Case Report on CAKUT Syndrome with Known Case of Chronic Kidney Disease

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Abstract

Background: Congenital abnormalities of the kidneys and urinary tracts (CAKUT) are conditions brought on by errors in the growth of the kidneys and the outflow tubes that drain the kidneys. Although the issue may not become apparent until later in life, CAKUT is a congenital abnormality brought on by improper urinary system development. Case presentation: We are presenting the case of a 23-year-old man who has chronic kidney disease and is undergoing maintenance hemodialysis. The patient was apparently alright 2 weeks back when he had complained of breathlessness. fever 2 episodes, lower which was insidious in onset and gradually progressive. On arrival, the patient was provided oxygen therapy via a face mask for breathlessness and to make him feel comfortable with the head elevation. On laboratory investigation, the patient’s blood values are altered. The patient underwent computed tomography of the kidney-ureter-bladder (CT-KUB), and Ultrasonography of the kidneys (USG). A permanent tunnel hemodialysis catheter under local anesthesia was placed and he received 2 cycles of hemodialysis. During the course in the hospital, the patient’s condition was improving and was stable clinically and vitally hence, the patient was discharged. Conclusion: CAKUT can occur alone or in conjunction with heart and cardiovascular malformations, gastrointestinal tract malformations, central nervous system malformations, skeletal system malformations, lung, face, genito-reproductive system malformations, abdominal wall malformations, chromosomal abnormalities, MCA, and other organ system malformations.

Keywords: Congenital anomalies of the kidneys and urinary tract syndrome, Cortical echogenicity, Shrunken, Hemodialysis.

INTRODUCTION

Congenital kidney and urinary tract abnormalities, which affect 1 in 500 fetuses that undergo ultrasonography exams, are widespread in humans. However, little is known about the molecular pathophysiology of these diseases, even though CAKUT is a significant contributor to chronic renal failure in newborns and young children. (1) There are frequently additional defects present in newborns with CAKUT. CAKUT is the most common cause of end-stage renal failure, occurring in up to 1 in 100 live newborns and 1 in 500 fetal ultrasonographic examinations. (2) Chronic renal failure in newborns and young children is primarily caused by CAKUT diseases. 10-15% of those who are affected develop associated abnormalities of other organ systems. (3)

CASE PRESENTATION:

We are presenting a case of a 23-year-old male who is a known case of chronic kidney disease and is on maintenance hemodialysis. He came with the chief complaints of breathlessness for 2 weeks, 2 episodes of fever, and bilateral lower limb swelling for 2 weeks.

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The patient was apparently alright 2 weeks back when he had complained of breathlessness. Fever 2 episodes, lower which was insidious in onset and gradually progressive. The patient has a history of ventricular septal defect 12 years ago when he was 11 years old, he was operated on for surgical repairing of VSD and he had not faced any complications after that.

On arrival, the patient was provided oxygen therapy via a face mask for breathlessness and to make him feel comfortable with the head elevation. Later a physical examination was carried out, and the patient’s look was dull. He has elevated blood pressure to 140/86mmHg. On inspection, the patient had bilateral lower limb swelling. In the abdominal palpation, both the kidneys were palpated where a left kidney is found less in size than the right one. The patient is advised to do ultrasonography of the kidneys. And afterward, the blood sample was drawn and sent for examination.

On laboratory investigation patient’s hemoglobin level was decreased to 5.8 gm % total RBC count was 2.91 million/cumm, urea was 100mg/dl, creatinine 6.5 mg/dl, albumin was 2.8g/dl also the urine routine shows the presence of albumin in the urine. These findings of blood tests were progressively decreased and then gradually got normal.

On radiological examination, Ultrasonography of kidneys shows Right kidney is 7×3.5 cm there is e/o raised cortical echogenicity with loss of CMD S/O Grade III Renal Pelvic Diameter. The left kidney is not visualized in the left renal fossa. After this examination patient was advised to do the CT-KUB, which reveals that the Right kidney measures 6.3×3.8 cm there is e/o minimal perinephric fat stranding. The left kidney appears small and shrunken measuring 2.8×1 cm. Shrunken left kidney and minimum perinephric fat stranding around the right kidney. Through this finding, the patient was diagnosed with CAKUT syndrome with a known case of CKD on MHD.

The patient has done 2 cycles of hemodialysis after the placement of the permanent tunnel hemodialysis catheter under local anesthesia. And afterward, he has treated with Inj Ferri 100mg, Tab Lasix 20mg, Tab Met Xi, Tab Minipress 5 Mg Hs, Tab Nicardia 20mg, Tab Shelcal 500mg, Tab Sobisis, and Tab Folic Acid. During the course in the hospital, the patient’s condition was improving and was stable clinically and vitally hence, the patient was discharged.

DISCUSSION:

About 30% of all congenital malformations identified during pregnancy are CAKUTs, making them one of the most prevalent. (4) Antenatal sonography, which examines the kidneys, outflow channels, and most important the quantity of amniotic fluid, is used to identify the most of CAKUT cases. Amniotic fluid is made mostly of urine from the fetal kidneys after the 18th week of pregnancy. In contrast to kidneys that are tiny, hyperechoic, and/or have cysts, deformed kidneys that exhibit these characteristics are referred to as "dysplastic kidneys." (5-11)

Prenatal detection of renal and urinary system abnormalities is 89% when routine prenatal ultrasound evaluation and modern ultrasound technology are used. The planning of integrated care for the baby and parent counseling both depend on an accurate diagnosis. (6) Problems with CAKUT frequently require surgical intervention or, in the worst-case situation, result in renal failure and demand hemodialysis and/or transplantation of kidneys. The improper interaction between the ureteral bud and the metanephric blastema results in several conditions, including renal hypoplasia, vesicoureteral reflux, and ectopic ureters, to name a few. However, the genetic and biochemical control of urinary tract development is not completely understood. (12-15)

Elke Wühl, Karlijn J. van Stralen et al suggest that there is defined information found on the long-term natural course of CKD because of CAKUT. Even though Renal Replacement Therapy incidence was higher in children than in adults, it was unexpected to see that 50% of patients did not need RRT until they were in their fourth decade of life, and more than two-thirds of patients advanced to ESRD by the time they were adults. (16)

CONCLUSION:

CAKUT can occur alone or in conjunction with heart and cardiovascular malformations, gastrointestinal tract malformations, central nervous system malformations, skeletal system malformations, lung, face, genito-reproductive system malformations, abdominal wall malformations, chromosomal abnormalities, MCA, and other organ system malformations. Children with various organ system defects, MCA, chromosomal aberrations, and neonates with substantial ear lobe abnormalities should be suspected of CAKUT. Early intervention helps to prevent kidney impairment and CKD, therefore knowing about this link is critical in managing affected patients.

REFERENCES


