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Original Article

HORSESHOE KIDNEY: A MULTIDETECTOR COMPUTED TOMOGRAPHY STUDY

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ABSTRACT

Background and Objective: Horseshoe kidney is the most common renal fusion anomaly with a reported prevalence of 1 in 400 persons with a male to female ratio of 2:1. In many cases its presence may go unnoticed and undiagnosed because the patient may remain asymptomatic throughout life. The objective of our study is to report radiological and anatomical features of horseshoe kidney detected incidentally during retrospective evaluation of multidetector computed tomography scans.

Materials and Methods: Contrast enhanced multidetector computed tomography scans of 682 patients, 355 males and 327 females, were reviewed retrospectively.

Results: Seven cases of horseshoe kidney were detected incidentally, six males and one female, with an incidence of 1.02 %. In all cases, malrotation of the kidneys were observed with the hilum facing anteriorly or anterolaterally. The isthmus was made up of parenchymal tissue in all the cases and the fusion was midline in four cases and lateral in three cases. Horseshoe kidney in all cases was supplied by multiple renal arteries, varying from 3 to 6. In three cases symmetrical arterial supply and in the rest asymmetrical supply was observed. Nephrolithiasis and hydronephrosis were noted in two patients. No other associated congenital anomaly was observed in all seven patients.

Conclusion: Contrast enhanced multidetector computed tomography evaluation of patients with horseshoe kidney provide excellent information about its vascularity, collecting system and other associated conditions.

KEY WORDS: Horseshoe kidney, Renal fusion anomalies, Multidetector computed tomography.

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BACKGROUND

Horseshoe kidney (HSK) is the most common fusion anomaly of the kidney, occurring in 1 per 400 persons with a male to female ratio of 2:1 [1, 2, 3]. Its prevalence in necropsies is reported to be 1 in 1000 cases [4] and in pyelography 1 in 300 cases [5, 6]. It exhibits great variability in morphological position and vascular supply. Such congenital anomalies of kidney is important as they may cause renal failure in middle age group [4] and may be associated with urinary tract obstruction, infection and stone formation [7] and other associated anomalies and complications. Knowledge of horseshoe kidney and its vascularity is important to radiologists to identify all vessels and surgeons who have

to operate for complications [8] or to use a horseshoe kidney for transplantation.

The horseshoe kidney has an incidence of 0.25 % in general population [9] and in many cases its presence may go unnoticed and undiagnosed because the patient may remain asymptomatic throughout life. The condition may be missed in sonographic evaluation especially if the isthmus is composed of only fibrous tissue and not parenchymal tissue [10]. Recently it was suggested that the Multidetector row Computed Tomography (MDCT) urography is the modality of choice comprehensively evaluating anatomical features of renal fusion anomalies in a single examination [11]. We present the anatomical and radiological features of horseshoe kidneys detected during retrospective review of MDCT scans of 682 patients.

MATERIALS AND METHODS

The present retrospective review was done in a single diagnostic centre during the period from October 2012 to June 2014. MDCT angiography scans of 682 patients (355 males; 327 females) were reviewed and the horseshoe kidney was detected in seven cases. All the patients underwent contrast enhanced computed tomography (CECT) by 64 channel scanner (GE optima – 60) for suspected pathologies of hepatobiliary, renal, pancreatic and gastrointestinal systems and received 85-100 ml of non-ionic contrast (Omnipaque, 300mg I/ml) at the rate of 4 ml/s intravenously. The diagnostic centre routinely obtains written informed consent from the patients before contrast injection. Sections of 0.625 mm thickness were obtained from diaphragm to upper part of thigh and delayed phase scans were also obtained. The scans were analysed in a separate work station (AW volume share 4.5) with multiplanar reformatting capability and maximum intensity projection (MIP) of axial, coronal and sagittal sections and volume rendered (VR) images were obtained.

