Congenital solitary functioning kidney; an incidental cadaveric dissection finding in an adult male Nigerian: Case report.

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Abstract
Background: Congenital solitary functioning kidney is one of the congenital anomalies of the kidney and the urinary tract. It is very rare and has a male predominance. An incidental case of a congenital solitary functioning kidney in an apparently healthy adult Nigerian male was discovered during cadaveric dissection for a study of kidney dimensions in Nigerians.
Conclusion: This report was premised on the rarity of this congenital anomaly, its existence as the only anomaly in an adult and its compatibility with normal adult life.This case report further strengthened the crucial role and benefit of cadaveric dissection in the teaching and learning of human anatomy.

Keywords: *Congenital solitary kidney, adult male, Nigerian, Cadaveric dissection*

Résumé
Contexte: le fonctionnement solitaire congénital du rein est l'une des anomalies congénitales du rein et des voies urinaires. C'est très rare et a une prédominance masculine. Un cas incident d'un rein de fonctionnement solitaire congénital chez un homme nigérian adulte apparemment en bonne santé a été découvert lors d'une dissection cadavérique pour une étude des dimensions rénales chez les Nigérians.
Conclusion: Ce rapport était fondé sur la rareté de cette anomalie congénitale, son existence étant la seule anomalie chez un adulte et sa compatibilité avec la vie adulte normale. Ce rapport de cas a renforcé le rôle crucial et le bénéfice de la dissection cadavérique dans l'enseignement et l'apprentissage d'anatomie humaine.

Mots-clés: *Rein congénital solitaire, Homme adulte, Nigérian, dissection cadavérique*

Introduction

Congenital solitary functioning kidney (CSFK) is one of the anomalies of the genitourinary organs and tract. It is characterized by the presence of only one kidney either due to failure of development of one kidney i.e unilateral renal agenesis or regression and complete disappearance of a malformed kidney before birth. In children, diagnosis is usually made at abdominal ultrasonography in the course of investigation for renal and urinary tract pathologies. In adults, its diagnosis may be incidental finding at surgery, postmortem examination, cadaveric dissection or radiologic investigations for either renal or non-renal indications. The exact incidence of CSFK is unknown, however, a postnatal autopsy series put it at 1: 1000 [1], an ultrasonographic study reported an incidence rate of 1:500 [2] while another ultrasonographic study put it at 1 per 1500-3200 live births [3]. In an ultrasound screening study of 4000 neonates, the incidence of CSFK was found to be 1: 1300 [4]. The incidence of CSFK in Nigerians is unknown. It may be an isolated anomaly as in the index case. It may be syndromic; associated syndromes and anomalies with which CSFK had been reported included acro-renal syndrome [5-8]; VACTREL anomalies [9]; MURCS syndrome [10], Poland syndrome [11] and Ear, Nose and Throat anomalies (ENT) [12]. It has been reported to be commoner in males than females [3,12].

A review of literature revealed that only one case, a 38 year old Nigerian male with unilateral renal agenesis coexisting with bilateral cryptorchidism had been reported [13]. Thus to this best of our knowledge, this is the second case of CSFK in an adult Nigerian that will be reported. It is worthy of note that this case being reported was an incidental finding in the course of cadaveric dissection for a study on assessment of kidney dimensions in Nigerians by us.

Case report

The index case was one of the cadavers sourced from Lagos, Nigeria for the purpose of supervised gross anatomy dissection by undergraduate medical students. Unconfirmed source had it that he was involved in deadly clash between rival cult groups at which dangerous weapons were freely deployed.

The body was that of a young negroloid, healthy looking man of medium to heavy habitus.

