An Autopsy Case of Horseshoe Kidney

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Introduction

Among various organ malformations of man, those in the urinary system appear rather frequently, and the fused kidney, mostly appearing as the horseshoe kidney with the fusion of the lower pole, is common. The fused kidney appears in 1 of 400 subjects, twice as frequently in males as in females (Warkany, 1971). According to the reports on Japanese so far, the incidence ranges between 0.22 and 0.28% (Taira et al., 1951), with a male to female difference similar to that seen in Europeans. We have observed a case of horseshoe kidney among the cadavers used for anatomical research in 1975 as reported below.

Findings

This fused kidney was found in the cadaver of a 72 year old male (cause of death, cerebral apoplexy). The right and left kidneys were fused between their lower poles, forming a horseshoe kidney.

1. Position and shape of the kidney

The upper end of the right kidney is at the level of the middle of the first lumbar vertebral body and the upper end of the left kidney is slightly lower than the right, at the level of the upper end of the second lumbar vertebral body. The fusion between the right and left kidneys has taken place in front of the abdominal aorta and inferior vena cava, as a bridge-like connection between these two (tentatively called the isthmus) (Figs. 1, 2). The lower end portion of the right kidney slanted towards the anterior and inferior direction and the lower end of the left kidney also slanted to the ventral inferior and medial

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direction, so that the long axis of neither the right nor the left kidney is parallel to the spine, with asymmetry in addition. Therefore, this horseshoe kidney, as a whole, appears as a slightly opened upward U shape. Both kidneys have slightly rotated towards the lateral direction, so that the renal hilus is directed anteriorly.

i) Right kidney: The length of the right kidney proper is 10.3 cm, width 5.0 cm and thickness 3.7 cm. The position and mild abnormal rotation were described above (Fig. 2).

In the outside appearance, the posterior surface is flat and the lateral surface is slightly swollen, with an approximately round tumor of 5.3 cm diameter from the lower end of the kidney. Since this specimen was fixed insufficiently, histological examination of this tumor was not possible. Except for this swelling, the right kidney, as a whole, appeared lens bean-like, with smooth surface. The inferior medial portion of the kidney, on the other hand, shows a transition to the isthmus which has become flat antero-posteriorly. The renal hilus is normally formed (length 4.2 cm, width 1.5 cm), but the artery entering there shows considerable deviation from the normal appearance (subsequently described). The renal pelvis consists of two large renal calyces, the superior and inferior ones, uniting at the hilus. The ureter continuing from the renal pelvis descends in front of the right kidney along the borderline with the isthmus after leaving the hilus.

ii) Left kidney: The left kidney proper is considerably deformed with the length of 10.5 cm, width 5.5 cm and thickness 2.5 cm. It is situated slightly lower than the right kidney as described above, and the hilus is somewhat rotated anteriorly (Fig. 1). In the outside appearance, the left kidney is somewhat flat antero-posteriorly. The superior medial (originally medial) and inferior lateral (originally lateral) margins form an approximately straight line giving a square appearance. The hilus is wide, giving an appearance of a long square with a length of 5.2 cm and the width of 3.2 cm. The development of hilar lips is poor. Renal sinus is not formed as in normal form with an exposure of several abnormal arteries and veins besides the small renal calyx, giving an appearance of renal hilar hypoplasia. The renal pelvis is distinguished into two large renal calyces in outline. Several upper and lower small calyces are completely exposed, and the ptotic renal pelvis formed by confluence of these protrudes from the shallow renal hilus, with a transition to the ureter at the inferior medial angle of the renal hilus. The ureter thereafter runs obliquely to the inferior medial portion of the kidney leaving a shallow pressure groove on the anterior surface of the kidney.

iii) Isthmus: A belt-like portion connecting the lower poles of right and left kidneys called the isthmus has a length of 5.2 cm in the long axis of the middle portion. The width is 3.1 cm at the central
portion and the upper margin gives an arch-like appearance with a slight concavity to the inferior direction, while the lower margin is almost rectilinear. Since the abdominal aorta runs behind the left half of the isthmus, a marked pressure groove by the aorta is found on the posterior surface of the isthmus. On the anterior surface of the isthmus, one pressure groove is found on the right end and another on the left end, by the right ureter and inferior mesenteric artery respectively. In the approximate center and the left lower end, two arterial branches originating from the anomalously running middle sacral artery supply (described subsequently).

