junction (PUJ) Obstruction. A Case Report and Review of Literature". *EC Paediatrics* 9.8 (2020): 204-207.

up ultrasounds showed progressive hydronephrosis and at 2 months, the right kidney showed thinned out parenchyma with anteroposterior pelvic diameter of 3.2 cm with a nondilated right ureter with normal left kidney and isthmus. An isotope renogram suggested, reduced differential renal function on right side (35%) with obstruction at PUJ. The child underwent cystoscopy, RGP and open pyeloplasty. Cystoscopy showed bilateral orthotopic ureteral orifices and RGP showed enlarged right pelvis and dilated proximal 1 cm of ureter, with abrupt narrowing and normal caliber ureter distally.

The right kidney was accessed extraperitoneally through transverse lower abdominal incision. On pyelotomy, a diaphragm type of ureteral valve was confirmed (corresponding to level of obstruction seen on RGP) causing obstruction to the proximal ureter (Figure 1). A standard pyeloplasty was performed with placement of a 3fr. double J stent. The child's post-operative recovery was uneventful and the stent was removed after 6 weeks. Follow up ultrasound at three months showed decrease in hydronephrosis in comparison to pre-operative ultrasound.

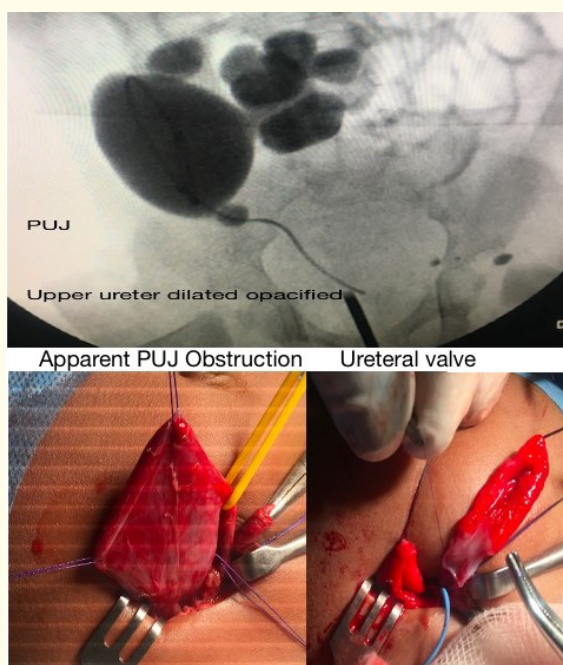


Figure 1: RGP showing dilated pelvis and adjacent ureter with cutoff level. Valve apparent on pyelotomy and ureteral spatulation.

Discussion

HSK occurs in 0.25% of the population (1 in 400 persons) [1]. They can lie anywhere from the pelvis to mid-abdomen and the isthmus is usually at L3 - L5 level, beneath the origin of IMA. Fusion occurs during 4 - 6th week of gestational age and in > 90%, the lower poles fuse [2]. In a study of 380 cases of HSK by Je BK., *et al.* [3], 99.2% had fused lower poles. Lateral fusion can result in formation of L-shaped horseshoe kidney, with ipsilateral moiety in vertical and other moiety in a horizontal orientation [4]. In our case the contralateral kidney was vertical, with ipsilateral kidney being horizontal. HSK may occur as an isolated entity, but approximately 1/3rd are associated with other congenital anomalies [5]. Horseshoe kidney occurs in 60% of females with Turner's syndrome (45 XO). Genitourinary anomalies occur more frequently in HSK (Vesicoureteral reflux (VUR) in over half, PUJO in 1/3 and ureteral duplication in 10% of patients) [1].

Hydronephrosis (seen in 70% of patients) can be due to high insertion of the ureter into the renal pelvis, its abnormal course anterior to the isthmus, and the anomalous blood supply to the kidney, which may individually or collectively contribute to the hydronephrosis [1].

