



# A case of pancake kidney with a single ureter in the retroperitoneal space

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## Abstract

The pancake kidney (PK) is a rare type of renal anomaly in which both kidneys completely fuse without an isthmus. In the previous reports, PKs have double ureters and are located in the pelvic cavity. We encountered a rare case of PK with a single ureter, which is located in the left retroperitoneal space, in a 95-year-old female cadaver, which was detected during a dissection course. In our case, the major calyces joined to form a single renal pelvis, which continued as a single ureter. To the best of our knowledge, this is the first report on PK with a single ureter that is located not in the pelvic cavity but in the retroperitoneal space. The knowledge of such anomalous presentation is important to avoid any complications during retroperitoneal surgery.

**Keywords** Fused kidney · Pancake kidney · Single ureter

## Introduction

Various forms of fused kidney have been reported (Natsis et al. 2014), with horseshoe kidney being its most common variant, with an incidence of 1:400–1:600 in a population (Natsis et al. 2014). Crossed fused renal ectopia is the second most common form of renal fusion anomaly, with an estimated incidence of 1:300–1:7500 in a population (Rinat et al. 2001). Pancake kidney (PK) is characterized by a complete fusion of renal parenchyma without an isthmus and accounts for only 2% of all fused kidney cases (Türkvatan et al. 2009). To the best of our knowledge, only 44 cases of PK have been reported in the literature to date. In general, PK is located in the pelvic cavity and drained by double ureter (Rinat et al. 2001; Türkvatan et al. 2009; Natsis et al. 2014). In this study, we report an extremely rare PK with a single ureter, which is located in the left retroperitoneal space.

## Case report

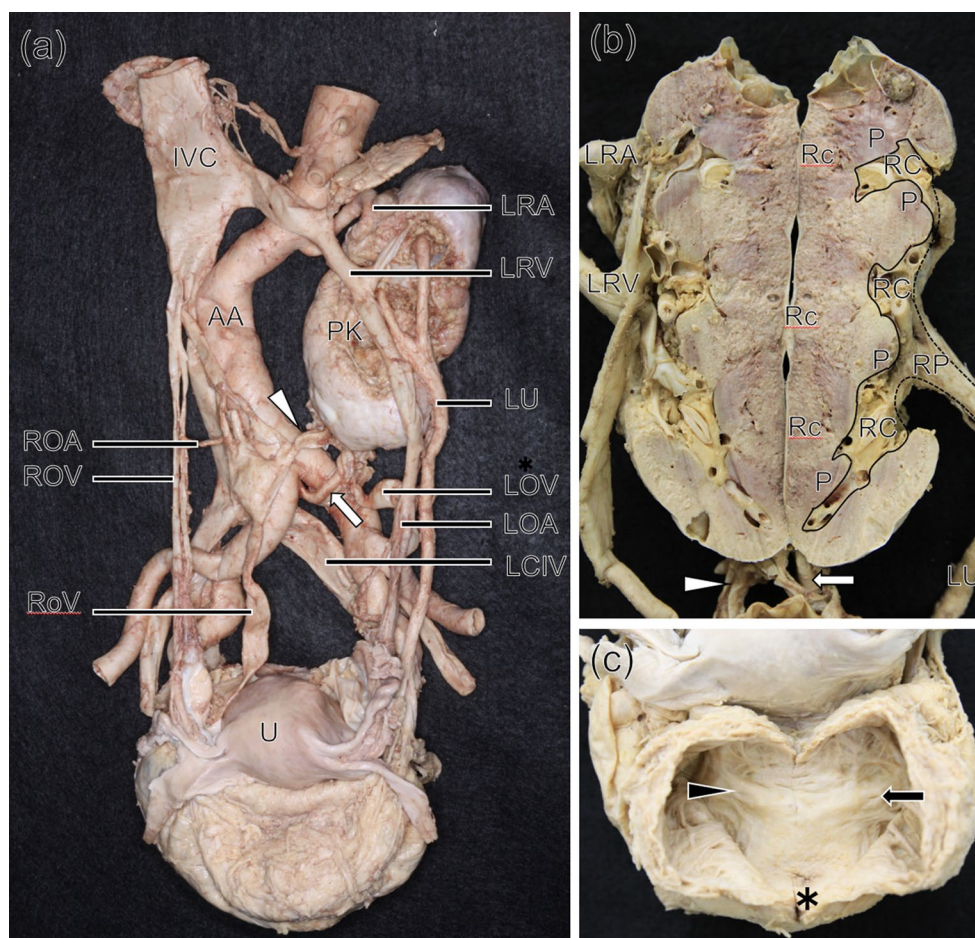
During a dissection course at Aichi Medical University, a rare case of PK was found in 90-year-old Japanese female who had died because of pneumonia. The PK was located in the left retroperitoneal space, at the level of Th12 to L3. The hilus was widely open to face ventrally. At the hilus, one left renal artery and three left renal veins were identified (Fig. 1a). The major calyces joined to form a single renal pelvis, which continued as a single ureter (Fig. 1b). The ureter opened at the left ureteral orifice of the cranial left side of the bladder triangle. The caudal side of the bladder triangle was located at the urethral pelvic region. There was no defect on the right side of the bladder triangle (Fig. 1c).

