Unilateral Renal Agenesis Associated With Anomaly of Left Testicular Venous Return

Author's Details:

Suresh Rao[1], Siva Konduru[2] & T. Ramesh Rao[1]

[(1)]Department of Preclinical Sciences, Faculty of Medical Sciences, the University of the West Indies, St. Augustine Trinidad & Tobago.[(2)]Consultant Radiologist, Medical Imaging Department, Sangre Grande General Hospital, Trinidad and Tobago.

Corresponding Author: Dr. Suresh R Rao

Introduction:

Unilateral renal agenesis is the most common congenital abnormality of the urinary tract [1, 2] and is characterized by the complete absence of development of one kidney accompanied by an absence of ureter and renal vessels. The incidence of unilateral renal agenesis in each year it is estimated at around one in two thousand and the incidence in male is slightly higher than females. Renal agenesis is caused by failure of the ureteric bud to induce development of metanephric blastema or by absence of the nephrogenic ridge [3].

The condition is usually asymptomatic and is commonly detected as a chance during renal ultrasound for the evaluation of a urinary tract infection, or in adults who present with hypertension. Most patients are asymptomatic if the other kidney is fully functional. Renal agenesis is occasionally associated with genital tract anomalies on the same side and also the other organs may show anomalies as well [4-6].

With the widespread use of antenatal ultrasound, renal agenesis can be identified in utero, although the presence of normal amounts of amniotic fluid and urine in the bladder results in the diagnosis being frequently missed, unless specifically and routinely looked for the features like absent kidney, absent ipsilateral renal artery and care must be taken not to mistake the low lying adrenal gland which is large in fetuses compared for a kidney. Careful examination of the rest of the abdomen should be carried out to ensure that an ectopic kidney is not present (more common than renal agenesis) as well as of the 'single' kidney to ensure it does not represent crossed fused renal ectopia. All imaging modalities such as postnatal ultrasound, CT and MRI will demonstrate the absence of a kidney, with the associated hypertrophy of the single kidney. Color Doppler interrogation may aid in showing absence of renal arteries.

Case Report:

A 61 year old male patient presented to urology department with the history of pain in the right loin and hematuria. Physical examination revealed a healthy looking with a weight of 82 Kgs, height 5.6 ft. and blood pressure 90/110. The patient was referred to the radiology department for a CT scan to rule out renal calculi or any bladder abnormality for the cause of hematuria. Patient has nil significant of past medical history. Family history was essentially negative. A renal protocol CT was performed on the patient. CT examination showed a small (<0.5cm) right upper pole renal calculus. Incidentally CT showed absent of left kidney either in its normal position or in ectopic location in abdomen and pelvis. There is no left renal artery or left renal vein evident. Also, the left gonadal vein appeared to be draining into the vein coursing from IVC to adrenal gland, suggesting left adrenal vein. Patient did not have any history of previous nephrectomy. There are no other associated anomalies in this patient. The case was discussed with the patient. Patient with left renal agenesis was diagnosed accidentally in sixties [Fig. 1].

Discussion:

Congenital unilateral renal agenesis is usually diagnosed on an incidental imaging examination and it occurs in 1-1.5 per 1000 autopsies [7–9]. The left kidney is more commonly involved than the right, and males are affected more than females. Normally embryogenesis of the kidney takes place in three stages: In Stage I the pronephros, develops from the intermediate mesoderm in the cervical region and then regresses at about four weeks embryonic age. It is replaced at about fifth week by the mesonephros, which develops from the paravertebral mesoderm of the upper thoracic to lumbar region and is also not functional. Stage II, the mesonephros disappears by the end of the second month of gestation. The few remaining caudal tubules adjacent to the testis and ovaries develop into the vas deferens in males and remnant tissue in females. Stage III the ureteric bud is an outgrowth from the mesonephric duct, invades a cord of mesenchymal cells called the metanephric blastema at 5 weeks of gestation. This invasion of the metanephric blastema by the ureteric bud leads to the formation of the definitive kidney, from the ureteric bud the major and minor calyces and the collecting ducts are formed. The invasion of the ureteric bud induces the mesenchymal cells of the metanephric blastema to form the glomeruli and renal tubules. This metanephric blastemal forms metanephros, which finally forms the definitive kidney, which starts to function at 6 to 10 weeks of gestation. The definitive kidney then migrates to the lumbar region.

