ANATOMICAL STUDY ON RENAL AGENESIS

Mary Hydrina D'Silva, Anbalagan J,SudhaRao, Rijied T Swer,Sadeesh T.

Department of Anatomy,
Mahatma Gandhi Medical College & Research Institute, Pillaiyarkuppam, Puducherry-607403

ABSTRACT

Congenital absence of kidney, unilateral or bilateral, is reported to be 1:1000 and 1:10000 births respectively. Unilateral agenesis is an ultrasonagraphic finding in adult with compensatory hypertrophy of the solitary kidney. Renal agenesis is generally associated with genitourinary anomalies like defective Mullerian duct, unilateral/bilateral absence or hypoplasia of seminal vesicle, vas deferens and adrenal agenesis. Unilateral agenesis is also reported to occur in X-linked Kallmann's syndrome. The present case is of a 33 weeks old male fetus medically terminated on the basis of ultrasonagraphy report of anhydraminos and suspected bilateral renal agenesis. Autopsy findings showed left unilateral agenesis, with dilation of the pelvic part of ureter and cryptorchidism on the same side. A study on microstructure of kidney, ureter, vas deferens, seminal vesicle, testes and suprarenal were carried out to observe the structural details.

Key words: Renal agenesis, congenital anomalies, genitourinary tract.

INTRODUCTION

Renal agenesis, unilateral or bilateral, is a clinical condition where the fetal kidneys fail to develop leading to oligohydraminos resulting in perinatal mortality. Renal agenesis is also due to lack of induction of metanephric blastema with ureteric bud or maldevelopement of mesonephric duct. The solitary kidney may also be the result of postnatal involution of multicystic kidney[1]. The incidence of unilateral agenesis of kidney is 1:1000[2]. The renal agenesis has also been reported to be due to teratogenicity of diabetes mellitus, use of renin angiotensin inhibitors with high doses of vitamin A derivatives[3], Chlorambucil[4], and cocaine abuse[5]. However the exact etiology of renal agenesis is still unknown.

Congenital absence of kidney is more prevalent in male.The incidence of bilateral renal agenesis is reported to be one or two in every 10000 births1. Renal agenesis is closely associated with ipsilateral congenital anomalies ofgenito-urinary systemin male and female[6,3,7]; anomalies of cardiac and skeletal systems[8]. In addition, the renal agenesis has also been reported to be associated with X-linked dominant trait and in Kallmann's syndrome [9,10].

Correspondence
Mrs. Mary Hydrina D'Silva
Assistant Professor of Anatomy
Mahatma Gandhi Medical College & Research Institute
Pillaiyarkuppam (Cuddlaore-Pondy Main Road)
Puducherry 607402
e-mail:hydriswer@gmail.com - phone:09894471980

CASE REPORT

A second gravida mother during her antenatal checkup was advised to undergo Medical termination of pregnancy on the basis of USG report showing anhydraminos, suspected renal agenesis (bilateral/unilateral),normal urinary bladderand two umbilical arteries and one vein. The 33 week old male fetus was terminated and was handed over to the department of Anatomy for detailed study. The external features of the fetus were normal, except the right testis was observed in the scrotal sac and theleft scrotal sac was empty.

OBSERVATIONS

The right kidney was observed in normal anatomical position, the upper part of ureter was slightly dilated and the ureteric opening was seen in the trigone of the bladder. The left kidney was absent, the pelvic part of ureter was dilated and ureteric opening in the trigone of urinary bladder was absent (Figure 2a, 2b). The left testis was found in the left iliac fosse. The suprarenal, seminal vesicles and vas deferens on both sides were normal. The thoracic, pelvic & abdominal organs were normal. The tissues from kidney, ureter, bladder, vas deferens, seminal vesicle and umbilical cord were processed for histological study.