OBSERVATIONS

In this retrospective study of 682 MDCT scans of 355 males and 327 females, we observed seven cases of horseshoe kidney (6 males and 1 female; 1.02 %). In all the cases the lower poles of both the kidneys were connected by an isthmus and were classified as Type A (b) according to Matsumoto et al [12]. The isthmus was midline in four cases (Fig.1B,Symmetrical horseshoe kidney) and lateral in three cases (Fig.6A, Asymmetrical horseshoe kidney). All horseshoe kidneys were located lower than usual site for normal kidneys. All cases of horseshoe kidneys were supplied by the several renal arteries varying from 3 to 6 arteries and the pattern of these arteries was almost similar to Graves' classification or a modification of this classification [13]. Lower right renal arteries in four cases and the main right renal artery in one case were found precaval crossing anterior to inferior vena cava before reaching the renal hilum (Figures 1C, 2C, 3B, 3C, 4C and 5C). No venous abnormality was noted contrary to Ichikawa et al who stated that major venous anomalies are frequently associated with horseshoe kidneys [14]. Right hilum was directed anterolaterally in 5 cases and similarly left hilum was anterolateral in 5 cases.

In three cases symmetrical supply was observed and in one of these three, each half received three renal arteries (Fig.1B). In rest of the two cases each half was supplied by two renal arteries (Fig. 2B and 3A). In one case left half was supplied by single renal artery and the right half was supplied by four renal arteries (Fig. 4B). In two asymmetrical cases right half was

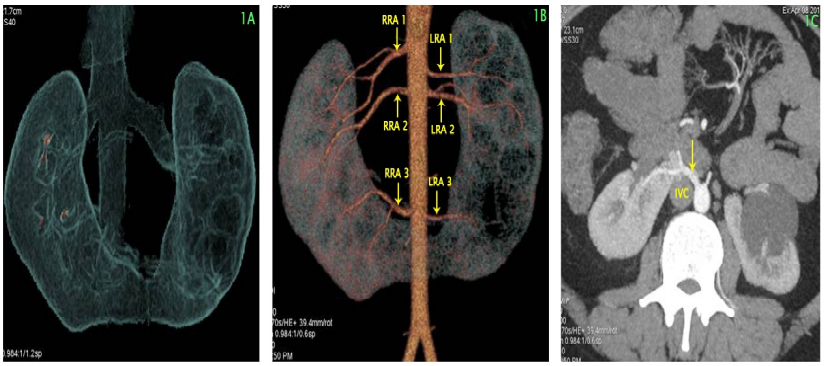


Fig. 1: VR (A, B) and axial MIP (C) images of a 33 year old male. 1-A show single renal vein draining each kidney into inferior vena cava. 1-B showing symmetrical arterial pattern with three renal arteries on each side. 1-C show precaval right renal artery (arrow) passing anterior to IVC.

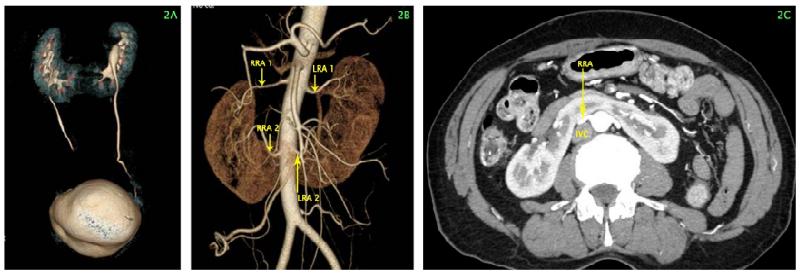


Fig. 2: Female aged 70 years (A) Urography showing anterolateral hila. (B) VR image showing symmetrical arterial pattern with each half supplied by two renal arteries. (C) Axial MIP image showing precaval course of lower RRA (RRA-2).

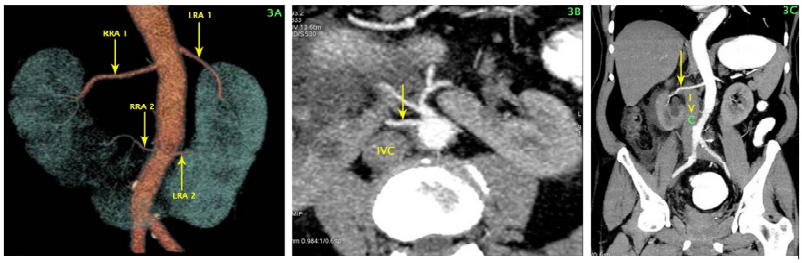


Fig. 3: Male aged 60 years (A) VR image showing symmetrical arterial supply of horseshoe kidney by two renal arteries on each side. Axial (B) and Coronal (C) images showing precaval right renal artery (arrow). Note the lower left renal artery giving a branch to lower pole of right kidney and isthmus (B).