In the inferior pole of the PK, three accessory renal vessels were identified, namely one artery and two veins. The accessory renal artery originated from the right common iliac artery 5 mm caudal to the aortic bifurcation. One accessory renal vein ran across the anterior aspect of the aorta to form the venous network with the right obturator vein and then opened into the right side of the vena cava distal to the opening of the right ovarian vein. In the superior pole of the PK, two accessory arteries branching from the left artery were identified. There were no remnant renal arteries and veins on the right side (Fig. 1a).

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**Fig. 1a–c** Gross appearance of pancake kidney in a 90-year-old female cadaver. **a** Photograph showing ventral view of the dissected urinary and reproductive systems. **b** Sagittal section of pancake kidney (PK). **c** Enlarged view of the bladder triangle. It was found that the renal pelvis was flat and formed a single renal pelvis including blood vessels from the inferior pole in **b**. There is no defect on the right side of the bladder triangle in **c**. *White arrow* Origin of the renal artery from the right common iliac artery, *white arrowheads* renal veins draining into the right side of the inferior vena cava, *black arrow* left ureteral orifice, *black arrow-head* scar of the right ureteral orifice, *asterisk* urethral orifice. AA Abdominal aorta, B bladder, IVC Inferior vena cava, LCIV left common iliac vein, LOA left ovarian artery, LOV left ovarian vein, LRA left renal artery, LRV left renal vein, LU left ureter, P renal pyramid, RC renal calyx, Rc renal cortex, ROA right ovarian artery, ROV right ovarian vein, RoV right obturator vein, RP renal pelvis, U uterus



## Discussion

We report a rare case of PK with a single ureter located in the left retroperitoneal space, found in an elderly Japanese female cadaver. Several cases of congenital anomalies of the kidney have been reported. Horseshoe kidney is formed by the medial fusion of the kidneys on either side of the midline, whereas the crossed renal ectopia is characterized by kidney and ureter crossing the midline to lie on the opposite side of the midline. PK was first reported by Glenn (1958) as an anomaly in which “the entire renal substance is fused into one mass, completely fused without an isthmus, and giving rise to ureters which enter the bladder in normal relationship”. Most previous reported PK are located in the pelvic cavity and drained by double ureters. Recently, Hamzeh et al. (2017) first reported, by computed tomography (CT) scan, a case of PK found in the retroperitoneal space, which had double ureters (Hamzeh et al. 2017). On the other hand, Goren and Eidelman (1987) first reported a case of PK with a single right ureter, which was detected by excretory urography. Silva et al. (2016) and Kanchan et al. (2017) reported a case of PK with a single

ureter, which was located in the pelvic cavity during CT scan or medicolegal autopsy, respectively. PK in our case was located not in the pelvic cavity but in the retroperitoneal space and had single ureter. In general, congenital solitary kidneys have a single ureter and are also located in the retroperitoneal space (Hiraoka et al. 2002). Therefore, PK in our case mimicked congenital solitary kidney. The characteristic findings of congenital solitary kidney are as follows: the lack of the ureteral orifice on the side of the renal defect, and the lack of half of the triangle of the bladder or abnormal morphology. Considering the high incidence of congenital solitary kidney, 1:1000 in a population (Shapiro et al. 2003), the previous cases of congenital solitary kidney may contain PKs similar to our case.

The ureteral bud finally becoming the kidney occurs from the caudal side of the mesonephric duct. Normal renal development depends on a normal ureteral bud, which undergoes orderly branching and penetrates the metanephric blastema. The ureteral bud is taken into the bladder wall along with the exstrophy of the mesonephric duct (Shapiro et al. 2003; Park 2016). During development, the kidney reaches its definitive position in the lumbar region following a complex sequence of movements that include ascent, lateral migration, and

internal rotation (Natsis et al. 2014; Park 2016). In our case, it was found that the bladder triangle showed normal form, and the right ureteral orifice remained as a scar. This finding indicates that the mesonephric duct and ureteral bud on both sides developed normally and then the right ureter disappeared. On the other hand, the hilus in our case was open to face ventrally and the accessory renal artery originated from the right common iliac artery. From these findings, it appears that renal fusion occurs within the pelvis, and that the right ureter disappears at an early stage of development. This disappearance may cause the ascent of the fused kidney to the left retroperitoneal space without rotation.

In most cases, PK is asymptomatic but can be accompanied by nephrolithiasis, hydronephrosis, and vesicoureteral reflux, resulting in recurrent urinary infections, all of which are attributable to the anomalous rotation of the collecting system and the short ureters, which are prone to stasis and obstruction (Tiwari et al. 2014). In this case, PK was located in the left retroperitoneal space, and the unilateral ureteral system was anatomically normal. Therefore, it was assumed that this woman was asymptomatic during her lifetime.

PK with a single ureter located in the retroperitoneal space is extremely rare. The present case report provides additional details regarding PK and its anatomical variants. Urologists and radiologists should have adequate knowledge regarding this anomalous form of PK. Clinical anatomists and embryologists may find this report interesting.

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### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

**Ethical approval and consent to participate** In this study, we examined a cadaver donated for medical research according to Japanese law (Act No. 56 of 1983). We require the consent for donation from the donor and their family during their life. Following the guidelines of the Japanese Association of Anatomists (item 4. 5), a rare case encountered during a routine dissection class does not need to obtain a statement on ethics approval.

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