Kidney: The right kidney was seen in sub-hepatic region. It was well covered with the renal capsule (dimensions 13x11x3 mm) and hilum showed renal vessels and ureter. The external surface of kidney was smooth without any lobulations. Microscopically the

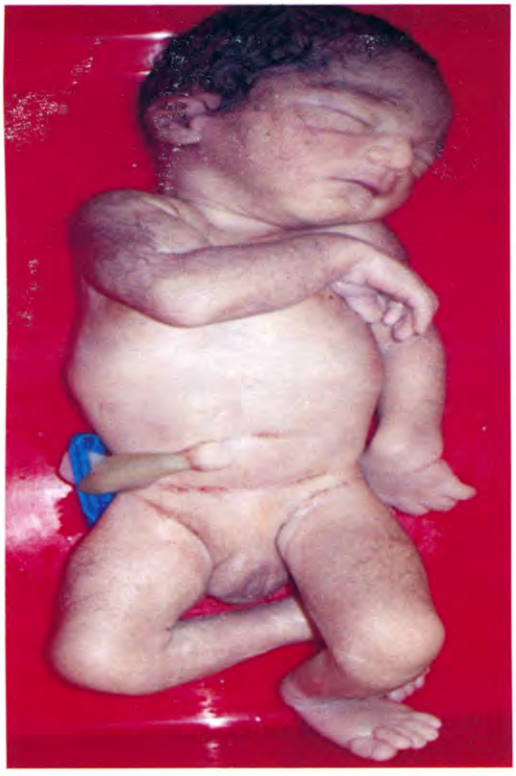

Fig.1. Phtograph of 33 wks fetus with normal features

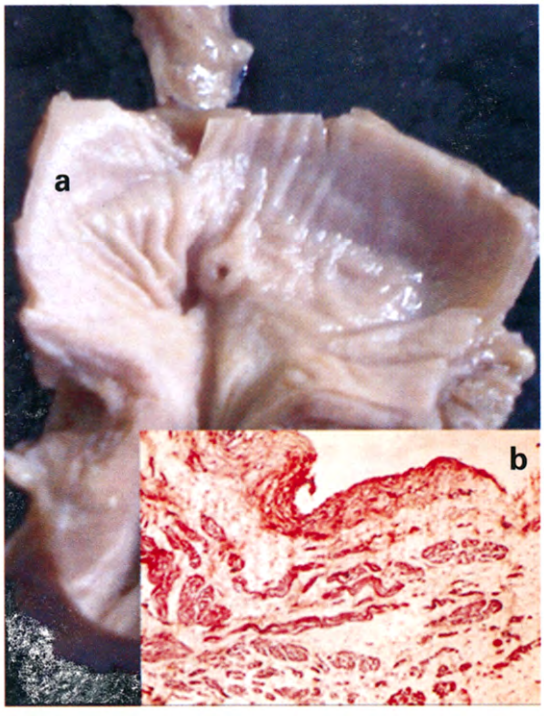

Fig 2

2a Photograph of trigone of urinary bladder showing blunt opening of left ureter.

2b (insert) Photomicrograph of urinary bladder showing normal structure.

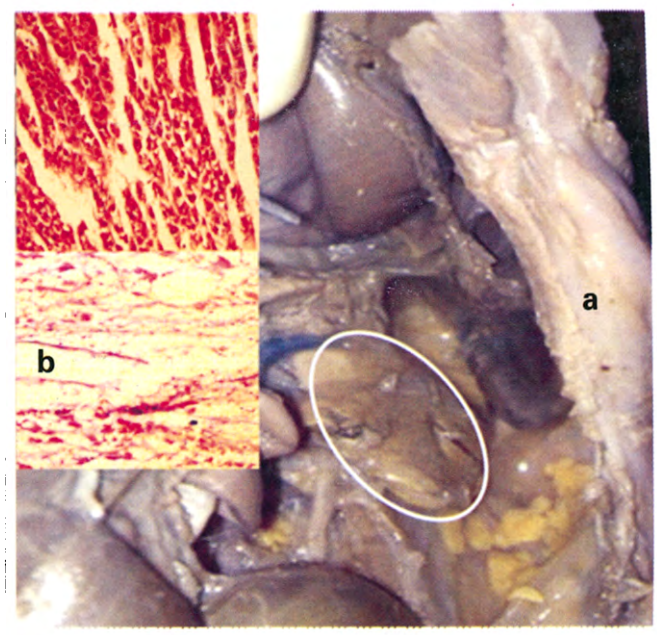

Fig 3

3a- Photograph showing left suprarenal (circled)

3b - (insert) Photomicrograph of suprarenal showing cortex & medulla.