The association of ureteral valves with HSK has not been reported but, has been mentioned only once in the world literature till date. In a review of 64 cases of ureteral valves over 80 years (1926 to 2006) by Rossi E., *et al.* [6], 40% had associated urinary anomalies including 1 case of HSK (Table 1). Rabinowitz., *et al.* reported 7 of their 8 patients with ureteral valves having associated anomalies (Reflux, renal hypoplasia/agenesis and ectopic ureters) [7]. The embryogenesis of valves is unclear and persistence of Chwalla’s membrane, physiological folds and abnormal ureteral embryogenesis⁶ has been hypothesized. Abnormal growth of ureteric bud may lead to valves in upper and mid-ureter [8], while persistence of Chwalla’s membrane have been proposed to cause lower ureteral valves [9].

Incomplete renal duplication	1
Complete renal duplication	4
VUR	7
Bifid renal pelvis	1
Horseshoe kidney	1
Renal agenesis	2
PUR	1
Urethral duplication	1
PUJ obstruction	3
Renal dysgenesis	3
Renal ptosis	1
Ectopic ureter	1
Total	26 (40%)

Table 1: Associated urinary tract anomalies in 62 cases of ureteral valves reported [6].

Valves are reported to be found in the upper ureter (50%), followed by distal ureter (33%) and least commonly in the mid-ureter (17%). Morphologically ureteric valves can be: cusplike (leaflet), diaphragmatic or annular type [6]. Our patient with a presumptive diagnosis of right PUJO, had upper ureteral obstruction, with significantly dilated pelvis, dilated upper ureter due to obstruction by diaphragmatic type ureteral valve.

Most upper ureteral valves reported, have been managed by dismembered pyeloplasty [6]. The standard dismembered pyeloplasty is also the preferred operation for PUJO in HSK. In some cases, dismembered pyeloplasty may not be suitable and an ureterocalycostomy is an alternative [10].

Conclusion

Hydronephrosis secondary to PUJO is a common occurrence in HSK. Upper ureteral valve causing hydronephrosis, which was difficult to differentiate from PUJ obstruction on imaging. A rare case of upper ureteral valve in HSK is reported here, highlighting the importance of RGP in such uncommon presentations.

Acknowledgements

Dr. Srinivas-Urologist for his contribution in editing the manuscript. Dr. Asha Thomas, Radiologist for help in imaging and diagnosis.

Conflicts of Interest

Nil.

Sources of Support

Nil

Bibliography

1. Shapiro E and Telegra S. "Anomalies of the Upper Urinary Tract". In: Wein AJ, Kavoussi LR, Partin AW, Peters CA., editors. Campbell-Walsh Urology. 11th edition Philadelphia: Elsevier (2016).
2. Taghavi K, *et al.* "The horseshoe kidney: Surgical anatomy and embryology". *Journal of Pediatric Urology* 12.5 (2016): 275-280.
3. Je BK, *et al.* "Incidence and Spectrum of Renal Complications and Extrarenal Diseases and Syndromes in 380 Children and Young Adults with Horseshoe Kidney". *American Journal of Roentgenology* 205.6 (2015): 1306-1314.
4. Hardik S and Vijayanadh O. "Multimodality imaging spectrum of complications of horseshoe kidney". *Indian Journal of Radiology and Imaging* 27.2 (2017): 133-140.
5. O'Brien J, *et al.* "Imaging of horseshoe kidneys and their complications". *Journal of Medical Imaging and Radiation Oncology* 52.3 (2008): 216-226.
6. Rossi E, *et al.* "Congenital ureteral valves: Two new cases and a review of the literature". *Journal of Pediatric Urology* 3.5 (2007): 344-349.
7. Rabinowitz R, *et al.* "Ureteral valves in children". *Urology* 51.5 (1998): 7-11.
8. Maizels M and Stephens FD. "Valves of the ureter as a cause of primary obstruction of the ureter: anatomic, embryologic and clinical aspects". *Journal of Urology* 123.5 (1980): 742-747.
9. Pandey V, *et al.* "Mid-ureteric obstruction due to diaphragmatic valve: Case report with review of literature". *IOSR Journal of Dental and Medical Sciences* 13.4 (2014): 110-111.
10. Dewan PA, *et al.* "Ureterocalycostomy in the management of pelvi-ureteric junction obstruction in the horseshoe kidney". *BJU International* 84 (1999): 366-368.

Volume 9 Issue 8 August 2020

© All rights reserved by Reshma Murali, *et al.*