ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 7, Issue – 5, September – 2022 , Page No. : 97 – 101

Horseshoe Kidney and Its Various Clinical Presentations – Our Institutional Experience

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Citation this Article: Rajkumar R, Periasamy Ponnusamy, Rajasekar Sundaram, Gopu Rakesh Vamsi, "Horseshoe Kidney and Its Various Clinical Presentations – Our Institutional Experience", IJMSIR- September - 2022, Vol – 7, Issue - 5, P. No. 97 – 101.

Type of Publication: Original Research Article

Conflicts of Interest: Nil

Abstract

Objective: Horseshoe kidneys (HSKs) are the most common congenital fusion abnormality; The objective of this study is to analyze various complications, associated anomalies, and need of surgical intervention.

Materials and Methods: This is a retrospective cohort study of ten patients who presented to our institute with the diagnosis of HSKs. The data were analyzed with regard to age at presentation, symptoms, associated anomalies, investigations and surgical intervention.

Results: Ten patients were included in the study in which eight patients were male and two female patients. Nine patients were symptomatic and during workup diagnosed as HSKs, in which six cases presented as pelviureteric junction obstruction (PUJO) and three cases presented with renal calculi. One case was incidentally diagnosed as HSKs while evaluating for road traffic accident (RTA). In our study all patients required surgical intervention.

Conclusions: HSKs the most common congenital renal anomaly and it is almost, always associated with other anomalies which requires prompt and through search, early diagnosis and adequate treatment.

Keywords: Horseshoe Kidneys (HSKs), Pelviureteric Junction obstruction (PUJO), Road traffic accident (RTA), Anderson-Hynes pyeloplasty)

Introduction

Horseshoe kidneys (HSKs) are the most common congenital fusion abnormality; it has a reported incidence of 1 in 400[1] and male to female ratio of 2:1[2]. HSKs occurs due to congenital fusion of lower poles of left and right kidneys and it is associated with various other anomalies. A whole range of anomalies has been known to be associated with increased frequency in symptomatic patients. It is doubtful that this anomaly represents a particular genetic predisposition, but it may be the result of a genetic expression with a low degree of penetrance[3]. The objective of this study is to analyze various complications, associated anomalies, and need of surgical intervention.

Material and Methods

This is a retrospective cohort study of ten patients who presented to our institute with the diagnosis of HSKs.

They were either referred with the diagnosis of HSK or were diagnosed with HSK during investigations for urinary tract infections (UTI) or associated anomalies. one patient was asymptomatic and incidentally diagnosed as HSKs while evaluating for Road Traffic Accident (RTA). Nine patients presented with various symptoms and had a wide variety of urological abnormalities. All patients were extensively worked up with relevant investigations such as U/S, Computer tomography (CT) plain, Contrast Enhanced CT micturating cystourethrography, intravenous pyelography, and isotope scan DTPA (diethylenetriaminepentaacetic acid scan). The data were analyzed with regard to age at presentation, symptoms, associated anomalies, investigations and surgical intervention.

Results

Ten patients were included in the study in which eight patients were male and two female patients. Nine patients were symptomatic and during work up diagnosed as HSKs, in which six cases presented as pelviureteric junction obstruction (PUJO) and three cases presented with Renal calculi. One case was incidentally diagnosed as HSKs while evaluating for road traffic accident (RTA). In our study all patients required surgical intervention. characteristics of these Ten patients were summarized in Table 1. The mean age was 42 years. The most common presenting symptoms were abdominal pain and UTI.

