Case Report

Congenital hepatic fibrosis – Two cases

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Abstract

Congenital hepatic fibrosis is an exceedingly rare disease where only very few cases with sufficient evidences and clinical data have been reported up to now. Here we reported a patient onset of hematemesis, with normal liver function. Computer tomography scans showed splenomegaly, intra-hepatic bile ducts dilation, thickening portal vein and tortuous spleen vein. Liver biopsy found significant fibrosis in the portal area and ectasia of bile ductules. With sufficient radiologic and pathologic data, our case revealed the features of congenital hepatic fibrosis associated.

Keywords: Congenital hepatic fibrosis ectasia, hematemesis, Radiology; Pathology.

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INTRODUCTION

Congenital hepatic fibrosis is a developmental disorder that belongs to the family of hepatic ductal plate malformations and is characterized histologically by a variable degree of periportal fibrosis and irregularly shaped proliferating bile ducts[1]. It is an important cause of portal hypertension in the infantile and juvenile age groups. In most patients, the first manifestations of the disease are signs or symptoms related to portal hypertension—especially splenomegaly and varices—often with spontaneous gastrointestinal bleeding[2]. The clinical manifestation of congenital hepatic fibrosis is, however, non specific, which makes the diagnosis of this disorder extremely difficult. However liver biopsy is highly specific for the diagnosis of congenital hepatic fibrosis. Because of the difficulty in the clinical and pathologic diagnosis of congenital hepatic fibrosis, imaging studies could play a crucial role in the diagnosis of this disorder if accurate and reliable signs can be determined. Unfortunately, little is known about the radiologic diagnosis of congenital hepatic fibrosis. Delphine Zeitoun et al described the CT findings in eighteen patients with congenital hepatic fibrosis[3]

CASES REPORT

CASE I: A 8 yr old female patient came to paediatric department of Mahatma Gandhi medical college and hospital Jaipur, Rajasthan with complain of recurrent hematemesis 10-12 episodes in last 2 yr in every 2 to 3weeks,non projectile non bilious perfuse in amount, contain blood clots and approx 100 -200 ml blood in every vomit. She had no history of previous hepatitis. And no record of hereditary liver disease was found in her family members. Physical examination showed abdominal wall varies, splenomegaly. Blood sample test showed pancytopenia, which indicated the existence of hypersplenism. LFT was normal in range. Hepatitis B surface antigen or anti-hepatitis C virus antibody are negative. Immunology test showed normal levels of serum ferrum and ceruloplasmin. Autoimmune antibodies associated with liver disease, such as anti-nuclear antibody, smooth muscle antibody, liver-kidney cytoplasm antibody etc, are all negative. On gastroduodenoscopy oesophageal varies grade II are noted.

Radiology investigation

USG whole abdomen showed coarse, heterogenous echotexture of liver parenchyma with periportal fibrosis. There were bilateral mild dilated intra hepatic biliary radicals. Liver is reduced in size. Portal vein was normal in size and showed normal colour flow and waveform. Spleen is enlarged in size measured13.56 cm and coarse echotexture. splenic vein was normal in diameter. There were multiple collaterals are seen at splenic hilum.

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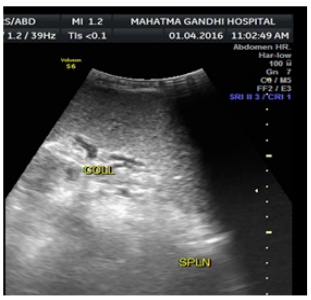
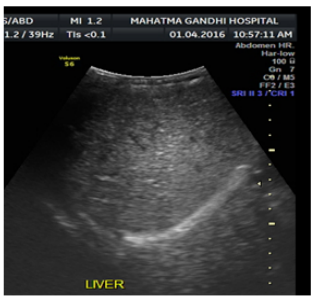


Figure 1



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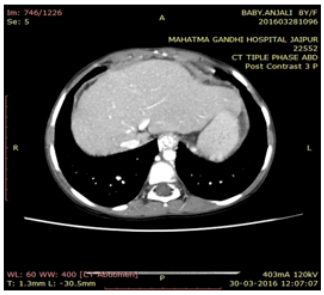

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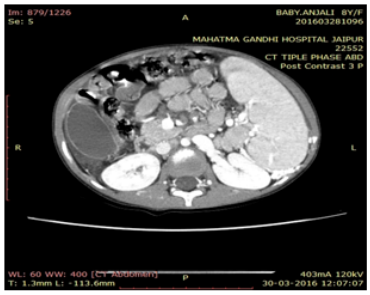
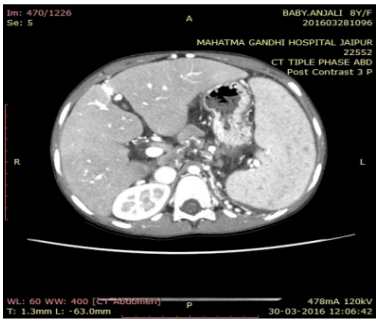


