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Caroli's Disease- Radiological Evaluation Of Rare Biliary Ductal Plate Malformation In A Child In A Tertiary Care Hospital In Tripura

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ABSTRACT

C*aroli's disease, a rare congenital condition, among the ductal plate malformations characterized by non-obstructive saccular or fusiform dilatation of larger intrahepatic bile ducts that occur at different levels in the developing biliary tree. The disease is also characterized by the formation of intraductal calculi and susceptibility to infection. It as an autosomal recessive disease which is often associated with congenital hepatic fibrosis & autosomal recessive polycystic kidney disease.Males and females are equally affected .More than 80% of patients present before 30 years of ages. Imaging modalities like Ultrasonography, Magnetic resonanace cholangiopancreatiography play very crucial role in detecting Early diagnosis of testicular cancer leads to proper management and improve the outcome by preservation of future fertility. Whereas ultrasonography (USG) helps in early detection of dialatation of intrahepatic biliary radicles, advanced diagnostic modality such MRCPconfirms the diagnosis of Caroli's disease as well as can predict the prognosis of this diasease.We report one case of Caroli's disease in a 6 year old female patient who presented with unusual presentation of hematemesis. Very few cases of Caroli's disease in child in Tripura have been reported in the literature till date which makes our case a rare entity.*

Key Words: *Caroli disease, USG (Ultrasonography), CT scan, MRCP (Magnetic resonance cholangiopancreatography)*

INTRODUCTION

Caroli's disease (CD), also known as communicating cavernous ectasia or congenital saccular dilatation of the intrahepatic bile ducts is a rare congenital disorder first specifically described in 1958.[1] It corresponds to type V choledochal cyst as classified by Todani et al[17]. Although mode of inheritance is still not clear but in majority of cases it is transmitted in autosomal recessive fashion[2]. The incidence of Caroli's disease is about 1 in 1,000,000 live births[3]. Caroli's disease is now considered as a genetic disorder involving the *PKHD1* gene (polycystic kidney and hepatic disease 1), which affects a protein called fibrocystin. (expressed in multiple organ systems: the renal tubular cells, liver cholangiocytes, and the pancreas). Genetic abnormalities in this protein result in fibrocystic changes in the kidney and liver. Thus it is seen that, Caroli's disease is frequently seen with autosomal recessive polycystic kidney disease (ARPKD)[4]. No gender predominance has been observed[5]. Caroli's disease may be diffuse or may be localized to a lobe or a segment of the liver, usually the left.The disease is usually seen in its two forms-simple (classic); in which the intrahepatic biliary duct dilatation is seen without hepatic fibrosis and Caroli's syndrome (fibrous); in which there is hepatic fibrosis in addition that may be accompanied by the cascade of events that follow portal hypertension. Early recognition of the disease and itscomplications is required for timely management of the patient[6].

CASE REPPORT

A 6 years old female patient presented to the Pediatrics emergency department of tertiary care of Tripura with complaints of multiple episodes of Hematemesis & mild pain abdomen for last 1-2 days. There was similar kind of history (2 episodes of Hematemesis) 3 Months back. Signs of pallor were noted on general examination. On per abdominal examination, Hepatosplenomegaly was present.Examinations of all other systems were normal. In laboratory investigation, patient was found to be anaemic(Hb% was 7.3 mg%). TLC was little bit on higher side(15000/mm^3).Then radiological Investigations were advised for this patient. An initial USG scan was done followed by MRCP & CECT whole abdomen.

On gray scale ultrasonography gross secular dilatation of intrahepatic bile ducts were seen in the both lobes of liver. Common bile duct was normal. No evidence of any calculus or mass lesion seen in the common bile duct. On color Doppler evaluation, central vascularity was noted in few of the dilated intrahepatic bile ducts.Hepatosplenomegaly along with minimal anechoic free fluid collection were also noted.No obvious renal cyst was found. Based on ultrasonography findings provisional diagnosis was given as caroli disease with hepatosplenomegaly & mild ascites. To confirm the diagnosis MRCP followed by CECT was done.