Chart 1: Age distribution

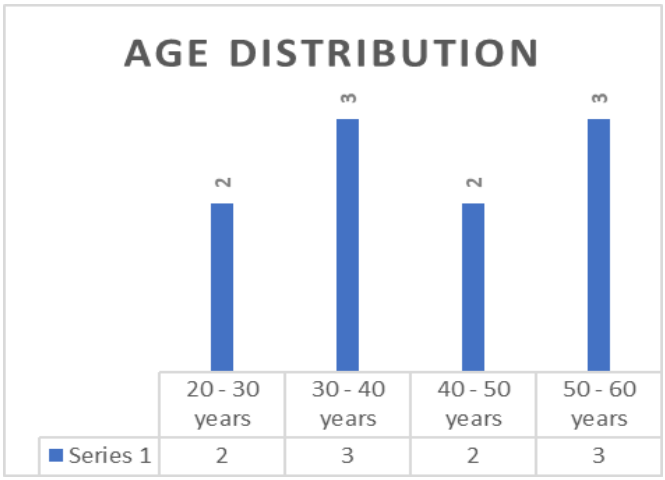

Chart 2: Surgical Procedure

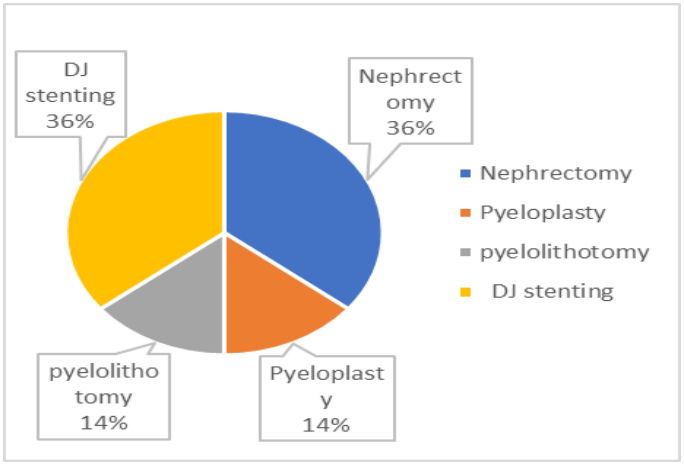

Chart 3: Urological anomalies

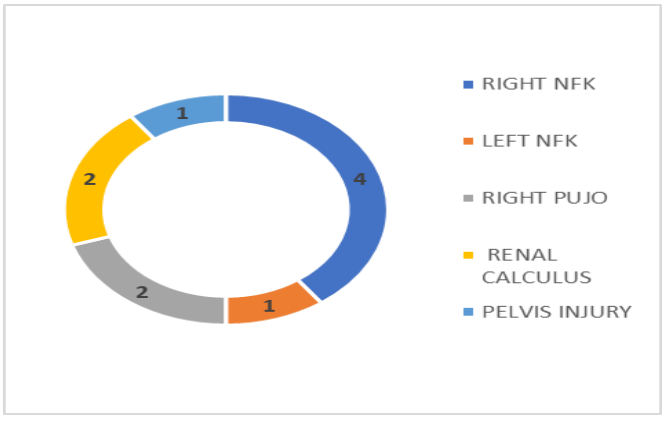

Table 1

Age	Sex	Clinical presentation		Urological anomalies	SURGERY	complications	
33	M	RTA with blunt	Injury abdomen	HSK with pelvis injury	Right DJ stenting	stricture	urethra
43	M	Right loin pain		Right NFK	Right nephrectomy	none	
57	F	Right loin pain		Right NFK	Right nephrectomy	none	
22	M	Right loin pain	Fever	Right PUJO	Right pyeloplasty	none	
36	M	B/L Loin pain	UTI	B/L Renal calculi	PCNL	none	
42	M	Left loin pain		Left NFK	Left nephrectomy	wound	infection
56	M	Right loin pain		Right NFK	Right nephrectomy	none	
25	M	Right loin pain		Right PUJO	Right pyeloplasty	none	
38	M	Left Loin pain		Left Renal calculus	Left pyelolithotomy	none	
60	F	Right loin pain, UTI		Right NFK	Right nephrectomy	none	
Total				10			

Discussion

HSK is the most common renal fusion anomaly. It has a reported incidence of 1 in 400[1.] The two renal masses are joined at the lower poles in more than 90% of cases. One study has shown that fusion of the lower poles occurs very early in gestation when they are in proximity, and this is the result of abnormal migration of nephrogenic cells[4]. Variability of vascular anatomy hints that an anomalous blood supply could be a possible cause of this abnormal renal position[5]. The HSK is usually positioned low in the abdomen. It has been hypothesized that the inferior mesenteric artery obstructs the isthmus and prevents further ascent. Rarely, the isthmus can be posterior to the aorta and/or inferior vena cava[6].

HSKs have been reported in identical twins and several siblings within the same family[7]. The abnormality is more common in males. Diagnosis is primarily by U/S; however, diagnosis can be difficult sometimes and Strauss has reported various findings on U/S which point to the diagnosis of HSKs[8]. DMSA isotope scan can identify the isthmus in 100% of the cases[9]. Computed tomography and magnetic resonance imaging can be used to aid in the diagnosis. Associated anomalies have been frequently found in children diagnosed with HSKs. The most commonly affected organ systems are gastrointestinal, skeletal, cardiovascular and central nervous systems. These findings have been echoed by Boatman *et al*. who reported a 33% incidence of associated anomalies in patients diagnosed with HSKs[10].

HSKs are also associated with Turner's syndrome[11]. Urological abnormalities are also encountered with increasing frequency in patients with HSK. VUR (10–80%) and PUJO (25%) are most commonly associated anomalies found in symptomatic patients[12-14]. Genital

anomalies include hypospadias and undescended testes (4%) bicornuate uterus or septate vagina (7%)[10]. Renal calculi occur commonly and metabolic abnormalities have been implicated in at least one-third of patients with renal calculi[15-16].

