

RESEARCH ARTICLE - Human anatomy case report

A rare instance of fused pelvic kidney without other associated congenital anomalies

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Abstract

Much of the fused pelvic kidney (pancake kidney) described in the literature are complicated by coexisting anomalies of other organ system and vasculature. During a routine anatomical dissection of a 69 year-old male we found a fused pelvic kidney without vascular or other organ system defects. A detailed dissection of the entire body showed normal anatomical features except pancreatic cancer and related lymph nodes. Further dissection of the pelvic kidney showed presence of right and left accessory renal arteries; however the remaining vasculature was normal. Possible embryological causes of pelvic kidney and clinical consequences of this anomaly are discussed.

Key words

Pelvic/pancake kidney, congenital defect, embryology.

Introduction

While the first reported case of a completely fused pelvic kidney in the modern literature dates from Huntington's Harveian Society Lecture of 1907 (Huntington, 1908), it is generally recognized that the first detailed report was that of Looney and Dodd (1926). The occurrence of an abnormal internal urinary system (IUS) is reported to vary from 10% (autopsy study) (Rubenstein et al, 1961) to 3.51 per 1000 births (Stoll et al, 1990). Urinary system abnormalities often coexist with anomalies of the cardiovascular, gastrointestinal, anorectal, genitourinary and other systems (Boatman et al. 1972; Cook and Stephens, 1977; Van Allen, 1993; Bauer, 1998). Among all the IUS abnormalities reported horseshoe kidney is the most prevalent (Kaufman and Findlanter, 2001). As noted earlier (Kaufman and Findlanter, 2001), the rarest IUS involves the complete fusion of the 2 kidneys to form a single entity commonly known as a 'pancake' 'pelvic' or 'discoid' kidneys. Taken together, the occurrence of solitary pelvic kidney - as the case reported here - is 1:22,000 (Bergman et al, accessed 2016). What makes the present case exceptional is the fact that no other organ system or part of vasculature were affected.

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Case report

During routine cadaveric dissection we observed the absence of kidneys in the lumbar region and upon further dissection a fused pelvic kidney was identified (Figs 1 and 2). The donor was a 69-year-old Caucasian male died of complications resulting from pancreatic cancer. We made a detailed documentation of our observations of the cadaver. All the other organ systems (except the cancerous pancreas) and the vasculatures of the head and neck, thorax and abdomen were unremarkable. The suprarenal glands were in the normal locations around the 12th thoracic and 1st lumbar vertebra. The abdominal aorta divided much lower than usual within the pelvis. The renal arteries from the aorta arose at the level of sacral promontory close to the aortic bifurcation (Figure 1). We also observed a right and a left accessory renal artery but it was hard to dissect their entrees into the renal mass. The right ureter arose from the hilum while the left ureter arose anteriorly slightly at a higher level, they entered the urinary bladder separately. The combined size of the fused kidneys was normal except

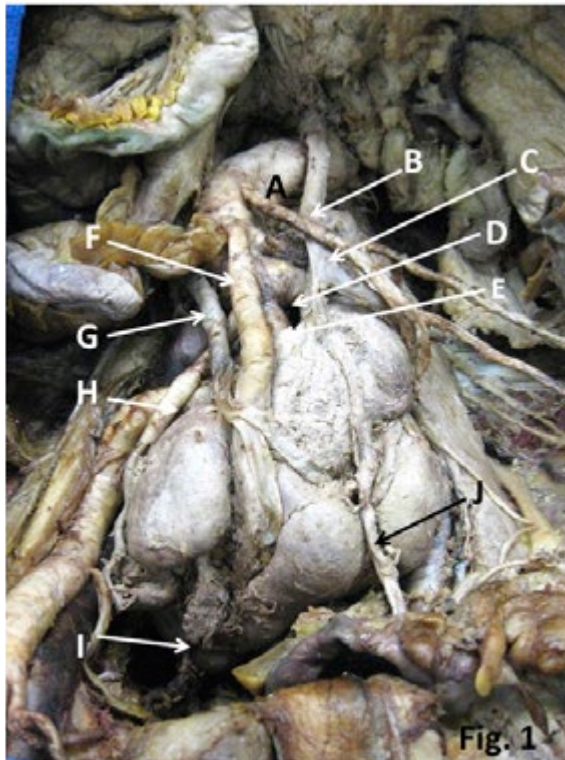


Figure 1 – Photograph of a detailed dissection of the fused pelvic kidney demonstrating the vasculature and ureters. A. Aorta, B. Left renal vein, C. Inferior mesenteric artery, D. Left renal artery, E. Left accessory renal artery, F. Right renal artery, G. Right renal vein, H. Right accessory renal artery, I. Right ureter, J. Left ureter. Note the lobulated appearance of the renal mass.

