Original Article

Horseshoe Kidney and Associated Anomalies: Single Institutional Review of 20 Cases

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Abstract

Background: Horseshoe kidney (HSK) is a common renal fusion anomaly. We undertook this study to discuss various anomalies associated with HSKs. The objective of the study is to study various anomalies associated with HSKs and to assess the need for surgical intervention in patients with these anomalies. **Patients and Methods:** This is a retrospective cohort study of twenty patients who presented to our institute with the diagnosis of HSKs. The data were analysed with regard to age at presentation, symptoms, associated anomalies, investigations and surgical intervention. **Results:** Twenty patients were included in this study. They were referred either with a diagnosis of HSK or were diagnosed during investigations for symptoms and during workup for associated anomalies. Eleven patients were incidentally diagnosed and were asymptomatic, and there were no associated anomalies diagnosed in these patients. Nine patients were symptomatic and were diagnosed with various associated anomalies. Our results concur with recent literature review which suggests that nearly half of the patients with HSKs have associated anomalies. In the present series, all symptomatic patients with associated anomalies required surgical intervention. **Conclusion:** HSKs being a common fusion anomaly, necessitates a prompt and thorough search of the search for any associated anomalies. Many of these anomalies may require surgical intervention.

Keywords: Horseshoe kidney, hydronephrosis, pelviureteric junction obstruction, urinary tract infections, urological anomalies

Introduction

Horseshoe kidney (HSK) is the most common renal fusion anomaly. A whole range of anomalies has been known to be associated with increased frequency in symptomatic patients. Here, we present twenty patients who were diagnosed with HSKs of whom eleven were asymptomatic. They did not have any associated anomalies. They were regularly followed up. Nine were found to have associated anomalies which were diagnosed during the investigation for symptoms or during investigations for associated anomalies. All nine required surgery for their anomalies.

Patients and Methods

This was a retrospective study of twenty patients with HSKs, of whom nine presented with various anomalies. They were either referred with the diagnosis of HSK or were diagnosed with HSK during investigations for urinary tract infections (UTI) or associated anomalies.

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Eleven patients were asymptomatic and were incidentally diagnosed. Investigations performed included urinary tract ultrasound (U/S). Since U/S did not reveal any abnormalities, these patients were on regular follow-up. Nine patients presented with various symptoms and had a wide variety of urological and non-urological anomalies. All symptomatic patients were extensively worked up with relevant investigations such as U/S, micturating cystourethrography, intravenous pyelography and isotope scans (diethylenetriaminepentaacetic acid and dimercaptosuccinic acid scans). All required surgical intervention.

Results

Out of a total of 20 patients, nine were symptomatic. Patient characteristics of these nine children are summarised in

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Table 1. The mean age was 5 years (range 1 day–10 years). There were five males and four females. The most common presenting symptoms were abdominal pain and UTI. Most common associated urological malformation was pelviureteric junction obstruction (PUJO) in three patients. Other urological anomalies included multicystic dysplastic kidney (MCDK), ureterocele, bilateral renal stones [Figure 1], bilateral vesicoureteral reflux (VUR) [Figures 2 and 3] and posterior urethral valves (PU valves). There were three patients with non-urological malformations. All of them were females. They included anorectal malformation (common cloaca), persistent urogenital sinus and myelomeningocele in one each.

Discussion

HSK is the most common renal fusion anomaly. It has a reported incidence of 1 in 400–1 in 1800.[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.[2] Variability of vascular anatomy hints that an anomalous blood supply could be a possible cause of this abnormal renal position.[3] The HSK is usually positioned low in the abdomen. It has been hypothesised 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.[4] There is 3.5% incidence of associated congenital malformation in adults discovered to have HSKs whereas associated malformations are more common in children. HSKs have been reported in identical twins and several siblings within the same family.[5] 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.[6] DMSA isotope scan can identify the isthmus in 100% of the cases.[7] Computed tomography and magnetic resonance imaging are also used to aid in the diagnosis. Associated anomalies have been frequently found in children diagnosed with HSK. The frequency of associated anomalies is higher in the newborn period. Zondek and Zondek found a high incidence of associated malformations (78%) in infants who were stillborn or who died within the 1st year of life.[8] Associated diseases of the HSKs were present in 22%.[8] 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.[9]

HSKs are also associated with Turner's syndrome.[10] 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.[11-13]

About 10% of patients can have ureteral duplication.[8] Kao *et al*. conducted a prospective study to assess the possibility of obstructive hydronephrosis in incidentally diagnosed HSKs by diuretic renogram. The authors concluded that there was a very

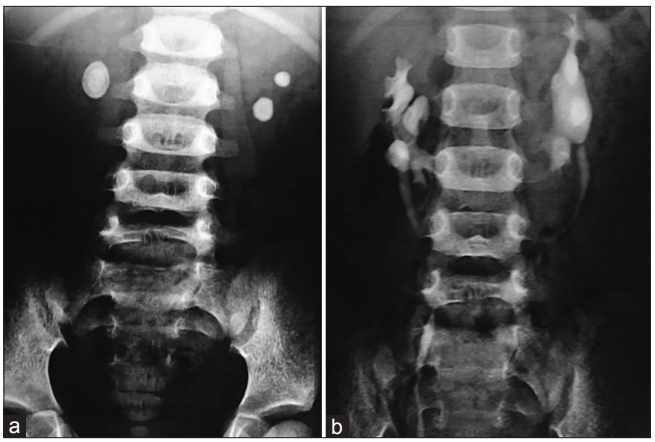

Figure 1: Showing radiopaque calculi in the horseshoe kidney (a) and intravenous pyelography in the same patient (b) in patient 7

