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Case Report

Case of Horse Shoe Kidney, Unilateral Duplex Kidney and aberrant renal vessel with Pelviureteric junction Obstruction of the lower moiety –a rare co-occurrence.

Dr Kapil Rampal^{1*}, Dr Devendra Prajapati¹, Dr Meghna Sharma²

¹Senior Resident surgery Deen Dayal Upadhyaya Hospital, Hari Nagar, New Delhi- 110064. ²Medical officer PCMS-1, CHC Kasel, Tarn Taran Punjab -143401.

*Corresponding author

Dr. Kapil Rampal

Email: balkarankapil@gmail.com

Abstract: Horse shoe kidney, pelviureteric junction obstruction, aberrant/crossing renal vessel and duplicated collecting system as individual entities are amongst commonest of renal anomalies, however very few co-occurrences have been reported. We report case study of a 17 year old male who presented to us with all these anomalies packed in a single individual. Successful surgical intervention was performed in the form of open pyeloplasty and the patient had an uneventful recovery.

Keywords: Horse Shoe Kidney, Double Moiety, Pelviureteric junction Obstruction, Hydronephrosis, Pyeloplasty.

INTRODUCTION

Horse shoe kidney has been reported to be the most common congenital anomaly affecting approximately 1 in 400 live births, being silent asymptomatic condition in majority [1]. In more than 90% cases the fusion happens at the inferior pole. Though no hereditary traits have been discovered as yet, male is to female ratio of 2:1 is reported [2].

Duplication anomalies of collecting system are the most common upper tract anomalies with a reported incident of 0.5-0.8% in general population [3]. There appears to be a strong genetic predisposition in those with duplex kidneys. Up to 30% have relatives with compete duplex kidneys and two thirds with bifid systems.

Pelviureteric junction obstruction is the most common cause of pediatric hydronephrosis occurring in 1 per 1000-2000 live births as variously reported having a 2 times male preponderance, with more cases now being diagnosed antenatally [4].

Despite the commonality of horse shoe kidney, duplication of collecting system or a duplex system and pelviureteric junction obstruction, co-occurrence of these in same one patient is exceedingly rare [5].

CASE PRESENTATION

A 17 year old male presented to us in surgery department with complaints of constant dull aching left flank pain with intermittent exacerbations for 02 year. No associated history of fever, haematuria, gastro-

intestinal complaints or lower urinary tract symptoms present. His general and systemic examination was essentially within normal limits. All hematological and biochemical parameters were normal. The ultrasound abdomen was suggestive of bifid left kidney with severe hydronephrosis of the lower moiety, right kidney and bilateral uretrers were normal. The intravenous urogram (Figure 1) showed duplicated pelvicalyceal system of the left kidney with normal pattern in the upper moiety but the lower moiety showing gross clubbing of the calyces. Both the ureters joined opposite lumbar 3rd and 4th vertebrae. Left kidney was also malrotated (Figure 2). Right kidney and bilateral ureters were normal.

CT urogram performed over the patient showed both kidneys to be fused at lower poles with isthmus infront of lumbar 3rd vertebra, left kidney bifid with hydronephrotic lower moiety (Figure 3). CT urogram also confirmed malrotation of left kidney with anterolaterally directed pelvis (Figure 4).

DTPA Scan showed left kidney to be bifid with a normally functioning upper moiety and grossly hydronephrotic but normally functioning lower moiety. Right kidney had normal functioning. The split functions obtained from DTPA scan are depicted in Table 1.

Patient planned for open pyeloplasty of the lower moiety. Per operative findings were (Figure 5 and 6) horse shoe kidney, double moiety left kidney, aberrant renal vessel crossing anterior to left lower moiety with dilated proximal part i.e. pelvis and a normal distal part i.e. ureter. Clamping of the crossing

vessel resulted in ischemia of the left lower pole. Patient underwent open dismembered pyeloplasty bringing pelviureteric junction to anterior of the crossing vessel, with placement of a double J stent. The post operative period was uneventful and the stent was removed after 04 weeks.



Fig-1: duplicated left pelvi calyceal system



Fig-2: bifid ureters joing at L3-L4

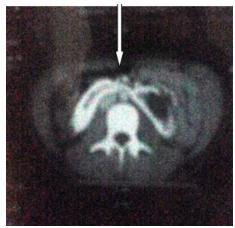


Fig-3: showing isthmus at L3 marked with arrow

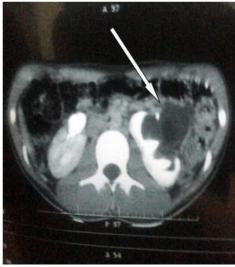


Fig-4: showing hydronephrotic malrotated left kidney marked with arrow

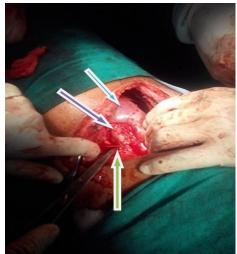


Fig-5: showing left kidney (blue arrow), hydronephrotic left pelvis (purple arrow), aberrant crossing vessel (green arrow)



Fig-6: hooked aberrant crossing vessel (blue arrow) and bifid ureters taped separately (green arrow) with ureters being seen joining distally (black arrow)

Table-1

	Left Kidney	Right Kidney	Total
Split Renal Function	48.23% upper moiety 23.50% Lower Moiety 76.50%	51.77%	100%
Glomerular Filtration (Ml/Min)	48.2	52.8	100

DISCUSSION

Horse shoe kidney is a common congenital anomaly and is a resultant of many diverse teratogenic factors and can be associated with a number of metabolic disorders and Wilm's tumour. Association of horse shoe kidney with duplex collecting system that in itself is a common entity is extremely rare. Christoffersen and his fellows have brought out that this combination is a rare occurrence [6]. Identical views were expressed by Kuzel and his colleagues who described a case of horse shoe kidney with bilateral duplicated collecting system [7]. Pode and his fellows described a case of unilateral triplification of collecting system in a horse shoe kidney [8]. Vipin Tyagi and his associates reported complete unilateral duplication of ureter with horse shoe kidney in 2013 [9]. Pelviureteric junction obstruction in majority has been attributed to abnormal intrinsic musculature [10]. Inferior aberrant vessel has been found to cross pelviureteric junction in 6.8% population. But the incidence of aberrant crossing vessel causing pelviureteric junction obstruction in a fusion anomaly has been placed at a higher level of 25% [11].

CONCLUSION

Only a few cases have been reported describing co-occurrence of horse shoe kidney with a

duplicated collecting system. Our case of horse shoe kidney with double moiety with pelviureteric junction obstruction with an aberrant renal vessel thus makes it a rarest amongst the rare. We aimed for a widely patent, tension free, funnel shaped pelviureteric transition with a dependent drainage through open dismembered pyeloplasty. The patient recovered well and is asymptomatic on 06 months follow up.

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