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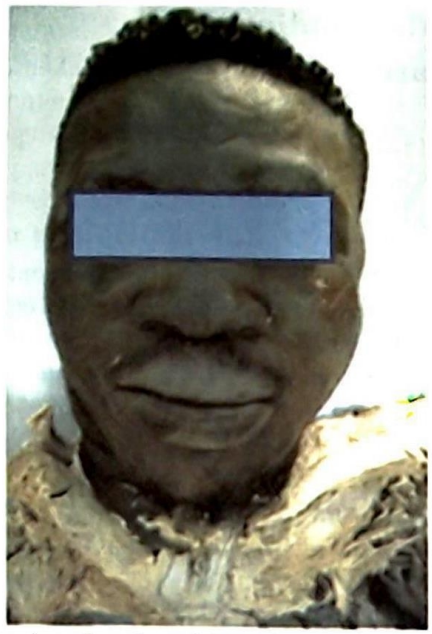

Fig. 1a: Anterior view of the Index Case.Note the facial appearance

Fig. 1b: Posterior view: The sutured deep lacerations are indicated by **** While §§§ indicate an extensive ragged-edge gaped laceration.

He had a stylish hair cut with dark full grown and groomed beard (Figure 1a). Physical examination of the body did not reveal any scar that would have been suggestive of previous abdominal surgery however, about forty eight deep matchet cuts in all the regions of the body were noted. They were mostly sited in the head, neck, back (posterior trunk) and the upper limbs. Prominent amongst the lacerations were a ragged- edge one approximately 10.0 cm in length and 3.0 cm deep at the level of posterior hair line and another one over the left parotid region with a length of about 6.0 cm. (Fig. 1b).

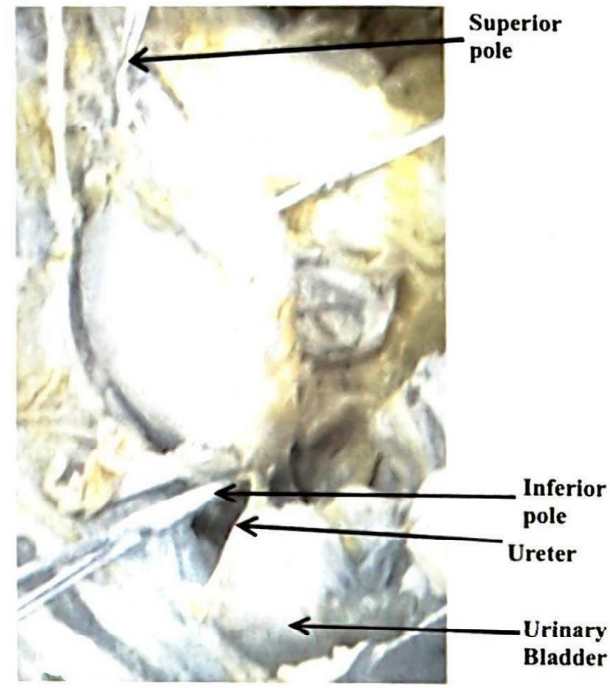

Fig. 2: Anterior view of the Solitary Kidney (In-situ) The superior pole is conical while the inferior pole

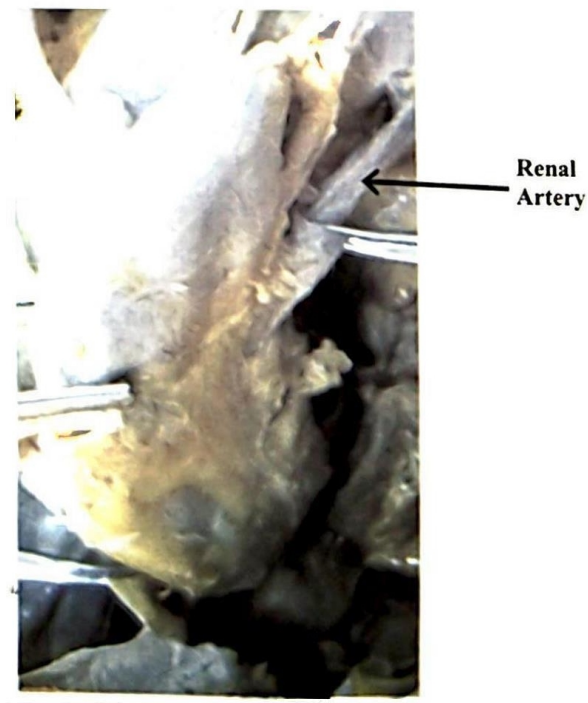

Fig. 3: Side view showing the single appears roundish. Renal artery

During the dissection of the abdominopelvic cavity, both lumbar (paravertebral) regions were explored for the respective kidneys but none was

