

CASE REPORT

DEVELOPMENTAL DISORDER OF MEDULLARY CYSTIC KIDNEY WITH EXTRA BILATERAL LOBE: A RARE CASE REPORT

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ABSTRACT: The knowledge of anomalous kidney vital to Anatomist, Neurologist, and Urologist for better treatment and understanding of the function of the organ. The both kidneys contains extra two lobes of renal dysplasia on its upper pole. In Adult onset of Medullary cystic kidney disease (MCKD), shows abnormal sacs called cysts, which present in the collecting tubes were observed in regular dissection for first year undergraduates in 58 years old female cadaver and incidence rate is 1: 20,000 in live birth. The MCKD is Non-inheritable condition is developmental disorder characterized by the cystic dilation of the collecting tubules in renal pyramids in both kidneys.

KEYWORDS: MCKD, Collecting tubules, renal dysplasia.

INTRODUCTION: The kidneys regulate the fluid and salt to balance the body and blood flow through the kidney is one fifth of the cardiac output per minute. Thus 1700litres of blood passes through the kidneys in one day, out of which 170 liters of glomerular filtrate is formed and only about 1.5 liters of urine is excreted. The dilation of the collecting tubules gives spongy appearance causing Medullary cystic kidney (MCK)⁽¹⁾ and do not usually appears until the age of 30 to 40 years. MCKD constitutes a group of renal cystic diseases that share the macroscopic feature of cyst development at the corticomedullary border of the kidneys.⁽²⁾ Autosomal dominant medullary cystic kidney disease (ADMCKD) with hyperuricemia and very late age of onset (mean 62.2 and 51.5 years).

CASE REPORT: During regular dissection of Abdomen region for first year medical undergraduate in department of Anatomy, Government Medical College, Bidar (Karnataka), the bilateral extra lobes of renal dysplasia additionally connected with upper pole of right and left kidneys was observed in female cadaver and absence of right supra renal gland. The right kidney is about 11cms in length and left kidney 12cms in length and both lateral side of the kidney are irregular. The internal structure shows cyst present at cortico-medullary junction and some cyst observed at medulla, which is 4 to 6mm in diameter and the cyst effecting renal pyramids and small area of cortex in both kidneys. The cysts is rounded or oval of varying size and large renal cyst present in bilateral extra two lobes and cyst shows neoplastic changes in both kidneys. The MCKD causes calcium to deposit and repeatedly form stones and growth of abnormal blisters of fluid (cysts) in the kidneys.⁽⁴⁾ Necropsy findings reveal gross or microscopic cysts in the medullary region and cortico-medullary junction of both kidneys. The development disorder of cyst presents Familial nephronophthisis (40%),⁽²⁾ Sporadic (20%), Retinal renal syndrome (15%) and Adult onset medullary cystic disease (15%).

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Clinical significance of congenital renal variants

Renal cyst - A fluid-filled sac arising from a dilatation in any part of the nephron or collecting duct.

- Antenatal hydronephrosis common
- Renal function: Obstructive lesions often associated with renal dysplasia

Developmental disorders in collecting variants

1. Medullary sponge kidney
2. Multicystic dysplastic kidney

Due to dilated collecting ducts shows "blush" in papillae on IV contrast studies

Increased risk of MCKD

Nephrolithiasis (50-60%)

Hypercalciuria (at least 33%)

Urinary tract Infection (20-33%)

Hematuria (0-18%).

The cysts were unilocular and smooth-walled. There was no communication between cysts and pelvis.

