Congenital Hepatic Fibrosis: An Uncommon Cause of Chronic Renal Failure

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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 girlwas 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 fibrosisand 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.

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