Iran J Radiol. 2017 May; Special issue(5):e48281. doi: 10.5812/iranjradiol.48281.

Published online 2017 April 13.

Abstract

Unilateral Enlargement of Kidney: A Rare Manifestation of Autosomal Recessive Polycystic Kidney Disease

Elham Zarei,[1] and Razieh Shahnazari[2,*]

[1]Assistant Professor

[2]Resident

[*] *Corresponding author*: Razieh Shahnazari, Resident. E-mail: eliday7@yahoo.com

Received 2016 December 21; **Accepted** 2017 February 08.

Abstract

Background: Autosomal recessive polycystic disease (ARPKD) is a heritable but phenotypically variable disorder characterized by varying degrees of nonobstructive renal collecting duct ectasia, hepatic biliary duct ectasia and fibrosis of both liver and kidney. The phenotypes of the ARPKD are distinguished by the age of the diagnosis at presentation and the predominance of renal over hepatic manifestations or vice versa. Sonography is currently the imaging technique of choice in children with renal cystic disease. Classic ARPKD is evident at birth with symmetrically enlarged kidneys with increased Parenchymal echogenicity at ultrasound with using a curved array transducer. In previous reports, nephromegaly is a constant imaging finding at the time of diagnosis. Asymmetrical enlargement has only been reported in one case report and the current case report is the second observation in this regard.

Case Presentation: A preterm neonate with suspicious RT renal mass and oligohydramnios at prenatal ultrasound was hospitalized due to respiratory distress and hypertension. Although postnatal ultrasound showed enlarged echogenic Rt kidney and normal sized echogenic Lt Kidney, regarding to linear array transducer findings, ARPKD was considered rather than renal mass. The diagnosis was confirmed after nephrectomy and histologic examination.

Conclusions: Because the abnormality in ARPKD affects the renal tubules, high resolution sonographic techniques with linear array transducer are well suited to the imaging of this disease. Although, the patient showed asymmetric renal enlargement, unique findings on linear array transducer help to diagnosis of ARPKD regardless of renal size. Also, the finding of asymmetric renal enlargement in this patient adds another radiologic finding that could be associated with ARPKD.

This is an abstract presented in the 33rd Iranian congress of radiology (ICR) and the 15th congress of Iranian radiographic science association (IRSA).