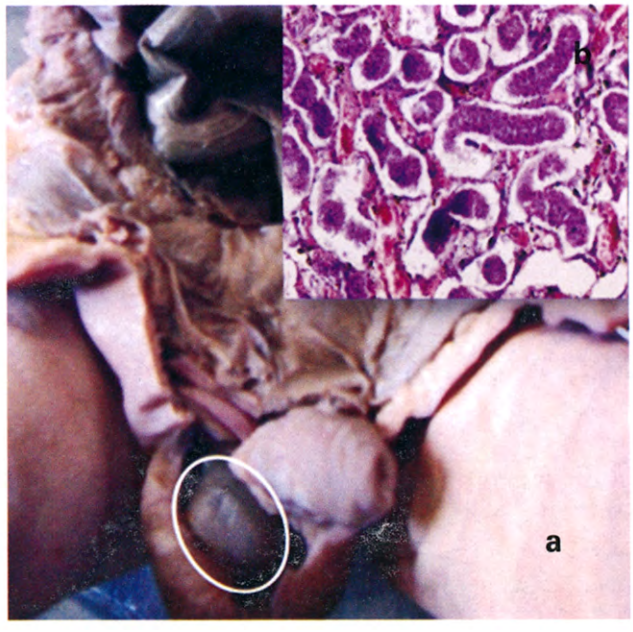

Fig 4

4a - Photograph showing descended right testis (circled)

4b - (insert) Photomicrograph of right testis showing undifferentiated seminiferous tubules

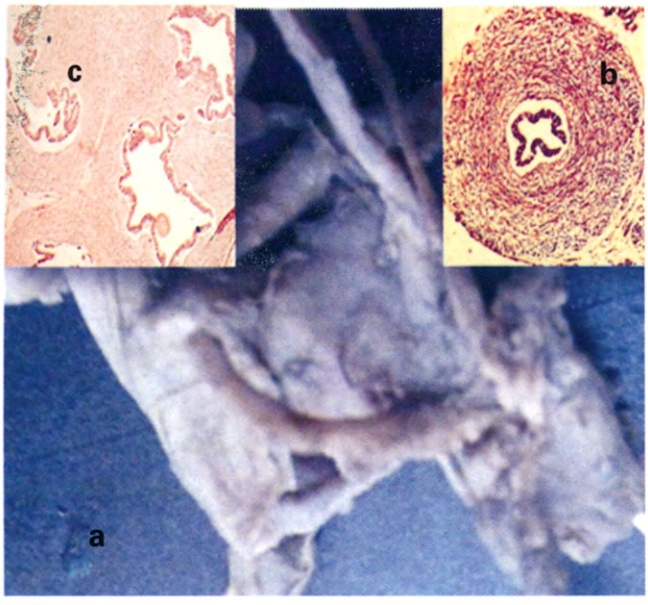

Fig 5

5a - Photograph showing vas deferens and seminal vesicle.

5b - (insert) Photomicrograph of vas deferens showing normal features.

5c - (insert) Photomicrograph of seminal vesicle showing normal features.