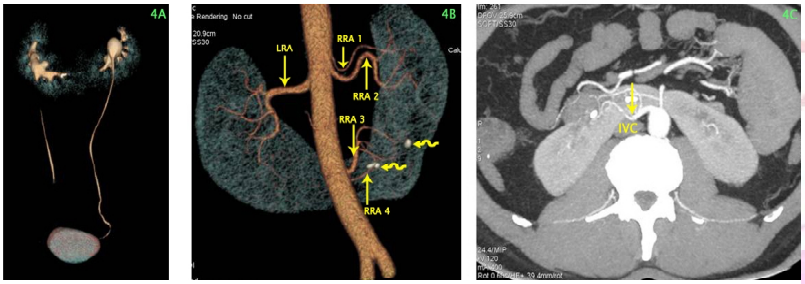


Fig. 4: Male aged 52 years (A) urography; (B) VR image posterior view showing asymmetrical arterial supply by a single renal artery on the left side and four arteries on the right side. Small calculi are observed in lower part of right kidney; (C) Axial image showing precaval course of lower right renal artery (arrow).

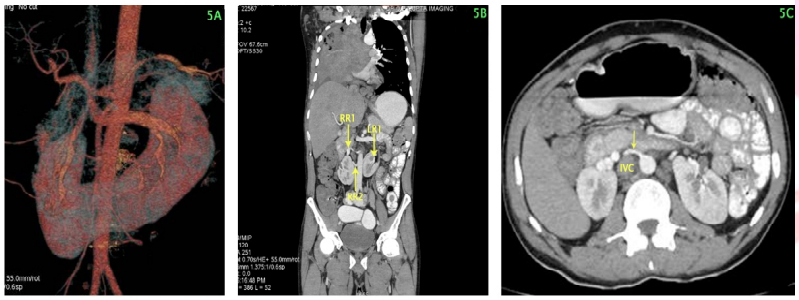


Fig. 5: Male aged 35 years. (A) VR image sowing single renal vein draining each half of the horseshoe kidney. (B) Coronal image showing asymmetrical arterial supply by a single renal artery on the left side and two arteries on the right side. (C) Axial MIP image showing precaval course of right renal artery.

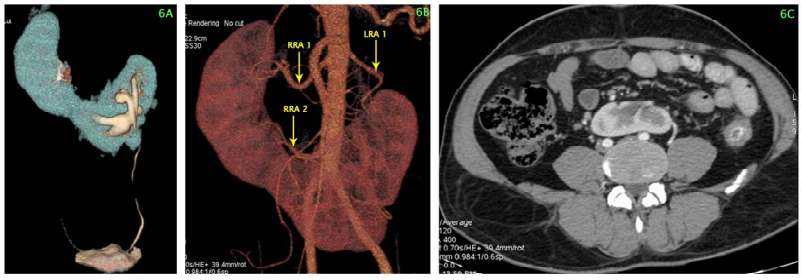


Fig. 6: Male aged 42 years (A) Urography (B) VR image showing asymmetrical arterial supply with a single artery on the left side and two arteries on the right side. (C) Axial image showing the position of isthmus in front of common iliac arteries.

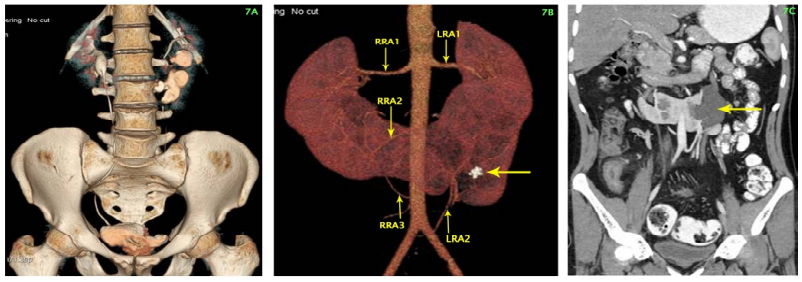


Fig. 7: Male aged 22 years. (A) Urography (B) VR image showing two renal arteries on the left side and three arteries on the right side. The lower renal arteries of both sides arise below the isthmus, the lower right renal artery (RRA3) from aorta and lower left renal artery (LRA2) from left common iliac artery. Note the presence of a calculus at the lower pole of left kidney (Large arrow) (C) Coronal MIP image showing dilated renal pelvis on the left side.