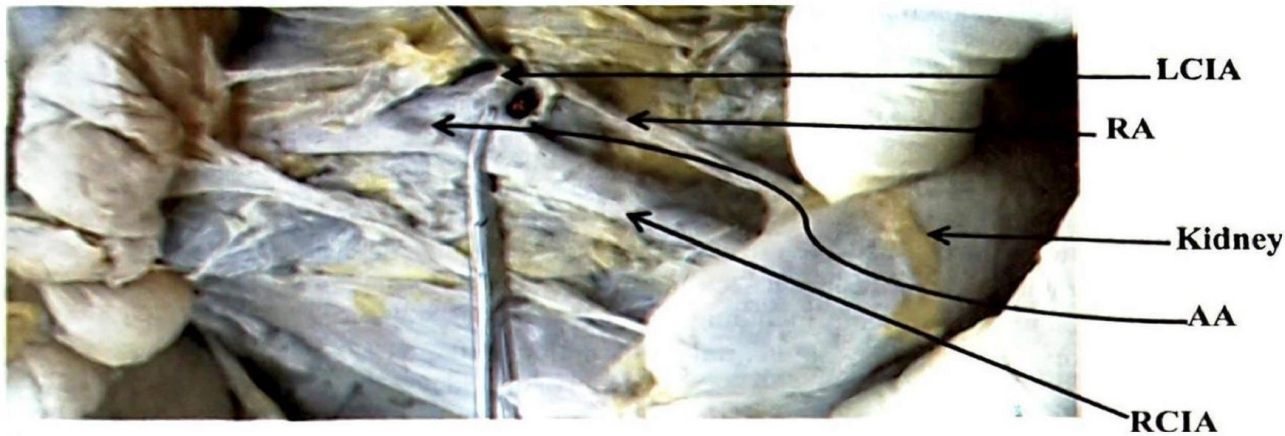

Fig. 4: The renal artery and its origin. *The small rent in the wall of the of the LCIA just at its origin was iatrogenic.LCIA=Left common iliac artery. RA = Renal artery. AA = Abdominal aorta. RCIA=Right common iliac artery.

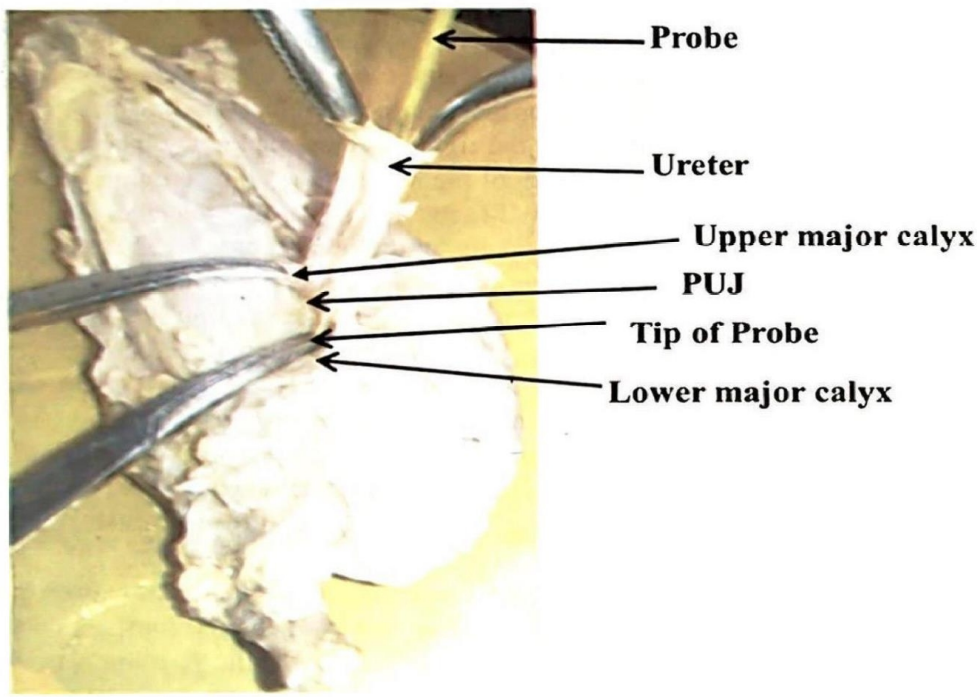

Fig. 5: Posterior view of the solitary kidney. Showing the ureter, the pelviureteric junction (PUJ) and two major calices

