



Focal Caroli's Disease Presenting as Fusiform Dilatation of Intrahepatic Biliary Radicles

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Authors' contributions

This work was carried out in collaboration between all authors. Author SA drafted the article and wrote the first draft of the manuscript. Author RK managed the literature searches. Author SK helped the author RK in literature search and author RG revised the article for important intellectual content. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Background Caroli's disease is a rare congenital disorder characterized by segmental, non obstructive, cystic dilatation of intra hepatic bile ducts. It belongs to the spectrum of fibropolycystic liver disease which results from in utero malformation of ductal plate. Two forms of this disorder have been described, the less common 'Pure' form involves only the large intrahepatic bile ducts and the more 'complex' form is associated with congenital hepatic fibrosis (CHF), and is known as Caroli's Syndrome. Case A young female presented with features of cholangitis and in addition to all the routine investigations, non invasive imaging in the form of ultrasonography, computed tomography and magnetic resonance imaging was done. Diagnosis Final diagnosis of focal Caroli's disease with cholangitis was made on clinical and imaging findings. Treatment Conservative treatment was given and patient referred to gastro surgery department for partial hepatectomy. Conclusion An early recognition of Caroli's disease with non invasive imaging can bring down the morbidity.

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1. CASE

A 29 yrs non diabetic normotensive female was admitted in Medicine department of Guru Gobind Singh Medical College and Hospital, Faridkot (Punjab), India with pain right hypochondrium, jaundice, loss of appetite for 8 months, and fever for 15-20 days which was high grade with chills and rigors. There was no history of weight loss, anorexia, vomiting, bleeding from any site, rash, tremors or altered sensorium and no previous h/o surgery, tattooing or blood transfusion. The patient has been having similar complaints repeatedly for about 8 months for which she used to take medication from a general practitioner. There was no history of liver, kidney or any other significant disease in the family. On examination patient had deep icterus with nontender hepatomegaly and mild splenomegaly. Rest of the clinical examination was unremarkable. Investigations revealed total S. bilirubin of 23.5 mg/dl (conjugated 0.58, unconjugated 13.6), AST-169IU/L(N 5-40), ALT-126IU/L(N 5-30), Alkaline phosphatase 912IU/L(N 60-150), INR- 1.2, TLC 20.7×10^3 /ul with 80% polymorphs. Renal function tests, blood sugar, electrolytes and urine examination were within normal limits. Hepatitis B surface antigen, Anti-Hepatitis C Virus (ELISA) & Human Immunodeficiency Virus (ELISA) were non reactive. Anti smooth muscle antibodies were negative. Ultrasonography of abdomen revealed hepatomegaly with normal liver echo pattern. Multiple cystic space occupying lesions with dilated biliary radicles in the left lobe were seen. There was dilatation of left and right hepatic ducts with normal CHD and CBD. The cysts showed internal echoes with intraluminal protrusions, and few cysts appeared to be communicating with the dilated biliary radicles. Gallbladder was contracted. There were few enlarged peri pancreatic and portal lymphnodes with mild splenomegaly with normal porto-splenic axis. Kidneys were unremarkable. On color doppler, portal vein branches were accompanying the cysts. Contrast enhanced CT abdomen showed oblong cystic lesions in the left lobe of liver with high attenuation, largest measuring 5.6x3.8 cm in segment II with mild dilatation of intrahepatic biliary radicles (IHBR) (Figs. 1 & 2). Vessels were seen at the periphery of the cysts with a central dot sign. MRI & magnetic resonance cholangio pancreatography (MRCP) revealed multiple fusiform dilatations of left intrahepatic biliary radicles (Figs. 3,4,5).

Upper Gastrointestinal endoscopy was normal and ERCP was not done. Based on the above clinical & imaging findings. A final diagnosis of focal Carolis disease with cholangitis was made and the patient was advised to conduct partial hepatectomy and referred to gastro surgery department. However, patient did not give consent for surgery and remained on conservative treatment.

2. DISCUSSION

Carolis disease also known as communicating cavernous ectasia of the intrahepatic ducts, is an autosomal recessive disorder and is among the ductal plate malformations that occur at different levels in the developing biliary tree, leading to several clinicopathologic entities [1]. There is arrest of or a derangement in normal embryological remodeling of intrahepatic bile ducts which causes varying degree of destructive inflammation and segmental dilatations with periductal fibrosis [2]. Two forms of the disease have been described. Type I also known as the pure or non hereditary form is often limited to one hepatic lobe usually left lobe with multifocal saccular dilatations of bile ducts. There is marked predisposition to cholangitis and liver abscesses in this form. Type II or the hereditary form is autosomal recessive trait involving central and peripheral intrahepatic ducts associated with congenital hepatic fibrosis and portal hypertension and is known as Caroli's syndrome [3,4]. According to Todani classification of choledochal cysts, Caroli's disease is the type V choledochal cyst variety [5], although the pathogenesis of Caroli's disease (autosomal recessive and often associated with renal disorders) and choledochal cysts (congenital and not associated with renal disorders) being different, make it unlikely that these entities are related to each other [2,4]. Other associations include autosomal recessive Polycystic kidney disease and Neurofibromatosis I. The incidence of Carolis disease is 1 in 1,000,000 with female preponderance. Clinically, patient presents with pain abdomen due to recurrent cholangitis, fever, tenderness in right upper quadrant and more rarely jaundice. Our patient also presented with cholangitis and jaundice. On imaging, the disease may be diffuse, lobar or segmental with more commonly involvement of left lobe. The dilatation of intrahepatic biliary radicles can be saccular (76%) or fusiform type (24%) [2]. Ultrasonography shows cystic spaces in liver