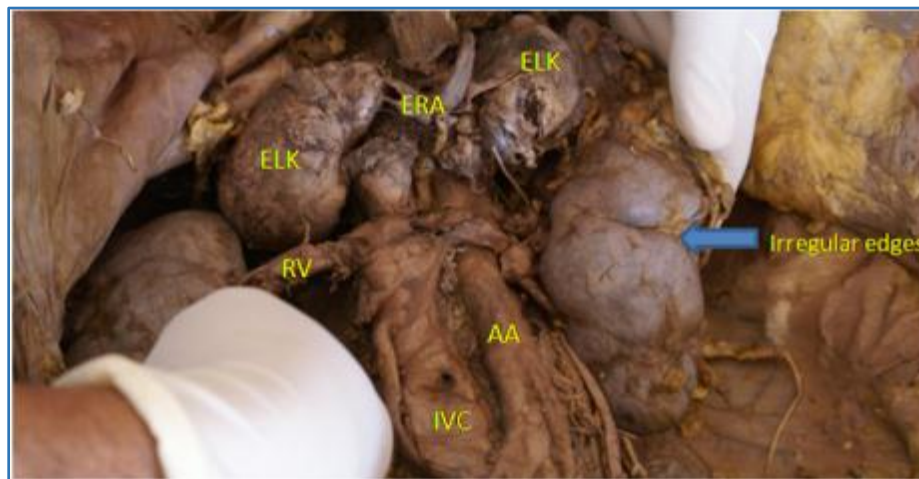


Fig. 1: Both kidneys consists bilateral extra lobe and its lateral edges are irregular

Abbreviations: ELK – Extra lobe of kidney; ERA – Extra renal arteries; RV – Renal vein; AA – Abdominal aorta; IVC – Inferior vena cava.

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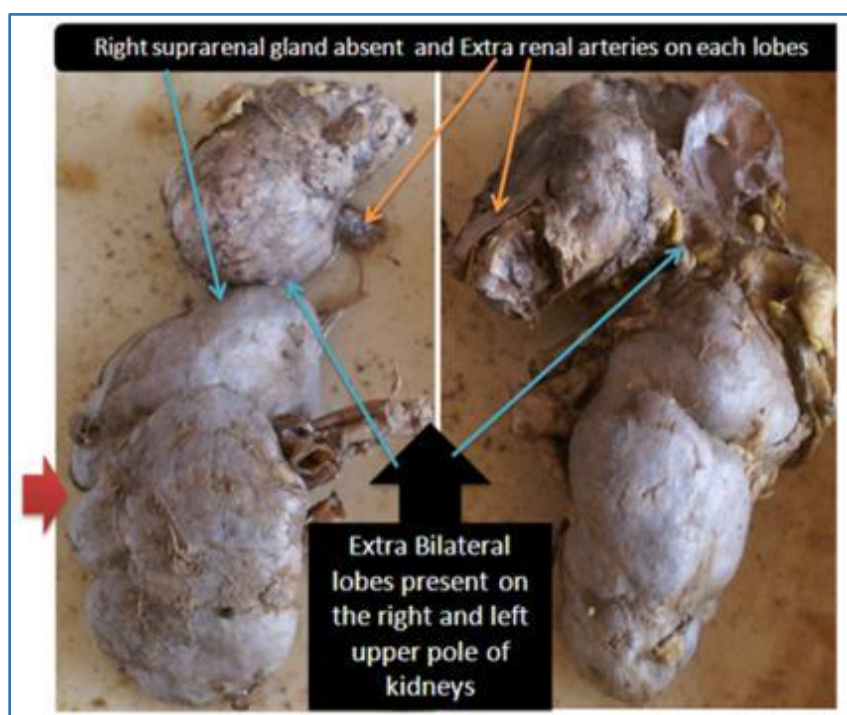


Fig. 2: Shows irregular lateral edges of kidney with extra bilateral lobes supplied by the extra renal arteries from abdominal aorta. Absence of right supra renal gland