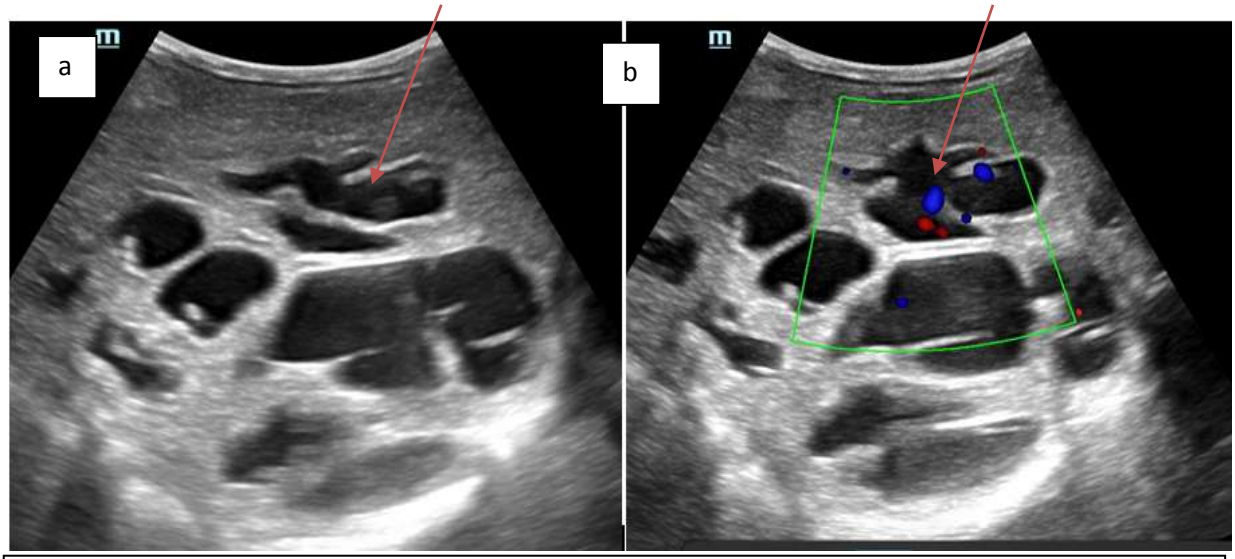

Figure 1: (a) Sagittal section of right lobe of liver showing IHBR dilatation (Gray scale USG),

(b) Axial section of scrotum showing internal vascularity inside dilated IHBR (Colour Doppler study).

On CECT whole abdomen prominent non obstructive secular dilatation of the intrahepatic bile ducts seen along with central in dot sign(central intensely enhancing "dot" surrounded by dilated IHBR) hepatosplenomeagly was also noted along withMinimal ascites. MRCP confirmed the non marked obstructive saccular dilatation of intrahepatic bile ducts.