supplied by two renal arteries and left half was supplied by single renal artery (Fig. 5B,6B). In the remaining asymmetrical case three right renal arteries and two left renal arteries were present. Lower most branches on both sides were arising below the isthmus, on right side from the aorta and on left side from common iliac artery (Fig. 7B). Renal calculi were observed in two cases. (Fig. 4B,7B). Hydronephrosis was seen in two cases (Fig 1C, 7C).

DISCUSSION

Renal fusion anomaly occurs when a portion of one kidney fuses with its opposite mate. The most common renal fusion anomaly is horseshoe kidney involving abnormal migration of both the kidneys (ectopy) resulting in fusion, usually of lower poles of the kidneys. Horseshoe kidney differs from crossed renal ectopia, which involves abnormal migration of one kidney across the midline and fusion with the normally located uncrossed kidney. In crossed renal ectopia the ureter from the crossed kidney opens at its normal position into the urinary bladder after crossing the midline. In horseshoe kidney the ureters from the right and left renal moieties do not cross the midline and open normally into the bladder.

Incidence: Horseshoe kidney is the most common congenital fusion anomaly of the kidney and its incidence varies from 1/400 to 1/800 [1]. The incidence at necropsy has been reported as 1/1000 and by pyelography as 1 / 300 [4, 5, 6]. Its reported prevalence in general population is 0.25 % and occurs more commonly in males [9] Based on the data from birth defect registry the reported incidence varies from 0.4 to 1.6 per 10,000 live births. An incidence of 1in 666 cases was found at a single institution after analyzing radiographic data of 15320 patients (23 cases of horseshoe kidney; 16 males and 7 females) [15]. Glodny et al estimated the period prevalence rates for horseshoe kidney in adults examined by sonography as 1: 708 and by CT as 1:474 [2]. In a cross sectional study on 12000 patients using ultrasonography and contrast urography, only 4 cases of horseshoe kidney was found [16]. In a retrospective review of 22 patients with renal anomalies, horseshoe kidney was found in 14 patients (12 men, 2 women) [17]. Studying Tc 99m DMSA renal scans of 400 children (mean age of 5.6 years), horseshoe kidney was seen in 4 cases (1.0%) [18]. In the present study we detected horseshoe kidney in 7 out of 682 patients (1.02%; 6 males,1 female) with an estimated incidence of 1in 97 cases. The prevalence is high probably because the subjects included were patients undergoing MDCT evaluation and not general population. The gender ratio was also high with 6 males to 1 female and similar such observation was reported by others also [17]. In contrast, an ultrasonographic study from Nepal has reported an incidence of 1 in 516 (61 horseshoe kidneys out of 31498 patients screened, 0.2%) with male to female ratio of 1:2 (20 males.41 females) which is just the reverse of ratio reported in the literature and this could be due to more number of females undergoing ultrasonography [19].

Morphology: Horseshoe kidney occurs due to abnormal migration and fusion of two kidneys which are also malrotated. Fusion is thought to occur before the kidneys ascend from the pelvis to their normal dorsolumbar position. It is generally accepted that the ascent of the horseshoe kidney is prevented by the origin of inferior mesenteric artery and hence are located at a lower level. Matsumoto et al [12] classified horseshoe kidneys into following types based on the position and features of isthmus: Type A (a) fusion of superior poles, Type A (b) fusion of inferior poles (most common),Type B (a) fusion by fibrous tissue, Type B (b) fusion directly and Type B (c) fusion by mediators. In more than 90% cases the inferior poles fuse to form horseshoe kidney. It is suggested that in MDCT scans if the fusion site of the kidneys showed the same contrast enhancement as the kidney itself it could be classified as consisting of renal parenchymal tissue. If the enhancement was lower the fusion site could be classified as fibrous [2]. All our cases belong to Type A (b) (fusion of lower poles) and B (b) (parenchymal isthmus) category. Hilum of kidneys is directed anterolaterally in most cases (10 hila in 14 are directed anterolaterally) because of malrotation. The isthmus of horseshoe kidney may lie over the midline (Symmetrical) or lateral to the midline (Asymmetrical) [20]. In the present study midline fusion was noted in 4 cases and lateral fusion in 3 cases. Glodny et al have observed midline fusion in 40% cases, left lateral in 38% and right lateral fusion in 22% cases [2].

