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Autosomal recessive polycystic kidney disease: a case report

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Abstract:

Background: Autosomal recessive polycystic kidney disease is characterized by renal collecting duct cysts, congenital hepatic fibrosis (biliary dysgenesis) and autosomal recessive pattern of inheritance. The disease usually manifests in infancy, and has a high mortality rate in the first year of life. Kidney involvement is always present and manifests as variable degrees of non-obstructive collecting duct ectasia, usually in bilateral and symmetrical fashion and interstitial fibrosis. Patients almost always have also hepatic involvement as congenital hepatic fibrosis (periportal fibrosis with anomaly and dilatation in intrahepatic biliary ducts) that can cause portal hypertension with esophageal varices, bleeding and hypersplenism. The relative degrees of kidney and liver involvement tend to be inverse: Children with severe renal disease usually have milder hepatic disease and vice versa. Case report: This paper presents a 23-month old female with progressive renal failure and hepatic involvement (portal hypertension, esophageal varices bleeding and hypersplenism), which has been reported rarely.

Keywords:

Polycystic kidney disease ، Autosomal recessive ، Hepatic involvement ، Biliary dysgenesis

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