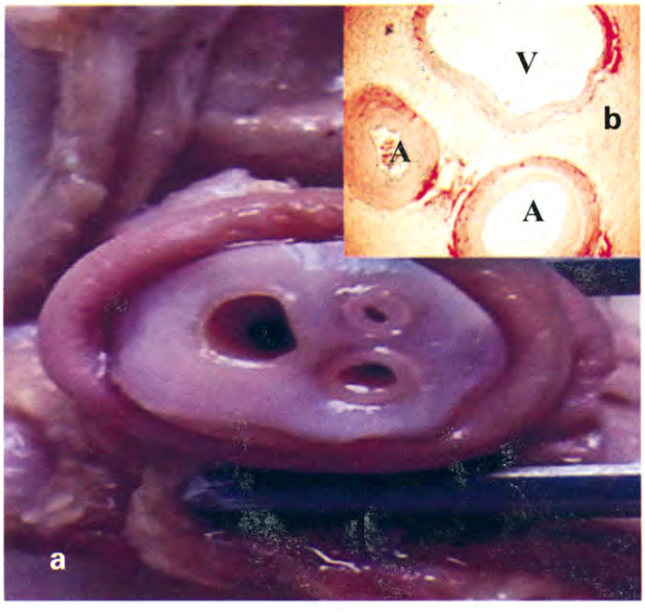

Fig 6

6a - Photograph of umbilical cord showing two arteries and one vein .

6b - (insert) Photomicrograph of cord showing dilated and thin umbilical artery.

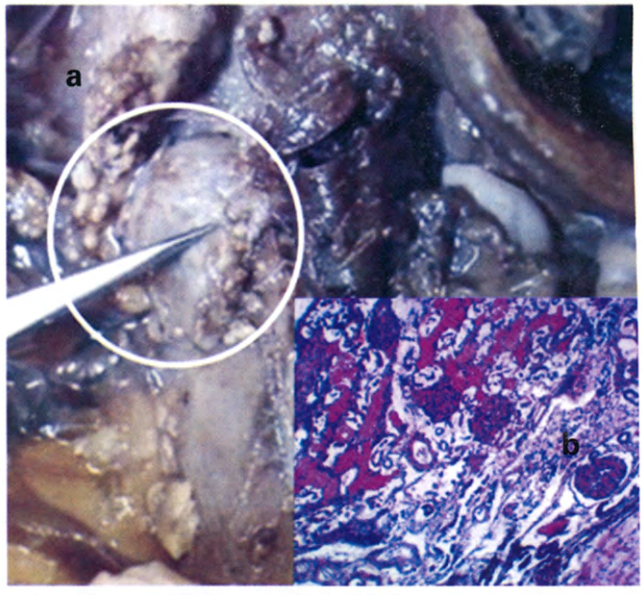

Fig 7

7a - Photograph showing Right kidney (circled)

7b - (insert) Photomicrograph of Right kidney showing few hypo plastic glomeruli & renal tubules.

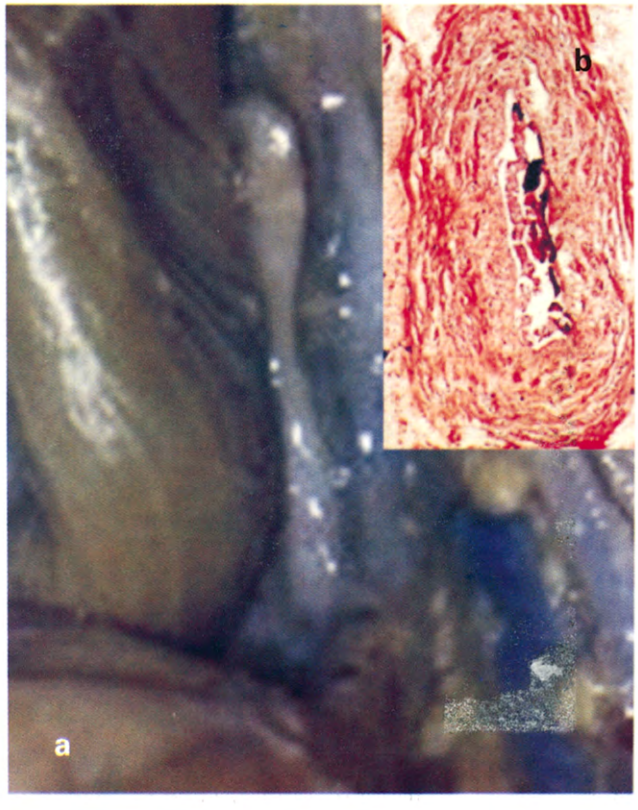

Fig 8

8a - Photograph showing pelvic part of left ureter.