which communicate with the bile ducts. Intraluminal bulbar protrusions of the wall may be seen causing irregular dilatations of the radicles. Our case also showed cystic spaces with intraluminal bulbar protrusions. CT scan shows multiple hypodense saccular areas inseparable from dilated biliary ducts with central dot sign [3,6,7]. The central dot corresponds to fibrovascular bundles which contain a portal vein radicle and accompanying hepatic artery branch surrounded by the dilated bile duct. This sign is considered highly specific for Carolis disease and the present case also showed the central dot sign. MR imaging provides information about the severity, location and extent of liver involvement [8]. MRCP demonstrates the continuity of cysts with the biliary tree which is considered the hallmark of the disease. Guy et al reported nine patients of Carolis disease who underwent MRCP along with contrast enhanced magnetic resonance (CEMR) and concluded that MRI can be used as the only diagnostic modality for

evaluation of such patients [9]. Multiple fusiform cysts communicating with the left IHBR were seen in this case also. Hence, invasive investigations like ERCP and PTC are usually not required as they carry high risk of bacterial cholangitis [6,8].

Differential diagnosis includes primary sclerosing cholangitis (PSC), recurrent pyogenic cholangitis (RPC), biliary hamartomas, polycystic liver disease and choledochal cyst [10]. In PSC there is associated atrophy of liver and inflammatory bowel disease in 50% of patients. The ductal dilatations are fusiform rather than saccular, diffusely involving the liver in PSC and RPC which were also seen in our patient, however the liver involvement was focal. In polycystic liver disease and biliary hamartomas the cysts do not communicate with the bile ducts which can be well demonstrated on MRCP with normal appearance of intra and extrahepatic ducts [11].

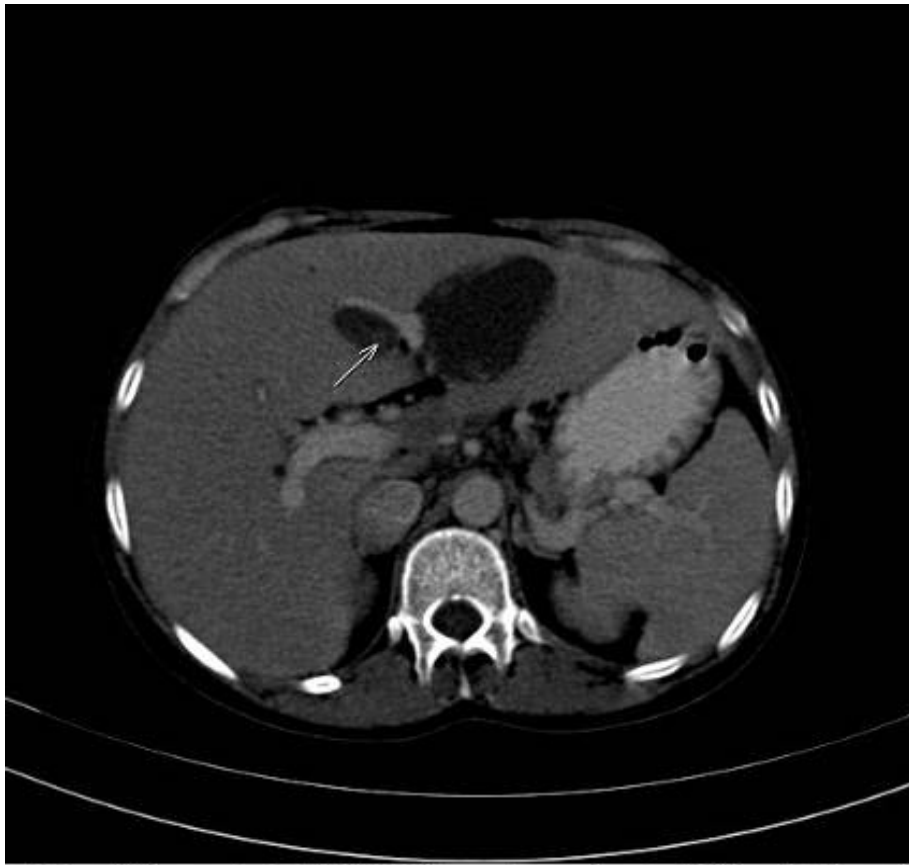


Fig. 1. CECT scan axial section of liver showing dilated IHBR of left lobe with a central dot sign and a cyst communicating with IHBR



