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Case Report

Anomalies by Birth in Urogenital System: Clinical Aspect

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Abstract – During routine dissection of 60 years old male cadaver in Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, it was found that the left kidney showed multiple cysts with dilated ureter and right kidney showed an extra renal artery entering the kidney from the upper pole. Both these findings are present in an individual since birth and are most leading causes of renal failure, hypertension, hydronephrosis and even jeopardisation of renal transplants.

Keywords – Multiple cysts, extra renal artery, pole, renal failure, hypertension, hydronephrosis, transplant.

1. Introduction

Polycystic kidney disease, a disorder that can be diagnosed in adult and pediatric patients, is an inherited disease that involves renal cysts without dysplasia. The condition is broadly divided into 2 forms: autosomal recessive polycystic kidney disease, previously known as infantile polycystic kidney disease, and autosomal dominant polycystic kidney disease, previously known as adult polycystic kidney disease. The knowledge about polycystic kidney disease is necessary since it is one of the leading causes of renal failure in children and adults. The arterial supply to kidneys is through renal arteries. The renal arteries arise from abdominal aorta below the origin of superior mesenteric artery, on each side. Near the hilum of the kidney, each renal artery divides into anterior and posterior branch, which in turn divides into number of segmental arteries supplying the different renal segments. The presence of unusual branching patterns of the renal arteries is not uncommon. In 70% of cases there is a single renal artery supplying each kidney. In various studies, a possibility of association between presence of multiple renal arteries and hypertension has been reported. Aberrant renal arteries to the inferior pole cross anteriorly to the ureter and may cause hydronephrosis. Knowledge of the variations of renal vascular anatomy has importance in exploration and treatment of renal trauma, renal transplantation, renovascular hypertension, renal artery embolization, angioplasty or vascular reconstruction for congenital and acquired lesions, surgery for abdominal aortic aneurysm and conservative or radical renal surgery [1]. During routine dissection, from a single male cadaver, 60 years of age. it was found that the left kidney presented with a polycystic kidney. The kidney on the right side showed an extrarenal artery. All other visceras show no abnormalities.

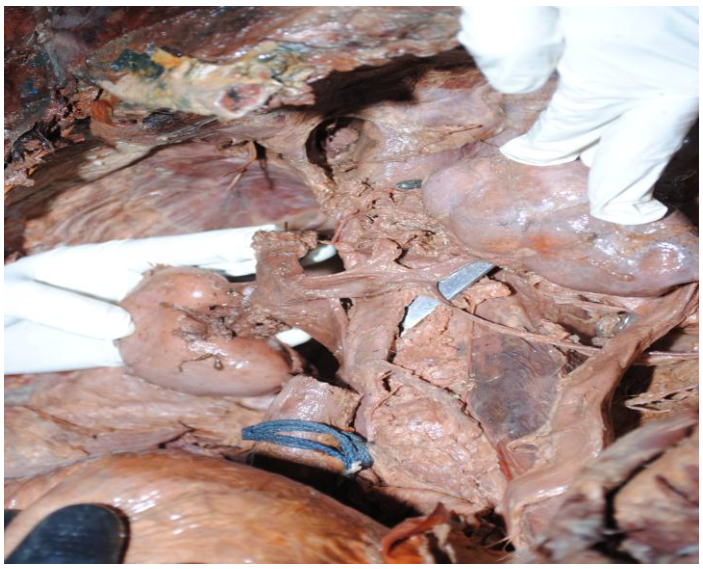
2. Observations

2.1. Left kidney

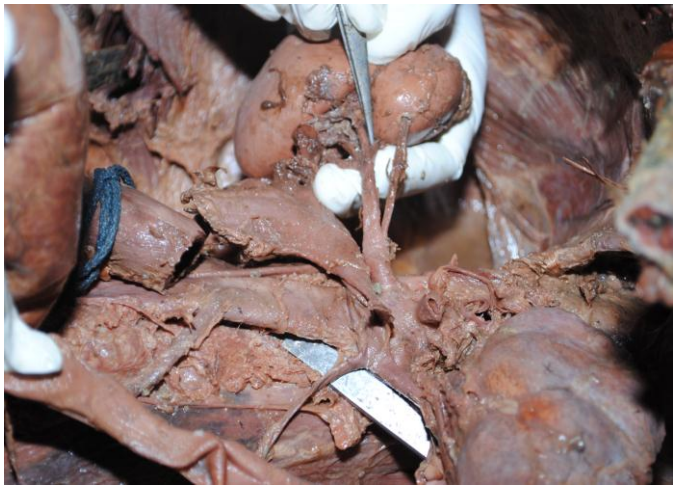
The left kidney showed a size of 10 x 6 x 5.5 cm. The ureter showed a size of 3cm. It is dilated. On gross examination kidneys showed a number of cysts. The left renal artery is also taking origin from abdominal aorta opposite the right renal artery and enters the kidney at the hilum of the left kidney. The length of left renal artery is 3 cm. Picture 3 shows the polycystic kidney and picture 4 shows dilated ureters continuing as dilated pelvis from left kidney.Every section should start with first paragraph and should use the right style.