2. Arterial supply to the horseshoe kidney

Into this horseshoe kidney including the original right and left kidneys and isthmus, the entrance of 5 arteries, 3 from the abdominal aorta, 1 from the inferior mesenteric artery, and one abnormal artery which is regarded as the middle sacral artery described in ii) are observed (Figs. 2, 3).

i) Right renal artery: Two arteries arising from the abdominal aorta at the level of the upper end of the body of the first lumbar vertebra and running tightly adjacent to each other, enter the hilus of the right kidney. The inferior one is tentatively called the first and the superior one the second renal artery. Both the first (Fig. 3; 1) and the second (Fig. 3; 2) renal arteries reach the right renal hilus running approximately parallel to each other behind the inferior vena cava. During the course, the inferior phrenic artery (Fig. 3; Ip) and the inferior suprarenal artery (Fig. 3; Sa) branch off from the second renal artery.

The first renal artery (with 5.0 mm diameter at the origin) is divided into the superior and inferior branches near the renal hilus, reaching the right kidney behind the renal vein and ureter. These arteries are further divided into small branches as shown in Figure 3. According to these findings, the first renal artery probably distributes in the anterior part of the upper one-half of the right kidney, the posterior half of the right kidney and the posterior part of the right upper end of the isthmus.

The second renal artery (with 2.5 mm diameter at the origin) flexes inferiorly near the renal hilus running in front of the first renal artery crossing it, and is divided into two branches reaching the renal hilus more anteriorly than the renal vein. These branches are further divided into several branches. The second renal artery mainly distributes over the lower one-half, its anterior part of the right kidney and, in part, in the anterior region of the right upper end of the isthmus. In view of the course and branching described above, the first renal artery appears to correspond to the normal right renal artery.
ii) Left renal artery: One ordinary left renal artery directly arising from the abdominal aorta (Fig. 3; 3) and a short, thin branch from the inferior mesenteric artery (Fig. 3; 4) enter the left kidney along with an extra artery with abnormal origin (Fig. 3; 5).

The left renal artery with the diameter at the origin of 7.0 mm arises from the left wall of the abdominal aorta at the approximately same level as the origin of the right first renal artery and runs towards the renal hilus behind the renal vein, giving off the left inferior suprarenal artery in its course. After reaching the renal hilus, it is divided into three main branches at the superior medial corner. These branches reach the deep portion of the upper half of the kidney from the surface, and in part from behind the renal vein deeply into the posterior part, even reaching the inferior lateral region.

The short and thin branch with a diameter of 1.5 mm arising from the inferior mesenteric artery (Fig. 3; Im) at 3 cm from its origin enters the renal hilus from the medial side of the kidney (Fig. 3; 4).

In the left kidney, furthermore, an extra abnormal artery described above is found. In the present case, the terminal branch continuous from the abdominal aorta or the middle sacral artery is not found at the normal position. Instead, an artery with a diameter of 4.5 mm is found to divide into the two and distribute over the left kidney (left branch) and the isthmus (right branch) after originating from a short common trunk probably representing the upward deflection of the middle sacral artery.

The left branch (Fig. 3; 5l) runs to the left superior lateral direction after branching, entering the left kidney from the lower pole and distributing over the lower one-third of the left kidney after being divided into two branches, the superficial and profound ones.

The right branch (Fig. 3; 5r) distributing over the isthmus is divided into a branch running towards the right end of the isthmus and the another one reaching its approximate center. These arteries are probably the ones to perfuse the isthmus.

3. Veins to the horseshoe kidney

The renal veins leave the renal hilus as two veins, superior and inferior, from the right and the left kidney respectively (Fig. 1). On the right, the lower vein receives the right testicular vein. On the left, two renal veins unite on the way to form one vein, which receives the left suprarenal vein. Both the right and left renal veins enter the inferior vena cava separately.
Discussion

The development of human renal anlage begins around the 4th week of embryonal life when the ureteric bud originating from the distal portion of the mesonephric duct reaches the metanephric blastema occurring on the posterior wall of the coelomic cavity near the lower end of the mesonephros. In the 5th week, the ureteric bud begins to ascend towards the lumbar region with gradual, internal rotation. Around the 8th week of embryonic development, the internal rotation is completed, and the anlage of the metanephros (permanent kidney) is formed facing the renal hilus to the medial direction (Patten, 1953).

