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Congenital Hepatic Fibrosis- A Case Report

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Abstract

CHF is one of the "fibropolycystic diseases" and is a rare disease of children and young adults. Patients usually present with signs of portal hypertension with frequent renal involvement. There is relative preservation of liver function and underlying architecture. Major complaints are due to hepatosplenomegaly and portal hypertension. We present a case of 16 year old male patient who presented with pain in abdomen, splenomegaly and signs of portal hypertension. LFTs were mildly deranged with pancytopenia. CT scan revealed moderate splenomegaly, mild hepatomegaly with renal cortical multiple bilateral cysts. Splenectomy was performed with wedge liver biopsy. Histopathological examination revealed portal tract widening with ductular proliferation along with fibrocongestive splenomegaly. Based on the clinical, radiological and histopathological findings, diagnosis of CHF with fibrocongestive splenomegaly was made.