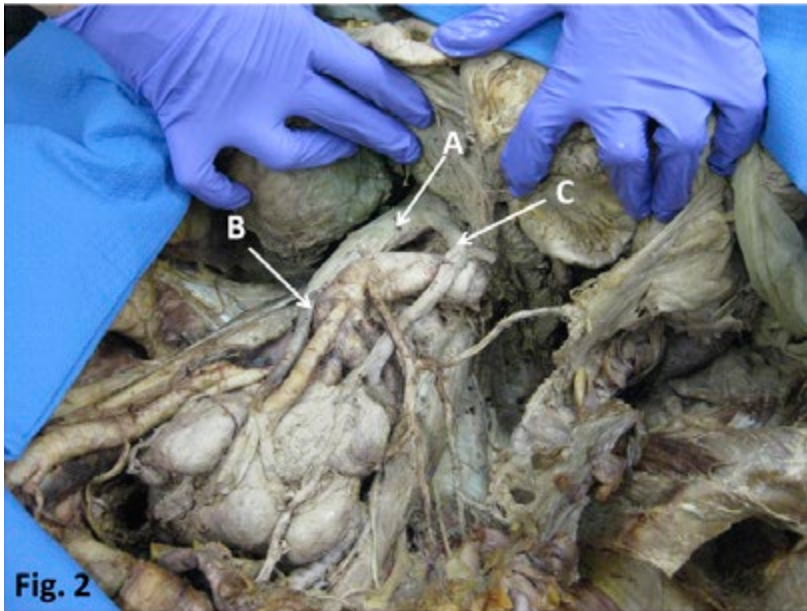


Figure 2 – Photograph of a dissection of the fused pelvic kidney demonstrating the venous system. A. Inferior vena cava, B. Right renal vein, C. Left renal vein with part of testicular vein.

it was hard and challenging to dissect. We also noticed the lobulated external appearance of the fused kidneys presumably due to embedded vessels.

Discussion

Much of earlier findings of fused pelvic kidney came from autopsy, cadaveric dissection or incidental findings during surgical or radiological procedures. In a number of these cases there were associated anomalies involving cloacal derivatives and lower limbs in what has been termed “caudal regression syndrome” (Duhamel, 1961; Braren and Jones, 1978; Brock et al. 1983). Generally patients with ‘fused kidney’ remain asymptomatic, although they often present with anomaly of the reproductive system (Rosenkrantz et al, 2010; Schwartz et al, 2010) and other organ systems (Boatman et al. 1972; Cook and Stephens, 1977; Van Allen, 1993; Bauer, 1998). In all reported cases of fused kidneys, the arterial supply and venous drainage were grossly abnormal, and this reflects the embryonic arrangement invariably seen in ectopic kidneys, often because of their very limited rostral migration during development (Kaufman and Findlater, 2001). In general, renal fusion anomalies are more common in males with a male to female ratio of about 2-3:1 (Kaufman and Findlater, 2001). In the case presented here the donor was a male and no other organ system or part of vasculature were affected. The fact that the donor lived a full life span of 69 years shows that this congenital anomaly, as in many cases, did not affect his longevity.

Patients with a fused (pancake) kidney are mostly asymptomatic, but may initially present with recurring urinary tract infection, fever and abdominal discomfort. The presence of a pancake kidney may predispose the patient to recurrent urinary tract infections and the formation of calculi due to the probable rotation anomaly of the collecting system and short ureters which are prone to stasis and obstruction (Eckes and Lawrence, 1997; Babu et al, 2015). Depending upon the origin of the ureter and its termination into the urinary bladder the patients may have calculi and obstruction, and in an extreme situation hydronephrosis resulting in severe abdominal pain. Sometimes patients may present with extra renal symptoms, such as amenorrhea, failure to conceive or iliac vessel aneurysm (Eckes and Lawrence, 1997; Tiwari et al, 2014). The presence of pancake kidney rarely leads to progressive renal failure. Since the cadaver was from an unidentified donor we have no knowledge of his past medical history except the cause of death.

A majority of the cases of fused pelvic kidney are associated with anomalies in other organs such as abnormal testicular descent, Tetralogy of Fallot, vaginal absence, sacral agenesis, caudal regression syndrome, spina bifida and anal abnormalities (Brock et al, 1983). The diagnosis of pancake kidney is not necessarily associated with a poor prognosis. However, complications that can be associated with anatomic malformations, such as urinary stasis, infection, formation of stones and vascular involvement, can cause serious clinical problems (Goren and Eidelman, 1987). Therefore, cases of cake kidney must be investigated in order to exclude concomitant anomalies and to prevent complications (Goren and Eidelman, 1987). It may pose problems to surgeons during abdominal surgery.

The development of the kidney begins in the 4th week of gestation by inductive interaction between the ureteric bud and the metanephric blastema. Initially the developing kidney lies in the sacral region and receives blood supply from the iliac vessels. Due to differential growth of the caudal end of the fetus the kidneys progressively ascends cranially from the pelvic region to their final position and at the same time undergo 90° axial rotation from horizontal to medial. During the ascent the kidneys receives blood supply from the lower end of the abdominal aorta and finally around week 10 the kidneys receive blood supply from the aorta through the renal artery. Congenital renal anomalies including renal ectopia can occur due to abnormalities of development, migration and rotation (Moore et al, 2016).

The accurate mechanism of renal fusion (pancake or pelvic kidney) anomalies is not clearly defined and understood even though many theories have been put forward. An abnormally located umbilical artery may force the metanephric masses to squeeze together and cause fusion (Srivastava et al, 1971; Kelalis et al, 1973; Goren and Eidelman, 1987). After fusion occurs, cranial ascent to the lumbar position is impaired by the inferior mesenteric artery. The vascular supply of the pancake kidney is consistent with its arrested ascent and derives from the common iliac artery or terminal aorta. Alternatively, this anomaly could result from the growth of the ureteric buds into a common metanephric blastema (Cook and Stephens, 1977; Kaufman and Findlater, 2001).

In conclusion we have presented a case of fused pelvic kidney in which, uniquely, other organ systems were not affected, whereas it is well known that pancake kidney usually coexists with other congenital anomalies.

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