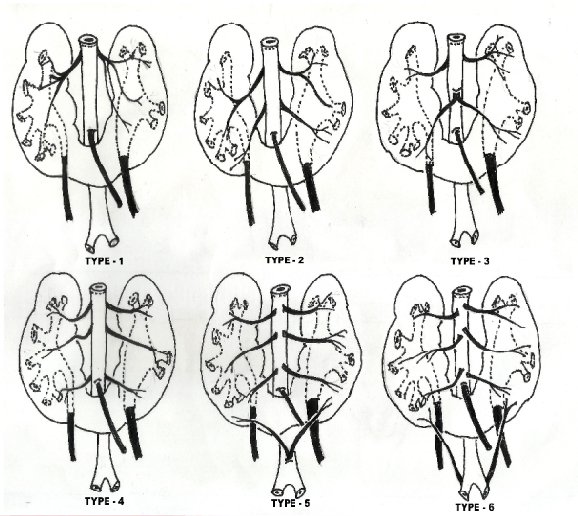
Vascular Supply: Horseshoe kidneys fuse when they are close to each other in the pelvic cavity and may acquire branches from distal aorta, common iliac or hypogastric (internal iliac) arteries [21]. In the process of ascent of the kidney during development, the blood supply continuously changes with the generation of

new arteries and degeneration of old arteries [22], so the number of renal arteries and trajectory of ureter can vary [23]. In fact single renal artery on each side can only be found for both kidneys in less than30% cases of horseshoe kidney. Papin's autopsy study of 139 horseshoe kidneys served as the basis for a classification system consisting of three groups. Group 1 kidneys have normal renal arteries and account for 20% of all horseshoe kidneys; Group 2 kidneys have three to five renal arteries and account for 66% of cases; Group 3 have more than five renal arteries and account for 14% [24].In the present study only one case belongs to Group 3 (Fig 1B) and the rest to group 2 according to this classification.

Graves [13] described 6 basic patterns of arteries in horseshoe kidney studied by means of resin cast (Fig. 8) He established that each artery supplies its own area, with no collateral circulation between the segments and the pattern is symmetrical. Pattern of blood supply may be similar to that of normal kidney with single artery supplying upper, middle and lower segments (Type 1). Upper and middle segments of each kidney may be supplied by a single artery, with a vessel from aorta entering each lower segment (Type 2). Sometime the arteries to lower segment arise from aorta by a common trunk (Type 3). All three segments are supplied by separate arteries arising from aorta (Type 4). The fused segment (isthmus) may also be supplied by arteries which arise above or below the isthmus , these may be unilateral or bilateral and may originate from the aorta independently or by a common trunk (Type 5). Finally the fused lower segment may be supplied on one or both sides by branch originating from the common iliac or rarely from hypogastric (internal iliac) or median sacral artery (Type 6). All the six patterns depict symmetrical supply to both renal moieties of the horseshoe kidney.

In two of our cases symmetrical pattern having two renal arteries on each side similar to Type 2 of Graves classification was observed (Fig. 2 B and 3 A) and another symmetrical case, having three renal arteries on each side, is similar to Type 4 of Graves classification in which all three segments of each half is supplied by individual renal artery (Fig. 1B). In one symmetrical pattern,

Fig. 8: Six types of arterial vascularisation pattern of horseshoe kidney. Adopted from Graves [13].



the lower left renal artery was giving a branch to the lower pole of right kidney moiety (Fig. 3B). In two asymmetrical cases left half is supplied by a single renal artery similar to Type-1 and the right half having two renal arteries is similar to Type-2 category(Fig. 5-B, 6-B). In another case where the two left renal arteries and three right renal arteries were present and lower most renal arteries arising below isthmus on right side from aorta and on left side from left common iliac artery, is a modification of Type-6 category of Graves (Fig 7- B). One case having one renal artery on left side and four renal arteries on right side, arterial pattern on the left side is similar to Type 1 and on the right side similar to Type 4 (Fig. 4B).