8b - (insert) Photomicrograph of left ureter showing ill defined transitional epithelium & muscular layer.

cortex of the kidney showed fewer undifferentiated, hypoplastic glomeruli surrounded by renal tubules (Figure 7a, 7b).

Suprarenal: Both the suprarenal glands were observed in anatomical position with vascular connections. The microstructure showed three cortical zones and scattered medullary cells surrounding the venules (Figure.3a, 3b).

Ureter: The upper part of right ureter was slightly dilated and the vesico-urethral opening was observed. The left ureter was present and showed pelvic dilatation, however vesicoureteral opening in trigone of urinary bladder was absent. Neither the remnants of degenerated nephric tissuenor any atrophic renal vessels were observed.

Histologically, both ureters exhibited all the basiclayers except left ureter showing less developed epithelium and the muscular layer. (Figure 8a, 8b).

Urinary bladder: The urinary bladder was normal,the trigone of the urinary bladder showed right ureteric orifice and the left ureteric opening was absent. Microscopically the mucosal folds showed transitional epithelium and a well-defined muscular layer (Figure 2a, 2b).

Testes: The right testis with all its coverings was observed in the right scrotal sac (dimensions: 10x4x4 mm). The epididymis, vas deferens, testicular vessels, appendices of testis and epididymis were present. The undescended left testis was found in the left iliac fosse (dimensions 9x5x2mm). Microscopically, both testes showed seminiferous tubules with undifferentiated clusters of spermatogonial cells. Interstitial cells of Leydig were observed in between the tubules (Figure 4a, 4b).

Epididymis: It was observed in both the testes and the microstructure showed cut sections of empty tubules lined by pseudo stratified epithelium with few cilia.

Vas deferens and Seminal vesicle: They were present on either sides of the posterior surface of urinary bladder. Histological features of seminal vesicles showed mucosal folds with some secretions in the lumen. The vas deferens showed narrow lumen with well defined muscular layer and a vascular connective tissue (Figure.5a, 5b, 5c).

Umbilical cord: Macroscopic and microscopic observations showed two arteries and one vein. However, one artery showed thin tunica media (Figure.6a, 6b).

DISCUSSION

Renal agenesis is associated with other congenital anomalies like maldevelopement of Mullerian duct, unilateral/bilateral agenesis of vas deferens, seminal vesicle, hemi/complete absence of trigone of bladder, oligohydraminos and one umbilical artery in the umbilical cord[7,6,5]. In the present case, the USG report showed anhydraminos and presence of two arteries and one vein in the umbilical cord. However, one artery appeared dilated.

Renal agenesis may be unilateral or bilateral. Bilateral renal agenesis is a rare anomaly incompatible with life. In the present case the male fetus showed left renal agenesis, with presence of pelvic part of ureter and absence of ureteric opening in the bladder. A solitary right kidney appeared normal without lobulations. Adrian S Woolf et al., (2006)[3] observed in his study that if the solitary kidney is of normal size, it is eitherhypoplastic/dysplastic. According to him a solitary functional kidney is always hypertrophied. In the present case, our findings are in accordance with Adrian S Woolf et al., (2006)[3] as the histological features of the right kidney appeared to be hypoplastic.

Renal agenesis occurs due to the absence of metanephric blastema, ureteric bud maldevelopement or lack of induction of metanephric blastema by the ureteric bud. In the present case, the absence of metanephric blastema and failure of induction of ureteric bud could have resulted in left renal agenesis. The renal agenesiscould also be due to the absence of transcription factor WT1 that influences growth factor FGF-2 and BMP-7 to prevent apoptosis of metanephric cells or failure to convert metanephric cells into nephric epithelium by regulatory genes PAX2 and WNT4 from ureteric bud[11].