found. It is pertinent to note that there was no evidence of previous exploration of the lumbar regions i.e both lumbar regions were 'virgin'. A solitary kidney with the following description was however found in the lower midline. The kidney was ovoid in shape extending from the upper border of the 4th lumbar vertebra down to the sacral promontory (figure 2). It had a bipolar dimension (length) of 10.4 cm, transverse dimension (width) of 5.1 cm, a thickness of 3.0 cm and weighed 96.0 g. There were two veins on the anterior surface and one on the posterior surface. The two anterior veins were 3.0cm and 3.5 cm distal from the upper pole respectively. They merged to form the proper anterior renal vein. The posterior vein arose from its posterior aspect approximately 5.0 cm from the upper pole. Both the anterior and posterior renal veins unite at the upper pole to form the main renal vein which emptied into the inferior vena cava at about 3 cm proximal to the formation of the inferior vena cava by the merging of the right and left common iliac veins. The main renal vein was 3.2 cm long, the proper anterior vein was 2.9 cm in length and the posterior renal vein was 3.9 cm long.The kidney had a single artery which

was a branch of the left common iliac artery and it pierced the parenchyma of the kidney on its posterior surface about 1.0 cm distal to the origin of the posterior renal vein (Figures 3&4). The ureter arose from the posterior surface and was about 6.0 cm long (figure 5). The kidney was in alignment with the urinary bladder along the vertical axis.All the other abdominopelvic structures and organs were grossly normal and intact. Examination of the head and neck, thoracic cavity, limbs and the perineum including the gonads did not reveal any gross congenital anomaly.

Discussion

The kidneys start to develop at the beginning of fourth week of gestation and they become functional by the beginning of the 9th week, nephrogenesis however continues till the 36th week.

The precursors of the paired human kidneys are the pronephroi, mesonephroi and metanephroi. The pronephroi found in the cervical region, degenerate after giving rise to the mesonephric ducts. The mesonephroi which are located in the thoraco-lumbar region give rise to the initial glomeruli and tubules (both constitute the mesonephric kidneys).These tubules open into the mesonephric ducts. The mesonephric kidneys will subsequently degenerate. The metanephroi located in the sacral region become the permanent kidneys. Each metanephros consists of metanephric diverticulum (MD) (an outgrowth of the mesonephric duct) and metanephrogenic blastema (MB) which is a mass of intermediate mesoderm. The MD forms the ureter, renal pelvis, calices and collecting tubules. .

The glomerulus and its capsule, proximal convoluted tubule, loop of Henle and distal convoluted tubule are derived from the metanephrogenic blastema. Both MD and MB interact and induce each other this is known as reciprocal induction. It is this process that gives rise to the permanent kidney [14]. Certain molecules known as renal developmental genes (RDG) regulate this reciprocal induction. These RDG include glial derived neurotropic factor (GDNF), fibroblast growth factor-2 (FGF-2), bone morphogenetic protein 7 (BMP7), hepatocyte nuclear factor-1β (HNF1β), Pax2, BMP4, FRAS1, FREM2, Six2 and Ret which is a receptor tyrosine kinase for GDNF [15-18]. Thus mutations of the RDG or defects in the transcription factors may result in congenital anomalies of the kidney and urinary tract.

The metanephric kidneys lie closely to each other ventral to the sacrum, as the embryo grows; they progressively ascend and become further apart. The normal adult position of 11th thoracic vertebra to the 3rd lumbar vertebra (T11- L 3) is reached by the 9th week. The hilium is initially anterior but with subsequent 90° medial rotation it becomes anteromedial. The origin of the renal artery changes with the accession of the kidney [14].