In four cases lower right renal artery and in one case main right renal artery having a precaval course was observed. Normally main and accessory right renal arteries pass posterior to IVC to reach right renal hilum and only the right gonadal artery cross in front of IVC. Analysing the arterial pattern of 90 horseshoe kidneys Glodny et al have observed that the second artery on the right side has a precaval course [2]. Presence of precaval right renal artery may be mistaken for gonadal artery especially in radiological imaging.

Development: Kidneys begin their development in the sacral region due to the inductive interaction between metanephric blastema and the ureteric bud and then ascend to reach their normal position. There are two theories about the embryogenesis of the horseshoe kidney and

the fusion is thought to occur as the kidneys ascend from their pelvic position. The classical theory of mechanical fusion proposes that when the kidneys pass through the fork between the two umbilical arteries, the lower poles will come into contact with each other and fuse due to any positional change in the umbilical arteries. Fusion can also occur due to lateral flexion and rotation of the caudal end in the 4-5 mm embryo disturbing the relative position of the nephrogenic blastema and ureteric buds [20]. More recently, it has been proposed that the horseshoe kidney is the result of a teratogenic event that involves abnormal migration of cells that form the isthmus [25]. It has been suggested that nephrogenic cells that have migrated across the primitive streak in the final phase of gastrulation and which arise from the posterior nephrogenic area of the epiblast are responsible for formation of parenchymal isthmus of horseshoe kidney [26]. Notochord is not necessary for nephrogenesis but is required for correct positioning of the metanephric kidney, while the axial sonic hedgehog gene signal is critical for kidney positioning along the mediolateral axis. Any disruption of this gene or notochord will result in kidney fusions [27]. Mc Pherson calculated empirical risk figures for first degree relatives based on the family history given by the subjects with horseshoe kidney and found three families in which horseshoe kidney recurred in two generations, suggesting that this condition may occur as a previously undescribed autosomal dominant condition [28].

Associated Anomalies: Horseshoe kidney may be associated with hydronephrosis and renal calculi [29,30]. Renal calculi were observed in two cases, on the right side in one (Fig. 4-B) and on the left side in another case (Fig. 7-B). Hydronephrosis was seen in two cases (Fig.1C, 7C). Horseshoe kidney can also be associated with congenital anomalies and the system most commonly affected is skeletal – hemivertebra, scoliosis, rib defect, club foot, congenital hip dislocation ; cardiovascular-Ventricular Septal Defect ;GIT- anorectal malformation, malrotation, and Meckel's diverticulum; CNS- neural tube defect; Genito-urinary- vesicoureteral reflex, duplication of ureter, hypospadias, undescended testis, bicornuate or septate uterus [8]. No such anomalies were noted in the present study. Thirty-three per cent of patients with Turner syndrome present some renal malformation and 7.1% of these patients present horseshoe kidney [31]. Horse-shoe kidney is also associated with a twofold increased relative risk of Wilm's tumor, a three to fourfold increased relative risk of transitional cell carcinoma, and a markedly increased relative risk (62-fold) of primary renal carcinoid tumour [32, 33].

CONCLUSION

Horseshoe kidney is one of the most common renal fusion anomaly. The condition may remain asymptomatic throughout life and go undiagnosed and therefore the exact incidence of this anomaly in general population cannot be estimated. This anomaly may be associated with other congenital anomalies and complications. MDCT evaluation of patients with horseshoe kidney provide excellent information about its vascularity, collecting system and other associated conditions which is invaluable for transplant surgeons, laparoscopic surgeons and interventional radiologists.

List of Abbreviations:

MDCT: Multidetector Computed Tomography
HSK: Horseshoe kidney
IVC: Inferior Vena Cava
LRA: Left Renal Artery
RRA: Right Renal Artery
Tc 99m DMSA: Technetium 99 Dimercapto succinic acid
MIP: Maximum intensity projection
VR: Volume rendered

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Conflicts of Interests: None

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