It has been reported in clinical studies, that the solitary kidney shows hypertrophy to compensate for the absence of the other kidney. In long run such kidney usually develops hypertension, proteinuria and glomeruloscelerosis[3]. The solitary adult kidney may show infarction, suppurative focal nephritis, and chronic interstitial nephritis with poor prognosis. The presence of solitary kidney usually goes undetected, sometime is detected accidently in ultrasonograph; otherwise unilateral agenesis is an autopsy finding.

In the present case, the solitary kidney showed histological features of hypoplastic kidney. The right ureter showed dilatation. It could be due to the obstruction of the uretero-vesical junction which is reported to be a common anomaly associated with

unilateral renal agenesis 8. The solitary kidney usually appears in ectopic position, but in the present case it was found in the normal position. The hypoplastic condition of glomerulus could have resulted in renal failure.

CONCLUSION

Although unilateral renal agenesis is common, its definitive etiology is unknown. USG report may fail to differentiate between renal agenesis and dysplastic/hypoplastic kidney as in the present case.

Unilateral renal agenesis may or may not be associated with urogenital defects as seen in the present case where the testes, vas deferens seminal vesicle were all normal. An early descent of the right testis was observed in the present study.

As the solitary kidney has to compensate for the absence of the other kidney, the patient with unilateral agenesis needs to have life-long nephrological care. Unilateral renal agenesis is inherited as X-linked dominant trait and also associated with Kallmann's syndrome. In such conditions, the parents and the siblings need to be screened for renal agenesis, and proper counseling needs to be given.

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REFERRENCE

1 .Mishra.A, Renal agenesis. The British Journal of Radiology, National Organ Transplant Program, Tripoli Central Hospital, Tripoli, Libya. 2007;80:e167e169.

2. Kiprov.D D, Calvin.R B, Mchuskey.R T. Focal and segmental glomerulosclerosis and proteinria associated with unilateral agenesis. Lab.Invest. 1982; 46:275-81

3. Adrian S. Woolf, Katherine A Hillman. Unilateral renal agenesis and the congenital solitary functioning kidney: Developmental, Genetic and Clinical perspectives. 2006.

4. Steege John F, Caldwell, David S. Renal agenesis after first trimester exposure to chlorambucil, Southern Medical Journal 1980 (PUB MED)

5. Maki kashiwagi, RabihChaoui, Thomas Stallmach, Sandra Hurlimann, UrsLauper, GundulaHebisch. Fetal bilateral renal agenesis, phocomelia and single umbilical artery associated with cocaine abuse in early pregnancy. Birth Defects Research (Part A) 2003; 67:951-952

6. Amin J B. Association of unilateral renal agenesis and genital anomalies.- Case Report.Clin.Pract.Rev 2002;3(2):57-60

7. McCallum T J, Milunsky J M, Munarriz R, Carson R, Sadeghi-Nejad, Oates R D Unilateral renal agenesis associated with congenital bilateral absence of the vas deferens: phenotypic findings and genetic considerations- Human Reproduction 2001; 16(2): 282-2884.

8. GrazynaKrzemień, Maria Roszkowoska-Blaim, IzabellaKostro, Urological anomalies in children with renal agenesis or multicystic dysplastic kidney. J Appl Genet. 2006; 47(2):171176.

9. Georgopoulous .N A, Koika.V, Galli-Tsinopoulou.A, Spiliotis.B E, Adonakis. G, Keramida.M K, Sgourou.A, Koufogiannis.K D, Papachatzopoulou.A, Papavassiliou,A G, Kourounis,G, Vegenakis.G A. Renal dysgenesis and KAL I gene defect in patients with sporadic Kallmann's syndrome. Fertil.Steril.2007;88(5):1311-17 E Pub 2007 July 2

10. Wegenke J D, Uehling D T, Wear J B Jr, Gordon E S, Bargam J G, Deacon J S, Herrmann J P, Pitz J M,FamilialKallmann's syndrome with unilateral renal aplasia. Clin Genet. 1975; 7(5):368-81

11. Sadler T W, Langman's Medical Embryology in Urogenital System. 9thEd. Lippincott Williams & Wilkins Philadelphia Pennsylvania USA 2000 pp 327-8