2.2. Right kidney

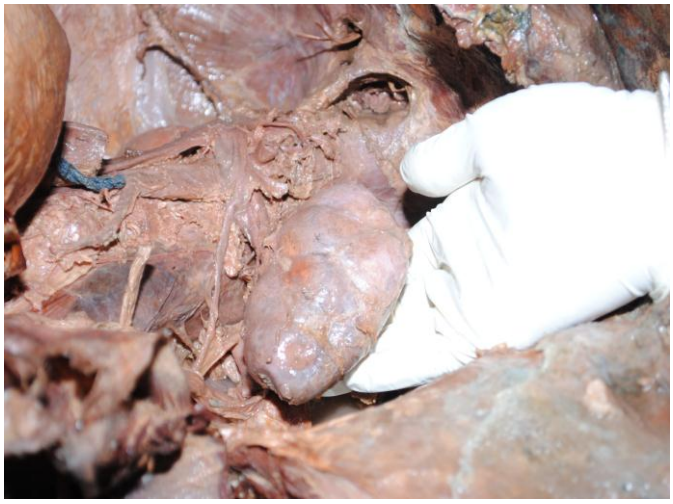
The right kidney showed a size of 9 x 5 x 3 cm. The right renal artery is also seen to originate from the abdominal aorta but after a distance of .1.5cms...it gives off a branch which enters the right kidney from the upper pole of right kidney. The length of right renal artery is 4 cm. The superior polar renal artery is 3.5cms.The right renal artery near the hilum of the right kidney divides into four branches which enter the kidney through the hilum of the kidney. No cysts are seen. Picture 2 shows accessory artery (superior lobar artery) supplying the right kidney.



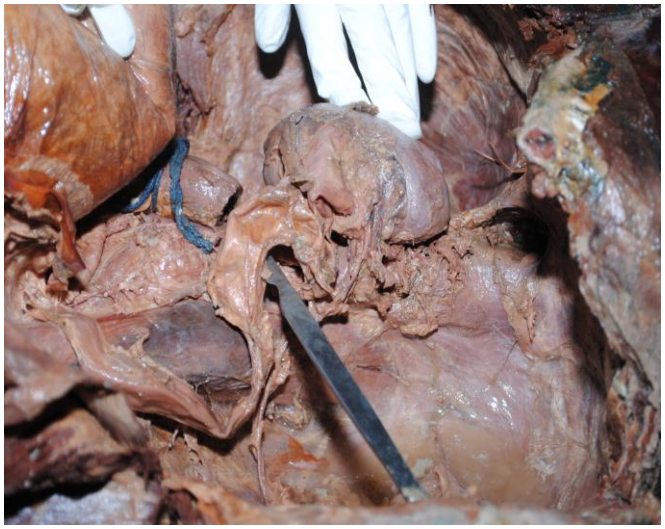
Picture 1- Showing both the kidneys and dilated ureter.



Picture 2- Showing accessory renal artery supplying right kidney



Picture 3 - Showing polycystic kidney left side



Picture 4- Showing dilated ureter left side

3. Discussion

Polycystic kidney disease is one of the life-threatening inherited disorders summated by the development of bilateral or unilateral renal cysts that might lead to renal failure in due course of time. One in 1000 people carry the APKD mutant gene. Adult polycystic kidney disease is usually asymptomatic until the third or fourth decade of life, and, although histological evidence of the disease is likely to be present from intrauterine life, the age of onset of gross morphological changes that is potentially detectable by ultrasonography. In 1974, Bear reported two cases of adult polycystic kidney disease. First case was 48 year man with unilateral ADPKD with agenesis of contralateral kidney. His father died of hypertension and renal failure and autopsy revealed polycystic kidney. The second case was unilateral ADPKD with contralateral nephrectomized kidney [2]. Todorov reported another case of unilateral ADPKD with contralateral renal agenesis [3]. Levine et al stated that unilateral renal cystic disease (URCD) had at least three aspects from ADPKD, that is unilateral localization, negative family history and no progression to chronic renal failure [4]. URCD patients usually have no cysts in other intra-abdominal organs [5]. Renal artery variations are common in the general population and they show variations in the frequency according to the social, ethnic and racial differences. It is more common in Africans (37%) and Caucasians (35%), and is less common in Hindus (17%) and the populations except Caucasians (18%).Accessory renal arteries are common (30%) and usually arise from aorta above or below the origin of the renal arteries[6].Renal artery variations are divided into 2 groups: Early division and Extra renal artery ERA.

