Unilateral Renal Agenesis with variations in the vascular pattern of Testis, Supra Renal Gland and Diaphragm - A Case Report

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ABSTRACT
Unilateral renal agenesis is so common, which occurs three times more in males and the left kidney is frequently involved. Here we report a case with unilateral renal agenesis and associated vascular anomalies. During the routine cadaveric dissection for undergraduates in Anatomy, congenital absence of left kidney and associated blood vessels were found in one of the male cadaver. Left suprarenal gland was present with altered shape and nourished by left inferior phrenic artery. The left inferior phrenic artery commences from the right renal artery as the left division of one of its branch. Embryological and genetic reasons behind the renal agenesis and its relationship with the vascular variations are discussed. A good knowledge of variations of the renal arteries and their branches as in our case helps in the surgical interventions of this region.

KEY WORDS: Renal agenesis, Suprarenal gland, Phrenic artery, Gonadal artery, Aberrant right renal artery, Suprarenal artery.

Introduction
The renal agenesis is one of the most frequent renal anomaly and has an incidence of 1/200 and 1/4000. By birth, renal agenesis is more common in male then in female. It happens more in left, than to right side [1]. The incidence of renal agenesis is increased in fetuses that have first degree relation with congenital renal anomalies. Renal anomalies are associated with some autosomal recessive and dominantly inherited disorder, including Meckel-Gruber syndromes, Townes–Brockes syndromes & Ivemark syndromes [2].

The most frequent genitourinary anomalies are renal, testicular & urethral respectively. About 10% of the populations have some kind of genital or urinary system anomalies. Urogenital anomalies in children and in patient having some syndrome, such as cystic fibrosis, chromosomal anomalies and neo plasma are very frequent [1].

Variations in number, source and course of the renal arteries are common. The renal artery
may give rise to branches normally derived from other vessels, such as the inferior phrenic, hepatic, suprarenal, gonadal, pancreatic and lumbar arteries. Familiarity about the possible variations in the renal artery pattern is especially important for the personnel dealing with kidney retrieval and transplantation, various endourological procedures and innumerable Interventional techniques [3]. In most of those situations, it is the comprehensive knowledge of the renal artery pattern which remains the key issue in determining the technical feasibility of surgical interventions as well as the post-operative management. Awareness of variation of testicular arteries is important during surgical procedures like varicose & undescended testis [4].

Clinically the right inferior phrenic artery was always found to be related with hepatocellular Carcinoma (HCC) and served as a major collateral artery for hepatic artery during surgical ligation or chemoembolization. The inferior phrenic artery (IPA) projected eight notable branches: ascending, descending, IVC, superior suprarenal, middle suprarenal, esophageal, diaphragmatic hiatal and accessory splenic [5].

The knowledge of variations, in relation to the renal, testicular, suprarenal and inferior phrenic arteries is important to the vascular surgeons and urologist and oncologist, during surgery of the retroperitoneal region [6].

**Case Report**

During the routine dissection of anatomy in slims, in an adult male cadaver, abdominal part of the aorta was traced. While tracing below superior mesenteric artery a ventral branch was running to the right kidney as it was traced, it is aberrant right renal artery, in the midway to the right kidney a small branch was running downwards, tracing further it entered into the spermatic cord, artery of testis [Fig. No.1].

In order to confirm that it is unilateral or bilateral the left side kidney was examined and it was found to be absent and the left testicular artery direct branch of the aorta [Fig. No.2].

On further inspecting on left side, only the left supra renal gland was present, the left testicular vein and left supra renal vein joins together and drain into the inferior vena cava.

After cutting through the right renal vein the right renal artery was traced and found that two branches was arising from the right renal artery, which supplies the right supra renal gland another continues up and it gives another branch to right supra renal gland which divides into right & left phrenic artery [Fig. No.3]. The left phrenic artery runs above the coeliac trunk over the abdominal part of aorta & inferior vena cava.
The left phrenic artery also gives a branch to left supra renal gland [Fig.No.4]. On tracing the left supra renal artery, it arises from the left lateral aspect of the abdominal part of the aorta.

**Discussion**

In 1883 P.S. Abraham reported the Congenital absence of kidney major were due to autopsy finding[7]. MEREDITH E. CAMPBELL reports in 1910 the incidence of solitary kidney were 1 in 1610 [4]. McPherson E, Indicates that, the incidence of bilateral renal agenesis in offspring of congenital solitary kidney probands is 0.8 % [8]. In 166 fetuses, Luciano A. Favorito reports renal anomalies were found in two fetuses 1.2%. Unilateral renal agenesis was found in a 25 WPC fetus [1].

In recent study shows agencies of kidney are associated with congenital anomaly. Congenital anomalies may sometimes be associated with single or multiple organ anomalies. The combination of urogenital anomalies, including unilateral kidney agencies associated with ipsilateral ectopic ureteric orifice, blind megaureter and bicornuate uterus [9-11]. Kouji Masumoto found absence of multiple organ anomalies of 3 or more major ipsilateral organs, namely the left lung, kidney, and upper limb or gastrointestinal anomalies are estimated to be 58.8%, 26.5%, or 10.3%, respectively [12].

Hung-Meng Huang indicates the prevalence of hearing impairment in the children with unilateral renal agenesis was 5.3% (4/75) [13]. HITOSHI OH-OKA has found the Diagnostic imaging revealed agenesis of the right kidney, associated with cystic dilation of the right seminal vesicle, and a prostatic utricle cyst [14]. Konanki Ramesh reports the association of moyamoya disease with renal agenesis and external iliac artery stenosis [15].