Congenital anomalies of the kidney may be in form of non-formation of one or both kidneys (renal agenesis), abnormal location (ectopic kidney), disruption of rotation (malrotated kidney) and polar fusion of the kidneys (horseshoe kidney). With serial ultrasound scanning, cases of renal aplasia have been observed to regress to renal agenesis [4].

In the case being reported, only one kidney was found, thus it is a case of unilateral agenesis. This most probably arose from failure of reciprocal induction between the mesonephric diverticulum and metanephrogenic blastema. The origin of the renal artery of the index case was the left common iliac artery thus the reciprocal induction failure must have involved the right metanephric kidney. The normal outgrowth of the MD requires transcription factors such as WT1gene and signaling molecules such as GDNF and its epithelial receptor ret. Thus non - expression or under expression of these molecules will hinder the reciprocal induction and result in agenesis. The expected location of the kindney was the left lumbar region however, the index case was located anterior to the lower lumbar vertebrae (4th and 5th) and the sacral promontory (midline) ; this is a form of ectopia. This ectopia could have resulted from ectopic ureteric budding, failure of vascularization or abnormal migration [19]. The single artery that supplied this index arose from the left common iliac artery. Thus the most probable factor responsible for its ectopic position was the arrest of its accession. Though, no anatomical structure that could have been responsible for the arrest of the accession was observed; this must have been atrophic and subsequently disappeared since the index case was an adult.

The exact causes of renal agenesis are unknown but certain genetic and environmental factors either maternal or inherent in the developing embryo may be responsible. Such implicated factors include foetal alcohol syndrome, maternal diabetics [20] , maternal usage of thalidomide [21], chromosomal abnormalities such as trisomies 10, 21 and 22; 45 X mosaicism and 22q11 microdeletion [22]. Certain drugs if administered during pregnancy may interfere with renal development. Such drugs include angiotensin converting enzyme inhibitors, dexamethasone, anticonvulsants and amino

glycosides [23]. Individuals with CFSK are at increased risk of renal injury. This injury includes hypertension, proteinuria and progression to chronic kidney disease [22]. This renal injury is precipitated by glomerular hyperfiltration which results into glomerular hypertension. No medical history was available in the index case; hence the presence or absence of renal injury in him could not be ascertained.

Urinary tract infections (UTI) and vesicoureteral reflux have been documented to retard kidney growth in children with CSFK [24], thus it is pertinent to treat UTI promptly and adequately in such patients.

At least one kidney is required for sustenance of life thus in patients with solitary functioning kidney (SFK) with renal tumours total nephrectomy is contraindicated. Nephron sparing surgery (NSS) is the gold standard for patients with SFK [25]. Operative modalities for NSS include open partial nephrectomy (OPN), laparoscopic partial nephrectomy (LPN) and robotic partial nephrectomy (RPN). Both LPN and RPN procedures have better perioperative outcomes [25] however, they require more expertise and equipment than OPN. These additional requirements (expertise and equipment) place a greater professional burden on the Urological Surgeon. Other procedures for SFK with tumours are cryotherapy and radiofrequency ablations [26, 27] both however have inferior outcomes to the previously mentioned procedures [28].

The definitive management for patients with chronic renal failure is replacement of only one kidney at one transplant surgery. The case being reported further strengthened this age-long tested transplant surgical procedure of replacing only one kidney and not the two kidneys in patients with end stage kidney disease. Also, this case report further attested to the previously held view that cadaveric dissection is crucial to the teaching and learning of gross anatomy and should not be replaced rather, available alternatives should be complementary [29].

Conclusion

From circumstantial evidences, it might be inferred that the index case being reported was apparently healthy. If his renal status was compromised, he would not have had the strength to live a violent life style as evidenced by the findings on physical examination and the multiplicity and the depth of the lacerations. Thus this case highlights the rarity of congenital solitary functioning kidney in adult Nigerians.

This case also attests to the fact that congenital solitary functioning kidney may be the only anomaly in an individual and such person may live a normal life with death being unrelated to the kidney.

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