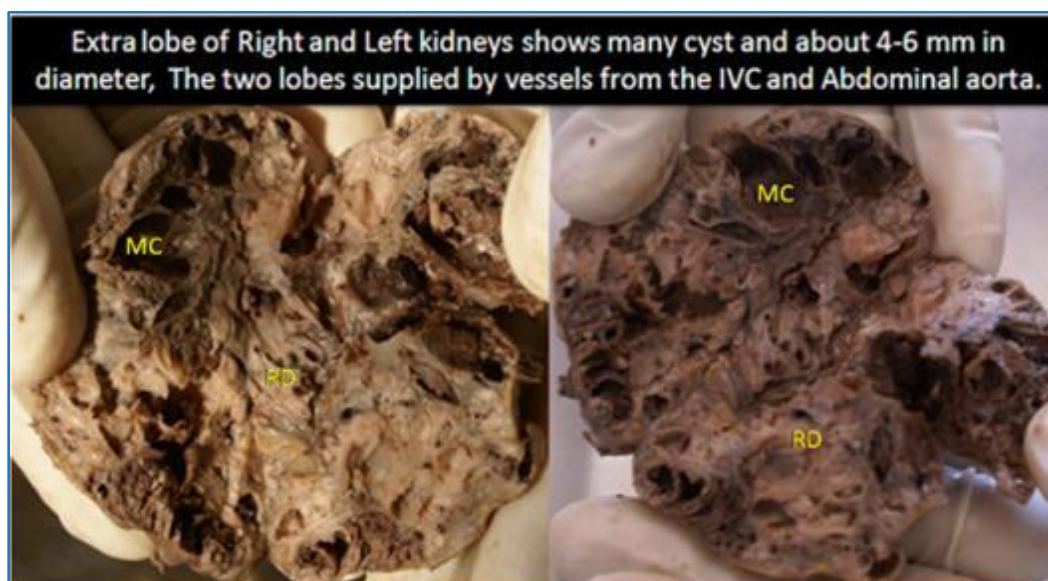


Fig. 3: Extra lobe of kidney present renal dysplasia

Abbreviations: MC – Medullary cysts; RD – Renal dyplasia

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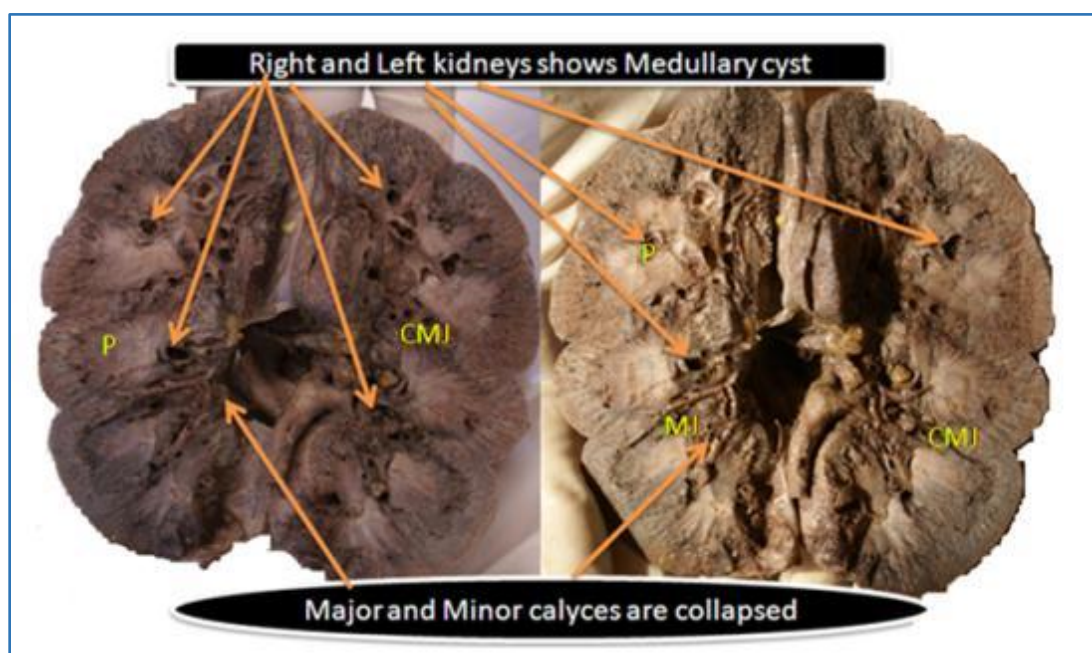


Fig. 4: The coronal section of both kidneys present medullary cysts in pyramid (Renal medulla) and corticomedullary junction. The medullary cysts about 4 to 6 mm in diameter