These kidney associated congenital anomaly are mainly based upon the mutation of genes like GDNF, SIX1&4, & PAX SERIES, c-ret & BMP4. Michael A. Skinner studied the DNA from 33 stillborn human fetuses with renal aplasia or severe dysplasia. Human RET mutations were found in 37% of fetuses with bilateral renal agenesis, 20% of fetuses with unilateral renal agenesis & GDNF Mutations in one fetus with unilateral renal agenesis (2). Hiroki Kobayashi reports Six1 mutation is associated with the branchio-oto-renal syndrome, which leads to kidney or urinary tract malformation by addition, deletion & overlaps of SIX1 and SIX4 loci which regulate the level of Gdnf expression [16].

D. Alan Stahl
Glial cell line-derived neurotrophic factor (GDNF) signals through a multi-component receptor system consisting of the transmembrane c-ret protein tyrosine kinase and glycosyl-phosphatidylinositol anchored co-receptor (Gfra1e4, and specifically Gfra1). In c-ret mutants, renal agenesis, severe hypoplasia, and blind-ending ureters were observed [17].

Existence aberrant renal artery of anatomic variations in the origins of the RA was first reported by Bartholin 1655–1738 [18]. The origin site of the right RA had several variation patterns. A double hilar artery in 11.1%, inferior polar artery in 10.5%, and superior polar artery in 3.3% of specimens. Anatomical variations were observed more frequently among male fetuses and on the right side [19]. Presence of aberrant renal artery on both sides is seen in 13–16% of cases [20]. Abberent renal artery enters into the kidney from the ventral side of the abdominal aorta [21]. Which is similar to our findings.

Aynur EmineCic-ekcibas-I says that incidence of gonadal artery arising from additional renal arteries has a wide range of variation in fetuses [22], SONI S reports in his study 8.7% of testicular arteries from the right inferior polar renal artery [3]. In Notkovich study it was found to be 14% [23], and in Pai M M study it was found to be 4.7% in right side. This anomalous pattern of origin of the testicular artery from inferior Abberent renal artery was seen in five specimens’ 7.4%, three on right side & two on left [24].

Keishi Okamotoa reports the InfRAs which arose at the lower level were observed in 24/270 cases on the right side & has classified the accessory renal artery. In Type I, the InfRA passed in front of the ureter and in Type II it passed behind the ureter. He also reports On the right side among the InfRAs of Type I, gives arise to gonadal artery (testicular/ovarian), [25] this is correlating with our study.

P Bordei E.D S studied 120 cases. The inferior suprarenal artery originated from the renal artery in 75 cases (62.5%); from the trunk of the renal artery in 58 cases (48.33%), [26–28]. J.C. Manso states the superior suprarenal artery originated from the posterior branch of the ipsilateral inferior phrenic artery in 83.3% on the right and 80% on the left. The origin of the middle suprarenal arteries from the trunk of the inferior phrenic artery is 26.7% on the right and 36.7% on the left. The arteries of the inferior group were branches of the ipsilateral renal artery in 70% on the right and 50% on the left [29] which relates to our study.

SONI S states the right inferior phrenic artery origins from the aorta, celiac trunk, & right renal artery; in 9% of the cases studied, it arises from right renal artery [3]. Anupama D examined the IPA Origin accessory RIPA which was arising in common with accessory renal artery and middle suprarenal artery on the same side of the Right renal artery [30], on contralateral which resembles our study. M. Loukas, to conduct an anatomical and a comparative study of the distribution of the IPA in normal and HCC cadavers. 17% of inferior phrenic artery commences from renal artery [5]. Akio Hiwatashi inspected CT of RIPAs during routine radiological scan and found that out 180 patients, 26 have different origin of RIPA, abdominal aorta in 18, celiac artery in 4, right renal artery in 2 and left gastric artery in 2 [31]. Masashi Hieda found 20 cases (14.3%) anterior branch of the LI RIPA commences from the RIPA. Of these 20 cases,
the RIP A arise from the right renal artery in 2 cases 10% [32] and in Thejodhar Pulakunta the RIP A arise from the right renal in 1 case [33], which resembles our study. S. KIMURA, the RIP A originates from RRA in 19 (11%) which resembles our study. In almost all cases RIP A originated from the celiac artery (CA) or right renal artery RRA [34].

The right renal artery bifurcated distal to its origin behind the inferior vena cava into upper and lower divisions. The upper division passed laterally towards the kidney and ended slightly proximal to the renal hilum by dividing to 2 branches. As the upper division passed laterally, it gave a branch (right superior polar artery) which ran towards the superior renal pole and entered the kidney. Interestingly, this branch also gave origin to inferior suprarenal and a muscular branch to diaphragm like our study [35, 30, 5].

Conclusion

Knowledge of these variations may also provide safety guidelines for endovascular procedures like therapeutic embolisation and angioplasties. Multiple vascular variations near the hilum of the kidney, testicular, suprarenal and inferior phrenic arteries are important during surgical procedures like varicocele, undescended testis & renal transplantation. Interventional radiologic procedures and renal vascular operations more safely and efficiently. A sound knowledge of possible variations is very useful for radiologists, urologists, oncologist and vascular surgeons. This anomaly is important in surgical procedures related to the posterior abdominal wall, renal transplantation, abdominal aortic aneurysm, ureter surgery, retroperitoneal region, hepatocellular carcinoma and the vascular pedicles of the kidney.

Acknowledgment

This work is dedicated to my mother (late) Mrs.N.Selvambal, I also thank my father, brother, wife & colleague who helped to complete this article & I also like to thank the management of Sri Lakshmi Narayana Institute of Medical Sciences, Puducherry.

References