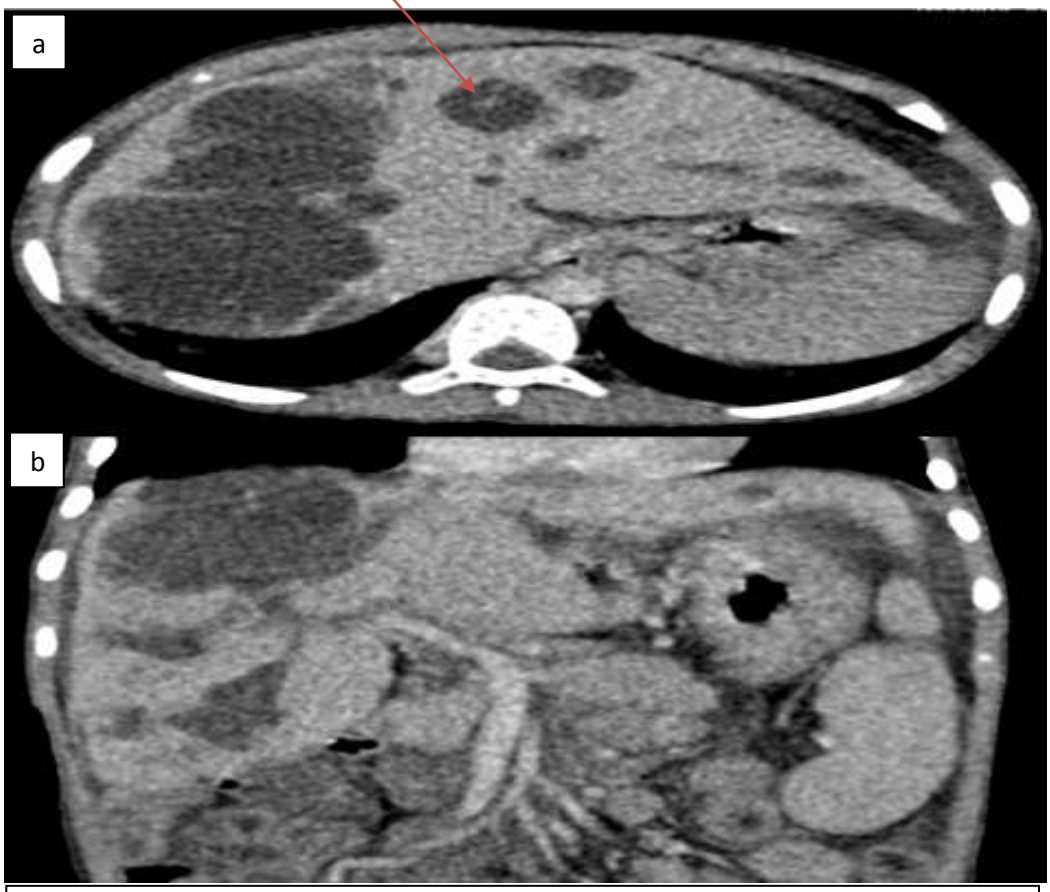

Figure 2:a) Axial section of liver showing grossly dilated IHBR with "Central in dot sign"- red arrow (CECT of abdomen)

b) Coronal section showing dilated IHBR with grossly dilated with hepatomegaly (CECT of Abdomen)

For confirmation of MRCP was performed. In T2 weighted images non obstructive gross dilatation of intrahepatic biliary radicles was noted with Hepatosplenomegaly. Central in dot sign was also noted which confirmed the diagnosis of Carolis disease.

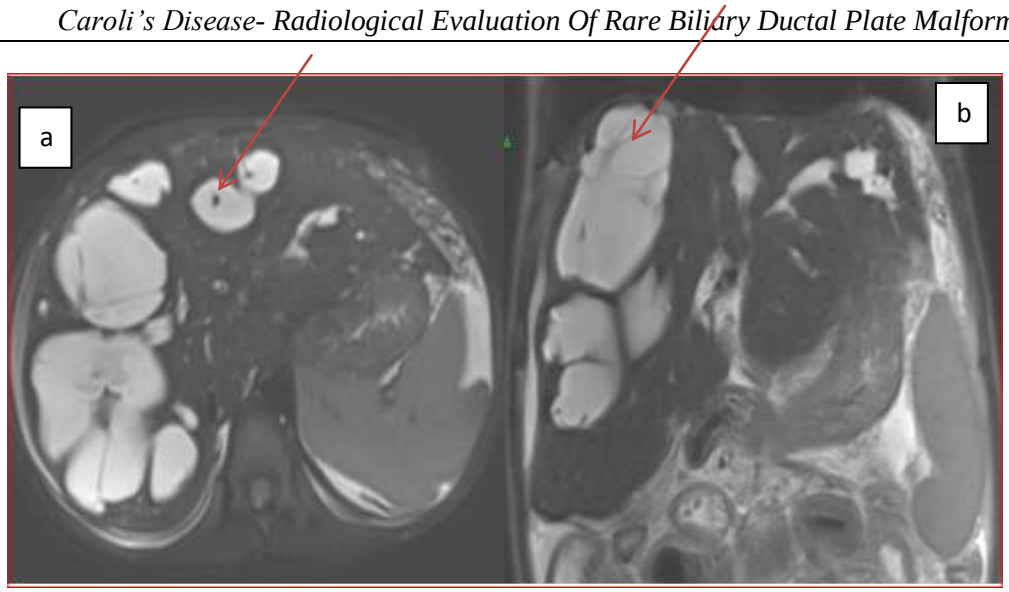

Figure 3: a) Axial section- showing gross saccular dilatation of intrahepatic biliary radicles (MRCP ; T2W FS image) with "central in dot" sign

b) Coronal section- showing gross saccular dilatation of intrahepatic biliary radicles with Hepatosplenomegaly (MRCP ; T2W image)