As to the time of development of the malformation and the cause of horseshoe kidney, various theories have been advanced. In most of the cases of this malformation, the hilus is abnormally rotated towards the ventral side and this represents one of the reasons for assuming that the fusion probably takes place either prior to or in the very beginning of the fifth week of embryonic development, when the rotation begins. As to the cause of the fusion of the right and left kidneys, the hypothesis of slight central displacement of the bilateral ureteric buds and subsequent growth (Willis, 1962), the hypothesis of mutual approach and fusion of the right and left metanephric blastemas at the time of arising from the sacral area (Tuchmann-D., et al., 1972, Patten, 1953), and the hypothesis of mechanical compression by the surrounding organ leading to fusion (Budde, 1913, Muthmann, 1907) are proposed.

In our case, no findings have ever suggested the time and mechanism of development of the malformation. In view of the presence of abnormalities in rotation, and the absence of the normally descending medial sacral artery replaced by an ascending artery distributing over the isthmus and the left kidney, it might be indicated that the fusion occurred at a stage with the position of the renal anlage below the sacrum.

The fused kidney has been classified according to the site and degree of fusion into various types. In the present case, the lower poles of bilateral kidneys are connected in a belt-like fashion, representing the most frequent type (Warkany, 1971).

According to previous reports, the horseshoe kidney has the following characteristics: 1. Lower position than the normal kidney, 2. Generally more ventral direction of the renal hilus than the normal kidney, 3. Passage of the ureter on the ventral side of the kidney, 4. Presence of abnormal blood vessels (Adachi, 1928) and 5. Frequent abnormalities in other organs. In our case, all these characteristics were present except for 5.

The present case is characterized by the presence of a considerable number of abnormal blood vessels. As a rare case, the middle sacral
artery was absent in normal position, instead, the artery probably corresponding to this ascended, entering the left kidney and isthmus. Entrance of a branch of the inferior mesenteric artery to the left kidney represents another one of the characteristics.

Among the cases of horseshoe kidney reported in Japan, abnormal arteries derived from the abdominal aorta were observed in 9 cases reported by Takahashi (1934), Nagatomo (1937), Mori et al. (1941), Takeda (1947), Nakayama et al. (1950), Kobayashi (1954), Honjin et al. (1955) and Matsumoto et al. (1963). Arteries arising from artery other than the abdominal aorta were observed in the cases reported by Aoki (1943) and by Matsuo (1944).

In the present case, a typical malformation with many characteristics of horseshoe kidney was found, with the absence of malformations in other organs permitting survival up to the high age of 72 years. Though the medical and social history during life is unknown, no disturbance ascribable to this malformation apparently occurred.

Summary

1) The horseshoe kidney with a fusion of bilateral lower poles forming the so-called isthmus was found in the cadaver of 72 year old male.

2) The position of this kidney was lower than the normal one by one vertebral body height on the right and by two vertebral body height on the left. Bilateral ureters descended the ventral side of the isthmus.

3) Abnormal arteries in this case included the a) two arteries distributing over the right kidney from the abdominal aorta, b) one artery from the abdominal aorta, a small branch from the inferior mesenteric artery and one branch (left) from the middle sacral artery distributing over the left kidney, and c) one branch (right) from the middle sacral artery distributing over the isthmus.

4) Both renal hili faced the ventral side, with a marked hypoplasia of the left hilus.

5) No malformations other than the horseshoe kidney were in other organs.

References

4) Honjin, R. and O'sugi, H.: A case of the horseshoe kidney, Jüzenkai Z., 57: 2160-
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Explanation of Figures

Abbreviations (Figs. 1-3)

Plate I

Fig. 1. Ventral view of the horseshoe kidney and its surroundings in outline found in a male cadaver at the age of 72.
Plate II

Fig. 2. The horseshoe kidney and its arterial system (details in Fig. 3); a view from the ventral side.
Fig. 2

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Plate III

Fig. 3. A drawing of the arteries distributing over the bilateral kidneys and isthmus.
1, 2: Right renal artery; 1 (First renal artery) and 2 (Second renal artery)
3: Left renal artery
4: An abnormal artery from the inferior mesenteric artery (Im)
5: An abnormal artery probably corresponding to the middle sacral artery,
   its right branch (5r) and left branch (5l).
Fig. 3