The cake or lump kidney is a rare form of fusion. The pancake kidney is always found in the pelvic cavity, in the greater or lesser pelvis. However, we report a case of pancake kidney which was found inside abdominal cavity while investigating 32-year-old male for lower limb pain. CT scan with IV contrast reveals ‘fused mass situated in the right side of abdominal cavity with course uncrossed ureters opening separately into the urinary bladder found incidentally with patient work up’. Pancake kidney in this case was managed conservatively without any complication. This malformation is usually present in pelvic cavity, so blood supply is almost constant but in rare cases might be different and follows location of this anomaly.

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Introduction

Crossed renal ectopia is the second most common fusion anomaly after horseshoe kidney. It occurs in 1:7000 with an M:F ratio of 2:1. Both kidneys are located on the same side and may occur with fusion (85%), without fusion (<1%), or extremely rarely be solitary or bilateral. The left kidney is three times more likely to migrate to the right than vice versa. Other fusion anomalies are extremely rare and include lump kidney, sigmoid kidney, disk kidney and L-shaped kidney. The pancake kidney was first described by Looney and Dodd. The ‘pancake kidney’ is a congenital anomaly that exhibits fused pelvic kidneys. In this rare anomaly, there is extensive fusion of the superior and inferior poles of both kidneys across the entire medial aspect, looking like a pancake. The pancake kidney has several other names, such as the cake, disc, doughnut and shield kidney, because it produces a ring- or doughnut-shaped mass.

Case report

A 32-year-old man, previously well, presented to our outpatient clinic with lower limb pain. Pain was located in right side of abdomen, intermittent, described as discomfort and persisted for the previous 3 months prior to presentation. The pain was not associated with urinary symptoms. Physical examination was unremarkable. Laboratory investigation showed microscopic hematuria and normal creatinine level. Abdominal ultrasound and CT scan with IV contrast showed fused lobulated mass situated in the right side of abdomen extending from L2 to L4 with course uncrossed ureters opening separately into the urinary bladder (Fig. 1).

CT scan with IV contrasts with urography phase show complex vascularity of the kidney (Fig. 2). A 3D reconstruction of the CT scan with urogram phase (Fig. 3).

MAG 3 scan was done to rule out obstruction and showed two well-formed renal units with split function as follows; Right kidney: 46.9% and Left kidney: 53.1%. Diuretic phase showed no evidence of obstruction.
After being thoroughly investigated the patient was counseled about his condition and the possible future complications and treated conservatively with regular follow up.

Discussion

The embryological development of the kidney results from the interaction between the mesonephric duct-derived ureteric bud, and the metanephros, the most caudal part of the nephrogenic cord. Development begins early in the fourth week of gestation and during the sixth and eighth weeks the lobulated embryonic kidneys ascend from the pelvic region upwards along the posterior abdominal wall to their normal position and undergo a 90° axial rotation from horizontal to medial. At the same time the ureteric bud divides sequentially to form the pelvicalyceal system. An ectopic kidney results from incomplete, excess or abnormal ascent. If during the process of ascent the kidneys come into contact, a horseshoe kidney or crossed renal ectopia will result. An ectopic kidney results from incomplete, excess or abnormal ascent. If during the process of ascent the kidneys come into contact, a horseshoe kidney or crossed renal ectopia will result.

Fusion anomalies of the kidney were initially described by Wilmer in 1938. However, in 1957, McDonald and McClellan modified this classification. In this modified classification, the kidneys are considered ectopic, and they can be a ‘simple ectopic’ or ‘crossed ectopic’ kidney. The ‘crossed ectopic kidney’ can be with or without fusion and may comprise a single or both kidneys. The ‘crossed fused ectopic kidney’ refers to the fused kidney if it is located on the same side, usually at the midline. The crossed fused ectopic kidney may have a pancake type of morphology. The pancake kidney has been reported to be an extremely rare variety of the ectopic fused kidney. The pancake kidney demonstrates complete fusion, including fusion at the upper pole, lower pole and hilum. The isthmus is not seen since there is complete fusion, looking like overlapping of the kidneys. Both kidneys fuse together to form a single mass. In all previous reported cases of pancake

Figure 1. Right lobulated mass extending from L2 to L4.

Figure 2. Showing two renal arteries originating from abdominal aorta with accessory left renal artery originated from left common iliac, along with two renal veins drain into IVC.
kidney, they were found in the pelvic cavity, in the greater or lesser pelvis. So the previous definition of this anomaly is fused pelvic kidney. But in our case the kidney was found in the abdominal cavity.

The arterial supply of the pancake kidney is almost always constant since it lies in the pelvis. The arteries may branch from the iliac arteries or the distal part of the aorta. The veins drain into the iliac veins or the inferior vena cava. Arterial supply of this case is different from usual cases come from its unusual location. However, in the case we present the patient has separate right and left renal arteries, an accessory left artery originated from abdominal aorta and two renal veins draining into the inferior vena cava. As suggested, the developmental explanation of pancake kidney is early fusion of both kidneys to form a single lobulated mass with two separate ureters that usually fail to ascend. However, it seems that it may ascend as reported in our case.

Conclusion

Pancake kidney is a rare developmental problem; this malformation is not necessarily associated with renal dysfunction but does require exclusion of concomitant anomalies. Long-term follow-up is needed for early detection of possible future complications such as PUJ obstruction, stones, infection and increased risk of malignancy.

Conflict of interest

This case report was done in the division of urology for academic purposes and was not funded by any external fund, the submission has no commercial interests, and the authors of this case report are not linked to any external agencies.

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