Congenital Hepatic Fibrosis: An Uncommon Cause of Chronic Renal Failure

Azarfar A1, kiani MA2, Keykhosravi Ag3, Ravanshad Y4

1Assistant professor of Pediatric Nephrology, Mashhad University of Medical Sciences, Mashhad, Iran.
2Associate professor of Pediatric Gastroenterology, Mashhad University of Medical Sciences, Mashhad, Iran.
3Associate professor of Pediatric Nephrology, Mashhad University of Medical Sciences, Mashhad, Iran.
4Mashhad University of Medical Sciences, Mashhad, Iran.

Abstract:
Congenital Hepatic Fibrosis (CHF) is a rare disease that affects both the liver and kidneys. Congenital hepatic fibrosis (CHF) is an autosomal recessive inherited malformation defined pathologically by a variable degree of periportal fibrosis and irregularly shaped proliferating bile ducts. Affected individuals also have impaired renal function, usually caused, in children and teenagers, by an autosomal recessive polycystic kidney disease (ARPKD). Impaired renal function associated with CHF in adults is caused by an autosomal dominant polycystic kidney disease (ADPKD).

Case presentation:
We report the case of a 8-year-old Iranian girl was admitted to our hospital for evaluation of renal failure. In patient hepatomegaly was noted incidentally on a routine physical examination and then kidney biopsy showed global sclerosis and A liver biopsy revealed proliferation of collagen fibres surrounding the portal area, a finding that was compatible with congenital hepatic fibrosis and our patient was scheduled for kidney and liver transplantation.

Conclusion:
The relationship of ARPKD to CHF is the subject of substantial controversy. Some clinicians suggest that the two conditions represent one disorder with a range of clinical/pathological presentations.

Key word: Congenital Hepatic Fibrosis Polycystic Kidney Disease, CRF.

*Corresponding Author:
Anoush Azarfar, MD, Assistant professor of Pediatric Nephrology, Mashhad University of Medical Sciences(MUMS), Mashhad, Iran. Email: Azarfar@iums.ac.ir