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Unilateral Renal Agenesis with Congenital Solitary Functioning Kidney: A Case Report.

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ABSTRACT

Unilateral renal agenesis is a form of renal agenesis characterized by the complete absence of development of one kidney accompanied by an absent ureter. The annual incidence is estimated to be 1 in 1000 live births. The male to female ratio is around 1.2:1. Approximately 56% of agenesis occurs on the left side. Congenital unilateral renal agenesis occurs in 0.93-1.8 per 1000 autopsies, and is usually diagnosed during as incidental investigation. Genital anomalies occur in 37-60% of females and 12% of males with congenital unilateral renal agenesis. This case is a 76 years male during investigation for urinary tract infection was found to have Congenital agenesis of left kidney and Solitary functioning right kidney with hypertrophy. Marital history was normal. Not a known hypertensive .No relevant family history.

Keywords: Unilateral Renal agenesis, Ultrasound, Genital anomalies, kidney

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INTRODUCTION

Unilateral renal agenesis is a form of renal agenesis, characterized by the complete absence of development of one kidney accompanied by an absent ureter. The annual incidence is estimated to be around 1 in 1000 live births [1]. The male to female ratio is around 1.2 :1. Approximately 56% of agenesis occurs on the left side. The disease is usually clinically silent and is commonly detected as a chance observation. It presents during routine fetal ultrasound with an empty renal fossa, during renal ultrasound for the evaluation of a urinary tract infection, or in adults who present with secondary hypertension. Most patients are asymptomatic if the other kidney is fully functional. However hypertension, proteinuria and renal failure may develop in the long run in 20-50% of cases at the age of 30, which may be based on glomerular hyper filtration . Renal agenesis is occasionally associated with genital tract anomalies on the same side like seminal vesicle hypoplasia and absence of the vas deferens . Other organs may show anomalies up to 44% , mainly cardiac such as atrial or ventricular septal defect and gastrointestinal such as anal atresia . The risk of renal failure in childhood is minimal; however patients may develop hypertension, proteinuria and renal failure. Unilateral Renal Agenesis can occur with dysplasia or hypoplasia of the solitary functioning kidney (renal dysplasia and renal hypoplasia), which makes the prognosis more serious. It may be associated with X-linked kallman syndrome. Because of better availability of diagnostic modalities better detection of anomalies is possible.

CASE REPORT

76 years male came to the hospital with complaints of burning micturition with fever on and off for the past 6 months. No past history of similar episodes. No other complaints. Marital history was normal. Not a known diabetic or hypertensive. No relevant family history.

On examination – Patient was febrile Not an aemic , no cyanosis/ clubbing or pedal edema Vitals-Temperature-101 $\rm \dot{c}$

Pulse rate-102/min BP- 130/80 mmHg CVS-S1S2 +, No murmurs RS- NVBS +, No added sounds P/A- Soft, no tenderness

INVESTIGATIONS

Patient was advised to undergo ultrasound abdomen and was found to have congenital agenesis of left kidney. Solitary right kidney with hypertrophy.

DISCUSSION

Unilateral renal agenesis is usually diagnosed on an incidental imaging examination. The left kidney is more commonly involved than the right, and males are affected more than females. Congenital solitary kidney is compatible with longevity, but may be prone to disease such as pyelonephritis, obstruction and calculus formation [2,3].

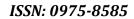
Currently, the exact cause of renal agenesis is not known. Renal agenesis is thought to be a "multifactorial" condition. The multiple factors like genetic and the environment are necessary for renal agenesis to occur. There have also been reports of families with hereditary renal agenesis (multiple family members have renal agenesis) [4].

Recognition of a congenital solitary kidney is important in order to monitor the affected individual for urinary infection, obstruction or calculi and warn the individual to avoid contact sports and similar activities that might endanger the solitary kidney. Physicians should be aware of the association of congenital unilateral renal agenesis with other anomalies of genital system. Early detection of a congenital solitary kidney by routine prenatal ultrasonography or incidental imaging should alert physicians to look for associated genital anomalies and avoid unnecessary procedures and surgery in patients presenting with abdominal and pelvic complaints [5].

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Prenatal diagnosis of the solitary kidney and other renal abnormalities by ultrasound is possible as early as 12 to 16 weeks of gestation [6]. Genital anomalies occur in 12% of males and 37–60% of females with congenital unilateral renal agenesis. Abnormalities in males include cryptorchidism, seminal vesicle cyst, hypoplastic vas, unilateral prostatic agenesis, cystic testicular dysplasia and hypospadias. Abnormalities in females include agenesis, duplication, rudimentary, unicornuate or bicornuate uterus, uterus didelphys (double uterus, double cervix and double vagina), double or absent vagina, absent or hypoplastic ovary, absent fallopian tube, persistent Gartner's duct cyst, and abnormal external genitalia [7].

The development of the urinary tract is a sequential and integrated process of the primitive renal elements. Abnormalities of this system result from defects occurring during embryogenesis between 15 and 94 days of fetal life. Interaction between environmental factors such as maternal illness and exposure to toxic agents, as well as genetic factors around this period result in malformations of this system [8].

Unilateral renal agenesis may be an expression of a single dominant gene [9]. McCallum et al [10] has postulated that unilateral renal agenesis and congenital bilateral absence of the vas deferens might have a non-cystic fibrosis mutation-mediated genetic basis that leads to abnormal development of the entire mesonephric duct before seven weeks of gestation. Solitary functional kidney is always hypertrophied ^[11] as in the present case.

CONCLUSION

Prenatal diagnosis of the solitary kidney and other renal abnormalities by ultrasound is possible as early as 12 to 16 weeks of gestation. Early detection of a congenital solitary kidney by routine prenatal ultrasound or by incidental imaging studies should alert physician to look for associated genital anomalies particularly in young females since one of every three with renal agenesis will also have a significant anomaly of the uterus, ovary or vagina . Such knowledge is useful to avoid unnecessary procedures and surgery in patients presenting with abdominal or pelvic complaints. Recognition of a congenital solitary kidney is also important in order to monitor the affected individual for urinary infection, obstruction or calculi and warn the individual to avoid contact sports and similar activities that might endanger the solitary kidney. Although unilateral renal agenesis is common, its definitive etiology is unknown. As the solitary kidney has to compensate for the absence of the other kidney, the patient with unilateral agenesis needs life long nephrological care. Unilateral renal agenesis is inherited as X- linked dominant trait and also associated with kallmann syndrome. In such conditions, the parents and the siblings need to be screened for renal agenesis, and proper counselling.

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