Abbreviations: P – Pyrmad; CMJ – Cortico-medullary junction; MJ – Major calyces

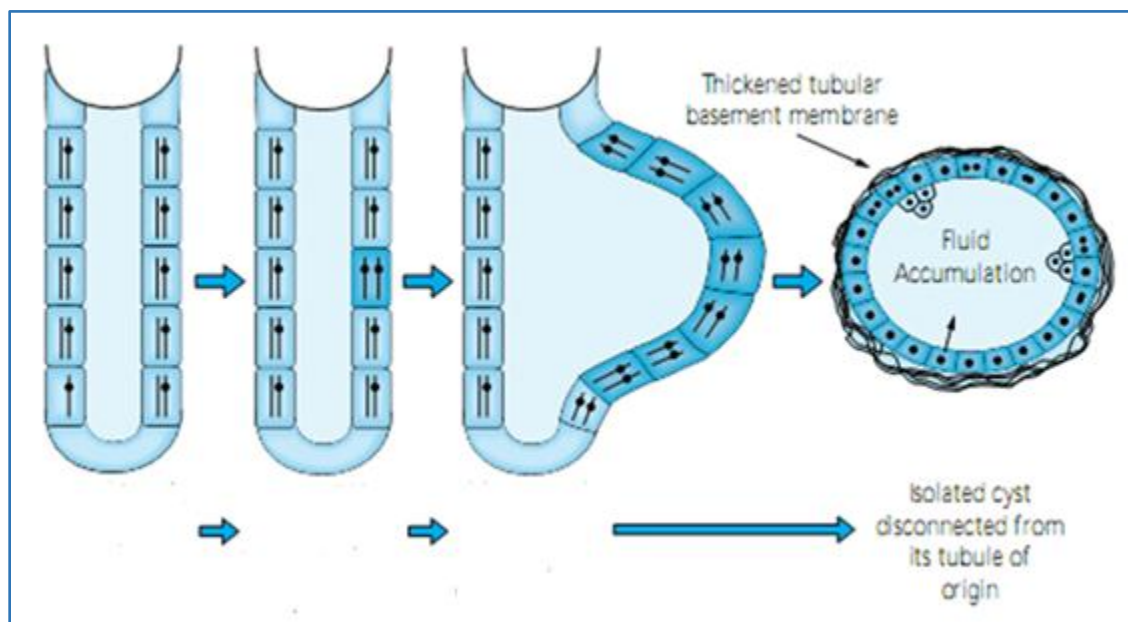


Fig. 5: Hypothetical study of the formation of medullary cysts, which contains accumulation of the fluid.

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DISCUSSION: Medullary spongy kidney disease is first recognized by G Lenarduzzi, a Radiologist and his colleagues Cacchi and Racci, in 1939; hence, also known as Lenarduzzi-Cacchi-Ricchi disease. MCK a progressive renal disorder, first described by Smith and Graham (1945) was reviewed by Strauss in 1962. The cyst changes in the kidneys are encountered in a variety of anatomical, Nephrology, and pathological disorders. The kidneys lose their ability to remove enough fluid from filtered wastes and pass great amount of undiluted urine.

MCKD is an autosomal dominant form of tubulointerstitial nephropathy characterized by formation of renal cysts at the corticomedullary junction. Mongeau & Worthen (1967) in their review of the literature found twenty cases with autopsy data and noted macroscopic and microscopic cysts in eight and twelve instances respectively.⁽¹⁾ The morphological findings in the kidneys were well summarized by Ivermark, Ljungqvist & Barry (1960). The gross and microscopic appearances of the kidneys in both our cases are strikingly similar to those described by Strauss (1962) and others (Hogness & Burnell, 1954; Kerlan & Russell 1963).

It is characterized by adult onset of impaired renal function and salt wasting resulting in end-stage renal failure by the sixth decade (Wolf et al. 2004). Ala-Mello et al.(1999) used the term 'nephronophthisis' for both the dominant disorder called medullary cystic disease.⁽²⁾ Parvari et al.(2001) studied a family of Jewish ancestry in which 15 members spanning 4 generations had chronic renal failure with onset between 18 and 38 years of age. Wolf et al. (2004) reported Belgian kindred with MCKD. Kirby et al(2013) reported 6 unrelated families with MCKD,⁽¹⁾ including the families previously reported by Kiser et al(2004) and Kimmel et al(2005).

Affected individuals had slowly progressive kidney dysfunction beginning in adulthood, absent or low grade proteinuria with bland urinary sediments, decreased glomerular filtration rate, and absence of other association signs or symptoms of systemic disease. The cause of MCKD is unknown, but genetic transmission occurs in only <5% of cases and majority of the cases considered as development disorder.⁽³⁾

The cysts are filled with fluid by increasing the width of the collecting tubules; the normal flow of urine that passes through them is slowed or stopped. When the flow of urine is slowed down or stopped, infections and kidney stones can develop. The infections develop because the toxins in the urine are not passing out of the body quick enough. In addition, since urine is warm and bacteria thrive in warm areas, having a place in the body where warm urine is sitting promotes bacteria growth.

CONCLUSION: The multiple small medullary cysts with extra bilateral lobe of renal dysplasia variants are not mentioned in the literature; hence such variation should be given importance for practical and clinical demonstration to better understanding of the MCK. This kind of case report may be used as Problem Base Learning for clinicians and Interns for update their first hand knowledge.

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