Figure 4



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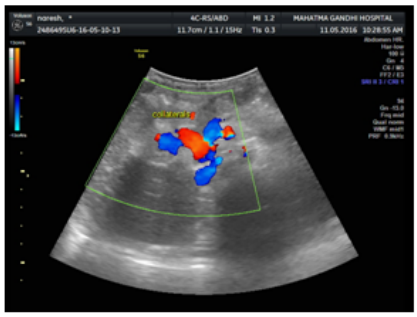

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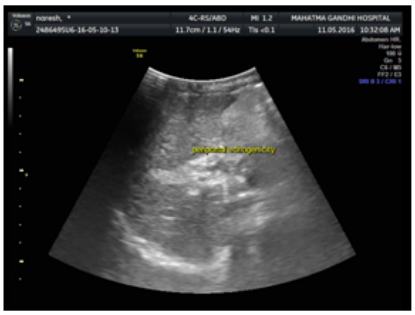

Figure 7

Legend

Figure 1: USG whole abdomen showing spleenomegaly with multiple collaterals and coarse echotexture of hepatic parenchyma
Figure 2: Cect abdomen showing
Figure 3: showing esophageal varieces.
Figure 4: Showing spleenomegaly, multiple hypoattenuated foci in entire spleenic parenchyma, multiple colletrals at spleenic hilum, spleenorenal
Figure 5: Cect abdomen showing recalanization of umblicalvein
Figure 6: USG whole abdomen showing multiple collaterals at splenic hilum
Figure 7: Periportal echogenicity

Abdominal triple phase contrast enhanced CT Scan showed Moderate ascites, splenomegaly. There were multiple small foci of hypoattenuation with no contrast enhancement were seen in entire splenic parenchyma. splenic vein normal in calibre. Multiple enlarged lymphnodes were seen in splenic hilum, largest measuring 10 mm in short axis. Multiple portosystemic collaterals were seen around GE junction with esophageal varices, perisplenic, perigastric, splenorenal, and periumbilical region with recanalized umbilical vein. Liver showed mild atrophic changes. IHBR were mildly dilated in both lobes of liver. Possibility of sequelae of congenital hepatic fibrosis to be considered.

On liver biopsy showed loss of architecture due to formation of nodules. The portal tracts showed severe fibrosis with formation of incomplete and complete nodules. There was proliferation of periseptal bile ductules and presence of bile pigments was seen in few ducts. Moderate degree of piecemal necrosis was seen around most of portal tracts. Features in suggestive of cirrhotic changes.

CASE II

A4 yr old male patient came to paediatric department of Mahatma Gandhi medical college and hospital Jaipur, Rajasthan with complain of recurrent hematemesis 5-10episodes in last 3yr. he had no history of previous hepatitis. no record of hereditary liver disease was found in his family members. Physical examination showed abdominal wall varies, splenomegaly. Blood sample test showed pancytopenia, which indicated the existence of hypersplenism. LFT was normal in range.

RADIOLOGICAL FINDINGS

USG whole abdomen showed altered, heterogenous echotexture of liver parenchyma with periportal fibrosis. Liver is reduced in size. Portal vein was 6mm in size.gall bladder was contracted with thickened wall. there are multiple peri gall bladder venous collaterals noted. Spleen is enlarged in size measured10cm. There were multiple collaterals are seen at splenic hilum. Abdominal triple phase contrast enhanced CT Scan showed splenomegaly. Multiple portosystemic collaterals were seen around GE junction with esophageal varices, perisplenic, perigastric, splenorenal, and periumbilical region. Liver showed mild atrophic changes. portal vein measures 5 mm with

periportal fibrosis. Possibility of sequelae of congenital hepatic fibrosis to be considered. On liver biopsy showed loss of architecture due to formation of nodules with severe portal fibrosis. There was proliferation of periseptal bile ductules and presence of bile pigments was seen in few ducts. Moderate degree of piecemal necrosis was seen around most of portal tracts. Features in suggestive of hepatic fibrosis changes.

DISCUSSION

Congenital hepatic fibrosis is a rare autosomal recessive disease named by Kerr in 1961[4]. Congenital hepatic fibrosis is a dynamic disorder that shows progression in the extent of liver fibrosis over time[5], with evolution into true cirrhosis of the liver.Okuda[6] recently stated, "no specific imaging feature is known" for the diagnosis of congenital hepatic fibrosis. Most patients represent as hepatosplenomegaly, hematemesis or hematochezia. Clinically, liver function is mildly damaged or normal because few hepatocytes are involved. Some distinct CT features were frequent in congenital hepatic fibrosis, such as hepatomegaly, varices, splenomegaly, associated ductal plate malformations, and renal abnormalities. The combination of these CT signs is very important for the diagnosis of congenital hepatic fibrosis. Pathologic changes include massive hyperplasia of fibrous connective tissue in portal area, hyperplasia of bile ductules, and/or cholangiectasis.

CONCLUSIONS

In these cases, we established the diagnosis on the basis of evidences as below: 1) Juvenile onset and variceal bleeding;
2) Normal liver function unparalleled with the severity of portal hypertension; 3) Intra-hepatic bile duct dilation, splenomegaly 4) Histopathologically, hyperplasia of fibrous connective tissues and bile ductules in portal area, 5) Excluded diagnosis of hepatitis, alcohol abuse, metabolic and hereditary liver disease, and autoimmune liver disease.

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