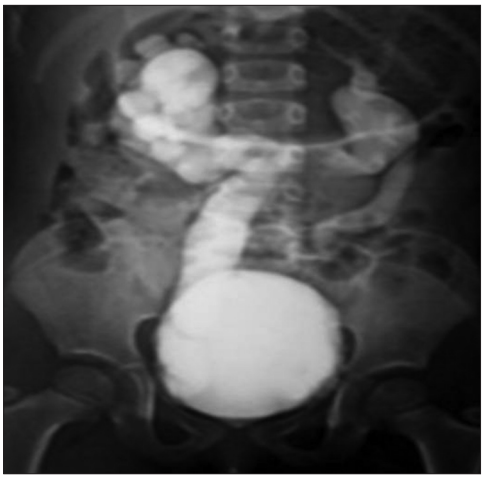

Figure 2: Micturating cystourethrography of patient 4 showing bilateral high-grade vesicoureteral reflux in a horseshoe kidney

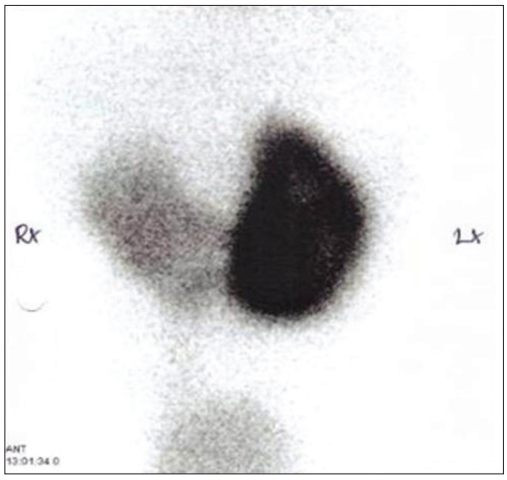

Figure 3: Dimercaptosuccinic acid scan of patient 4 showing horseshoe kidney with differential renal function of 20% of the right moiety and 80% of the left moiety

Table 1: Shows Patient characteristics

Age	Sex	Clinical features	Urological anomalies	Non-urological anomalies	Surgery	Follow-up and complications
1 month	F	Antenatal diagnosis	Right MCDK	none	Right nephro- ureterectomy	4 y none
2 y	F	Urogenital sinus	Right ureterocele	Urogenital sinus	Total urogenital mobilisation Right nephro- ureterectomy	5 y none
10 y	M	Pain left flank	Left PUJO	None	Left pyeloplasty	3 y none
1 day	F	ARM	Bilateral VUR	ARM (common cloaca)	Staged repair Bilateral ureteric reimplantation	3 y constipated
1 y	M	Pain abdomen	Rt PUJO	None	Right pyeloplasty	3 y none
8 y	M	Pain abdomen	Rt PUJO	None	Right pyeloplasty	2 y none
5 y	M	Pain Abdomen UTI	Bilateral renal pelvic stones	None	Bilateral pyelolithotomy	1y none
2½ y	M	Dribbling of urine since birth	Posterior urethral valves	None	PU Valve Fulguration	6 month none
10 y	F	UTI	Bilateral VUR	Myelo-meningocele Neurogenic bladder	Bilateral ureteric reimplantation Mitrofanoff stoma	6 month none

low percentage of obstructive hydronephrosis in incidentally diagnosed HSKs.[7]

Multicystic dysplasia, ureterocele and autosomal dominant polycystic kidney disease have also been r eported.[11,14] Genital anomalies include hypospadias and undescended testes (4%) bicornuate uterus or septate vagina (7%).[9] Renal calculi occur commonly and metabolic abnormalities have been implicated in at least one-third of patients with renal calculi.[15,16]

Cancers have also been reported in horseshoe kidneys with a higher incidence (1.76–7.93 times) as compared to general population.[17,18] Most children with anomalies require surgical intervention for correction of their urological anomalies.[11] Correction of PUJO is the most frequent indication for surgical intervention in a patient with an HSK. In the past, routine division of the isthmus was recommended to avoid the ureter having to cross the isthmus. The current view is that the isthmus does not contribute to obstruction and should not be routinely divided.[19] An extraperitoneal flank approach is utilised for unilateral operations. In those patients requiring bilateral procedures, a transperitoneal approach may be preferable to allow bilateral operations. Donahoe and Hendren reported that the kidneys remain in their original position following symphysiotomy because of fixation by the abnormal vasculature.[20] Panda *et al*. believe that Anderson-Hynes pyeloplasty with isthmectomy and lateropexy is an effective approach for treating PUJO in HSKs.[21]

However, Schuster recommends simple Anderson-Hynes pyeloplasty without symphysiotomy.[19] Most urological anomalies associated with HSKs are treated by minimal invasive procedures.[22,23]

In our own series of nine patients, five were males. Only one patient was diagnosed antenatally with hydronephrosis which later turned out to be MCDK of one moiety. PUJO was the most common associated urological anomaly (three of nine patients). Bilateral VUR (two patients), MCDK, PU valve, ureterocele and bilateral renal calculi were other anomalies detected. For PUJO, we performed Anderson-Hynes pyeloplasty through the extraperitoneal approach in all three patients. We did not feel the need for isthmotomy and lateropexy. Two out of three had aberrant vessels causing obstruction at the PUJ. Two patients with non-functioning kidneys underwent nephrectomies. We had one patient with bilateral renal calculi who underwent transperitoneal bilateral pyelolithotomy. We had three female patients with associated non- urological anomalies, namely, anorectal malformation, urogenital sinus and myelomeningocele who had surgical intervention for these anomalies.

Conclusion

HSK is the most common fusion anomaly of the kidneys and is associated with a wide variety of urological and non-urological anomalies. Most patients come to attention of the treating physician because they are diagnosed incidentally or 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 astute 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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Conflicts of interest

There are no conflicts of interest.

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