In our own series of Ten patients, eight were males. Only one patient was incidentally diagnosed as HSKs while evaluating for RTA and found with right renal collecting system injury and was managed with double J (DJ) stenting. Five patients were diagnosed as Non-functioning kidney (NFK), all patients were in age group of 40-60 years and all underwent open nephrectomies. Two patients in age group (20-25) years were diagnosed as PUJO with decreased functioning capacity of kidney confirmed with DTPA isotope scan they were subjected to pyeloplasty, this can be correlated with early age of presentation where function of kidney is preserved in young patients in comparison with old patients who presented with NFK. we performed Anderson-Hynes pyeloplasty through the extraperitoneal approach in Both patients. We did not feel the need for isthmectomy and lateropexy. both patients had aberrant vessels causing obstruction at the PUJ. Two patients presented with Renal calculi in that one with Bilateral renal calculi, underwent Percutaneous Nephrolithotomy (PCNL) in two sittings, And the other patient was subjected to Transperitoneal Pyelolithotomy due to presence stone near the isthmus.

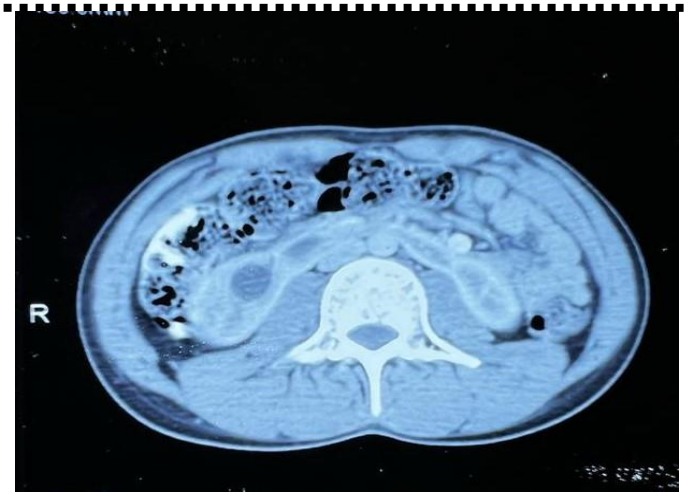

Figure 1: CECT-KUB showing HSK with right PUJO.

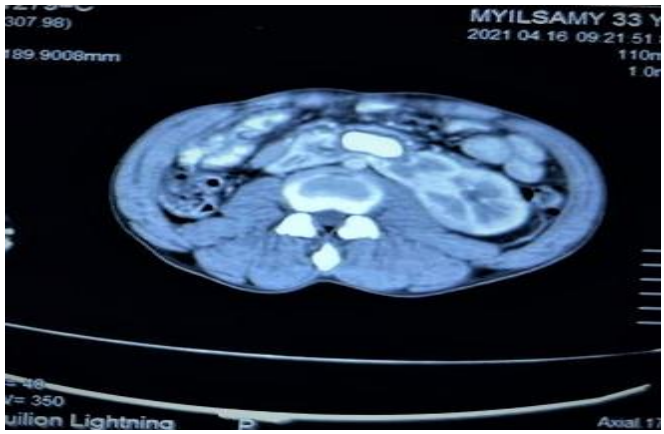

Figure 2: CECT-KUB with HSK with calculus at isthmus.

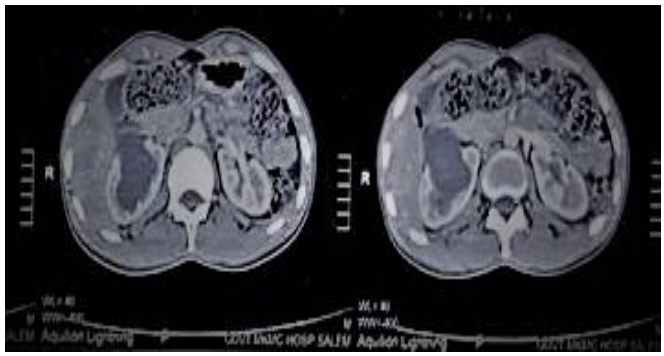

Figure 3: CECT-KUB showing right PUJO.

Conclusions

HSKs the most common congenital renal anomaly and it is almost, always associated with other anomalies which requires prompt and through evaluation, early diagnosis and adequate treatment. Most patients come to attention of the treating physician because they are diagnosed

incidentally and are symptomatic. Asymptomatic patients should be followed up regularly. Symptomatic patients often require surgery for their anomalies. We believe that the diagnosis of HSK should alert the clinician to conduct a thorough search for any associated anomalies. We recommend that all patients need a regular follow-up since complications may develop later in life.

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