Based on the clinical history and radiological investigations diagnosis was made as Caroli disease . Initially the patient was managed conservatively and 2 units of Packed RBC were infused after initial hematological investigations & Patient was stabilised. Patient party was counselled regarding the treatment options of this rare congenital condition of the patient. Then the patient was discharged and advised for follow up.

DISCUSSION

Caroli's disease was first described by Caroli et al., that is characterized by non-obstructive segmental cystic dilatation of the intrahepatic bile ducts[7]. Caroli's disease is extremely rare with prevalence of one in a million approximately with a female preponderance[6] .Two types of CD were later recognized. Type I, or simple CD, consists of pure cystic dilatations of the intrahepatic bile ducts, whereas type II, or complex CD, also known as Caroli's syndrome (i.e. CD with congenital hepatic fibrosis), is associated with hepatic fibrosis, or even cirrhosis, portal hypertension and oesophageal varices[16].Caroli's syndrome is more common than Caroli's disease. Most of the patients with Caroli's disease present with recurrent episodes of cholangitis, cholelithiasis, biliary abscess, and septicemia[9].In Caroli's disease first onset of the symptoms mostly occurs in adolescence or early adulthood[8] .Because hepatic resection can be curative in Caroli's disease, early diagnosis is essential to prevent complications, which include calculusformation, cholangitis, abscess, and cholangiocarcinoma[10].The laboratory findings may show leukocytosis in cholangitis(same as in our case).Imaging modalities plays crucial role in planning the surgical treatment, that consist of enterostomy, segmental or lobar hepatic resection, or liver transplantation[11]. Carbohydrate antigen 19- 9 and carcinoembryonic antigen are done for screening of cholangiocarcinoma[12].Many imaging modalities can be used for the evaluation of Caroli's disease in different clinical scenarios, which include Ultrasound (US), Computed Tomography (CT) , Magnetic Resonance Cholangiopancreatography (MRCP), Endoscopic retrograde cholangiopancreatography, radionuclide hepatobiliary imaging and intraoperative cholangiography. Among the imaging modalities ultrasonography remains the first line of investigation in Caroli's spectrum[13]. Although Ultrasonography and CT are commonly used to investigate patients with symptoms suggesting biliary disease, direct cholangiography is often necessary for an accurate diagnosis. As direct cholangiography (ERCP & PTC) carries higher risk of cholangitis noninvasive method likeMagnetic ResonanceCholangiopancreatography (MRCP) is widely used for confirmation of diagnosis of Caroli's disease.Advantages of MRCP consistof depiction of the entire biliary tree, a low complication rate, and a spatial resolution as high as 1.5 mm[14].The CT or MRI showsthe "central dot sign"- (representing portalradicles) [Figure 2] within the intrahepatic ducts[15].Magnetic resonance imaging (MRI) has excellent soft tissue resolution, multidirectional and multisequence scanning technology, making it a important supplementary method in the confirming the diagnosiss of Caroli's disease.

CONCLUSION

Caroli's disease is a rare inherited disorder that may cause severe, life-threatening cholangitis or hepatobiliary degeneration, or even carcinoma. Caroli's disease should not be forgotten in the differential diagnosis of recurrent cholangitis especially in children or young adults.Laboratory findings cannot be relied upon for diagnosis. Diagnostic imaging has an important role in the early diagnosis of caroli disease with several different imaging modalities employed over the spectrum of care in these patients including initial diagnosis and and long term surveillance.Among the imaging modalities most useful in the diagnosis of CD are MRCP and CT.

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