Fig. 2. Coronal reconstructed CECT image showing fusiform dilatation of IHBR (long arrow) of left lobe with a communicating cyst (arrow). Portal vein radicle is seen at the periphery of the cyst (arrow head)

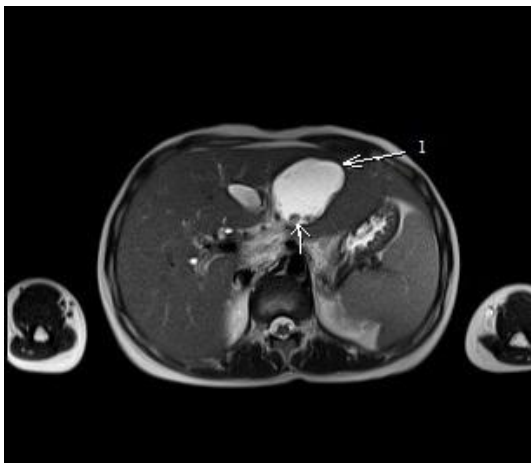


Fig. 3. T2 haste axial MR image showing dilated biliary radicles with a cyst in the left lobe with hypo intensities at the periphery of the cyst. Right lobe shows normal biliary ducts

Potential complications include cholangitis, cholelithiasis, biliary abscess, septicemia, liver cirrhosis, and cholangiocarcinoma (up to 7-24%) [3,5]. Treatment depends upon the disease

pattern, with segmentectomy or lobectomy as the preferred modality in localized form and orthotopic liver transplant in the diffuse form. Focal rather than diffuse liver involvement and fusiform instead of saccular dilations make this case different from the usual presentation.

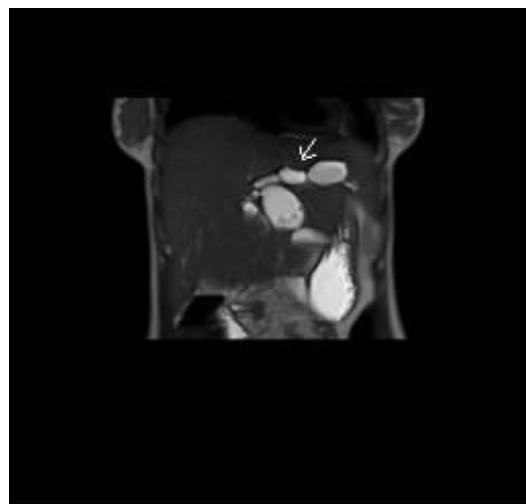


Fig. 4. T2 haste coronal MR image showing fusiform dilatations of IHBR of left lobe

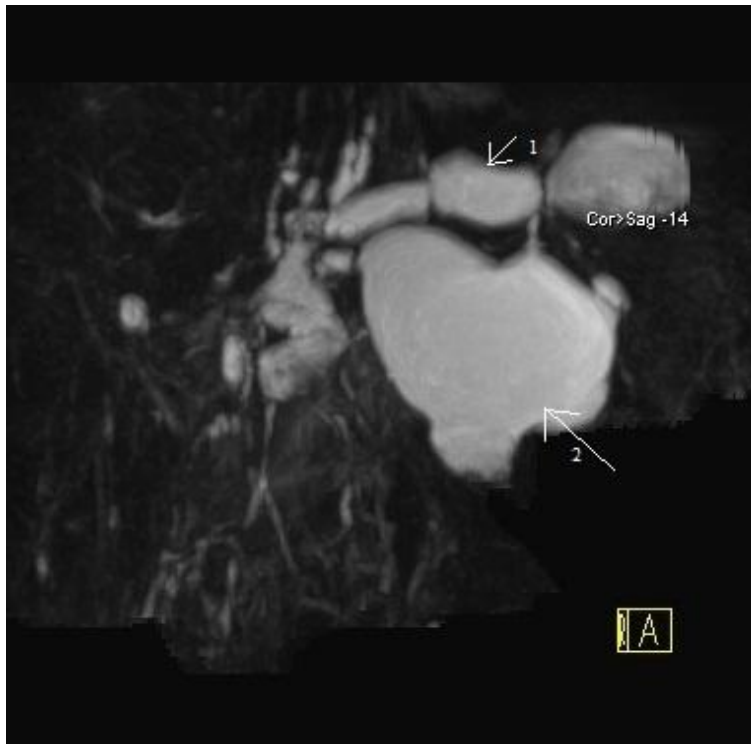


Fig. 5. MRCP showing dilated ducts of the left lobe with communicating cysts

3. CONCLUSION

In patients presenting with cholangitis, an early recognition of Caroli's disease with non-invasive imaging modalities, can bring down the morbidity associated with the disease.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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