Renal arteries as they reach the hilum further divide as segmental, lobar, interlobar, arcuate and interlobular [6]. While renal arteries divide into segmental branches at hilus level, a branching occurring more proximal to hilus is called “Early Branching”. Extrarenal arteries are grouped as follows: hilar(accessory) and polar(aberrant)arteries. While hilar arteries enter through the hilus with main renal artery and polar artery penetrate kidney directly through the capsule from outside the hilus [7]. Accessory renal arteries are commonly derived from the renal, abdominal aorta, common iliac and superior mesenteric arteries [8].Rarely they may originate from the external iliac, lumbar, spermatic, ovarian, inferior mesenteric, superior suprarenal, inferior phrenic, right colic, subcostal, contralateral renal, splenic and the thoracic aorta [9]-[11]. In the present case, an anomalous pattern of renal artery was observed, which might be the result of changes during embryonic development. The anatomic changes might be the result of developmental changes in lateral splachnic arteries [12]. Aberrant or accessory arteries have been of interest to the clinician for some years, mainly because of the possible part that the vessel may play in the causation of hydronephrosis. `Normally the kidney is supplied by a single renal artery which enters the hilum and that any additional artery entering the organ at one or other pole is necessarily an extra and aberrant source of supply to that provided by the main stem artery. Such vessels entering the upper pole of the kidney arise from either the aorta, renal artery or a suprarenal artery, whereas those entering the lower pole may stem from the aorta, common or internal iliac, superior mesenteric or spermatic artery [13, 14]. Renovascular hypertension is a condition of arterial hypertension induced by renal perfusion secondary to a vascular lesion [15]. In various studies, a possibility of association between the multiple renal arteries and hypertension has been reported [16]. According to one study, accessory or extra renal arteries usually have a longer

length and a smaller diameter than the main artery. The renal segment supplied by this artery shows decreased blood pressure which causes hypertension by stimulating rennin secretion but Gupta et al showed no association between accessory or multiplr renal artery and hypertension [17]. Anatomicical knowledge of multiple or accessory renal arteries is important before doing any transplant surgeries where microvascular techniques are required [18].

On embryological basis the permanent kidney develops from two sources, one in the mesoderm of sacral part of nephrogenic cord and other from the ureteric bud, a diverticulum arising from the mesonephric duct. Further, development involves epithelial mesenchymal interaction. Epithelium of ureteric bud interacts with mesenchyme of metanephric blastema. In the autosomal recessive polycystic kidney disease, cysts occur from collecting ducts, due to deficient or non-responsive cell adhesion molecules, syndecan and E-catherin which are essential for condensation of mesenchyme to epithelium [19]. The mesenchyme expresses a transcription factor that makes this tissue competent to respond to the induction. Characterization of the autosomal recessive polycystic kidney disease (ARPKD) gene has been complicated by genomic rearrangements on chromosome 16. Autosomal dominant polycystic kidney disease (ADPKD) is a genetically heterogeneous disease, with mutations in the PKD1 gene located on chromosome 16p13.3 accounting for the majority of cases [20].

Each primitive dorsal aorta gives off ventral splanchnic arteries,lateral splanchnic arteries, somatic arteries and caudal continuation.The lateral splanchnic arteries supply, on each side, the mesonephros,metanephros, the testis or ovary and the suprarenal gland. All these structures develop, in whole or in part, from the intermediate mesenchyme of the mesonephric ridge. One testicular or ovarianartery and three suparenal arteries persist on each side. Additional renal arteries are frequently present and may be looked on as branches of persistent lateral splanchnic arteries [21]. Arey gave the concept that anomalous blood vessels may be due to: a) choice of unusual paths in the primitive vascular plexuses; b) persistence of vessels normally obliterated; c) disappearance of vesselsnormally retained; d) incomplete development; and e) fusion andabsorption of parts usually distinct [22].

4. Conclusion

In conclusion accessory and polar arteries are a frequent occurrence but the presence of unilateral polycystic kidney is not seen so frequently. These anomalies may cause hypertension and even lead to renal insufficiency. The accessory vessels may cause hydronephrosis if it presses upon the pelvis or ureter. These multiple or accessory arteries may lead to failure of renal transplantation. Thus we should have a complete knowledge and awareness of causes of these anomalies of the renal arteries so that sufficient surgical management during renal transplantation, repair of abdominal aorta aneurysm, urological procedures and angiographic interventions can be done.

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