Currently, the exact cause of unilateral renal agenesis is not known, it may be due to embryological, genetical or environmental factors. However, embryologically unilateral renal agenesis results when the ureteric bud fails to develop and also fails to induce the differentiation of the metanephric tissue. However, the solitary kidney is compatible with longevity, but may be prone to disease such as pyelonephritis, obstruction and calculus formation [10, 11]. Around half of patients with congenital unilateral renal agenesis have associated urological anomalies including vesicoureteral reflux, ureterovesical junction obstruction, ureteropelvic junction obstruction and others [4], and 25% of them have associated cardiovascular, gastrointestinal, skeletal abnormalities [9]. Unilateral renal agenesis may be an expression of a single dominant gene [12]. The association of mullerian agenesis and renal agenesis could be an autosomal dominant disorder [13, 14]. Buchta et al [15] described many generations of two families with hereditary renal adysplasia with or without mullerian anomalies. They suggested dominant inheritance. The patient in this report had unilateral renal agenesis associated with left gonadal vein draining into the left suprarenal vein.

Conclusion:

Early detection of a congenital solitary kidney by routine prenatal ultrasound or by incidental imaging studies should alert the physician to look for associated genital anomalies. Such knowledge is useful in avoiding unnecessary surgical procedures in patients presenting with abdominal or pelvic complaints.

References:

1. Parikh CR, McCall D, Engelman C, Schrier RW. (2002) Congenital renal agenesis: Case control analysis of birth characterstics. Am J Kidney Dis. 39(4), 689-694.

2. Robson WM, Leung AC, Rogers RC. Unilateral renal agenesis. (1995) Advances in Pediatrics. 42, 575-591.
3. Atala A. Congenital anomalies of the kidney. In: Schrier RW (ed). (2001) Diseases of the Kidney and Urinary Tract, 7th edition. Philadelphia, Lippincott William and Wilkins. 649-662.
4. Cascio S, Paran S, Puri P. (1999) Associated urological anomalies in children with unilateral renal agenesis. J Urol. 162, 1081.
5. Dursun H, Bayazit AK, Büyükçelik M. (2005) Associated anomalies in children with congenital solitary functioning kidney. Pediatr Surg Int. 21,456.
6. Kaneyama K, Yamataka A, Satake S. (2004) Associated urologic anomalies in children with solitary kidney. J Pediatr Surg. 39, 85.

7. Doroshou LW, Abeshouse BS. (1961) Congenital unilateral solitary kidney: Report of 37 cases and a review of the literature. Urol Surv. 11, 219-229.
8. Barakat AJ, Drougas JG. (1991) Occurrence of congenital abnormalities of the kidney and urinary tract in 13, 775 autopsies. Urology. 38, 347-350.
9. Thompson DP, Lynn HB. (1996) Genital anomalies associated with solitary kidney. Mayo Clin Proc, 41, 538-548.
10. Dees JE. (1960) Prognosis of the solitary kidney. J Urol. 83, 550-552.
11. Emanuel B, Nachman R, Aronson N, Weis H. (1974) Congenital solitary kidney. A review of 74 cases. Am J Dis Child. 127, 17-19.
12. Fitch N. (1977) Heterogeneity of bilateral renal agenesis. Canad Med Assoc J. 116: 381-382.
13. Schimke, RN, King CR. (1980) Hereditary urogenital adysplasia. Clin Genet. 18: 417-420.
14. Buchta RM, Visekul C, Gilbert EF. (1973) Familial bilateral renal agenesis and hereditary renal adysplasia. Z Kinderheilk. 115, 111-129.
15. Biedel CW, Pagon RA, Zapata JO(9184) Mullerian anomalies and renal agenesis: Autosomal dominant urogenital adysplasia. J Pediatr. 104, 861-864.

Fig. 1