

A Middle Aged Man with Caroli's Disease: A Case Report

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Abstract:

Caroli's disease (CD) is a rare congenital abnormality characterized by non-obstructive dilatation of intra hepatic bile ducts, which may be complicated by stone formation, recurrent cholangitis, biliary abscess and higher risk for biliary malignancy. We report a 37-year-old man with recurrent bouts of upper abdominal pain, high grade pyrexia, mild icterus with normal liver function tests who was diagnosed as a case of Caroli's disease. The laboratory studies confirmed Caroli's disease with a SOL in liver suggestive of liver abscess and the patient received broad spectrum antibiotics with anaerobic and amoebicidal coverage. With 14 days course of antibiotics, he gradually recovered from his symptoms.

Key Words: Caroli's disease, Liver abscess, Cholangiocarcinoma, ERCP.

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Introduction:

Caroli's disease and caroli's syndrome belong to a group of fibropolycystic diseases^{1,2}. These are rare congenital disorders of the intrahepatic bile ducts. Jacques Caroli, a French gastroenterologist first described it as "non-obstructive saccular or fusiform multifocal segmental dilatation of intrahepatic bile ducts"³. The term caroli's disease is applied if the disease is limited to ectasia or segmental dilatation of the larger intra-hepatic ducts. In caroli's syndrome, malformations of small bile ducts, portal hypertesion and congenital hepatic fibrosis are also present⁴. This process can be either diffuse or segmental and may be limited to one lobe of the liver, more commonly the left lobe. Simple Caroli's disease is an autosomal dominant trait, whereas the more complex Caroli's syndrome is generally inherited in an autosomal recessive manner⁴. Females are more prone to have Caroli's disease than males³.

Case Report:

A 37-year-old, normotensive, non-daiabetic businessman admitted in Medicine department of BSMMU in October 2011 who presented with high grade intermittent fever with colicky non radiating right hypochondriac and

epigastric pain with progressive swelling of his legs for last 15 days without satisfactory response to 7 days course of oral ciprofloxacin. He had previous history of recurrent cholangitis with several hospital admission for it since 1996 with 3 operations (endoscopic removal of CBD stone with papillotomy, Cholecystectomy, Hepatico-jejunostomy) and he received 2 units of blood transfusion with history of multiple unsafe sexual exposure.

On general examination he is emaciated, moderately anaemic with digital clubbing, mildly icteric, edematous without any lymphadenopathy or bony tenderness. Per abdominal examination revealed mildly distended, tender epigastrium and right hypochondrium with a right subcostal incision mark and just palpable liver but no other organomegally or ascites. Laboratory investigations showed severe anemia (Hb-6.2 gm/dl), polymorphonuclear leukocytosis with very high ESR(140 mm in 1st hour). Liver function tests revealed SGPT: 16 U/L, SGOT: 49 U/L, S. Bilirubin: 8 µmol/L, Prothrombin time: 15.0 Sec, Alkaline phosphatase: 156 U/L, tumour markers (CA 19-9) & viral markers were within normal limit. Routine urine examination, kidney function tests and Coomb's test and reticulocyte count were normal. CXR P/A view showed 'elevated right hemidiaphragm with multiple air shadow in liver area' (Fig-1).

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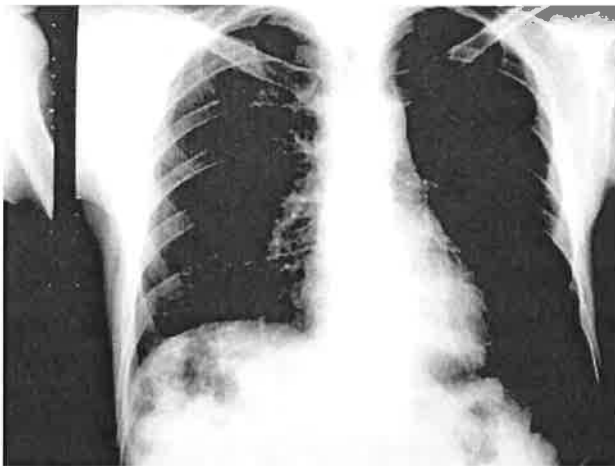


Fig-1: CXR P/A view showing elevated right hemidiaphragm with air shadow in hepatic area (Pneumobilia).

Ultrasonography of whole abdomen showed “ a mixed echogenic structure having both solid and cystic component measuring about 8.8× 6.9 cm present in the periphery of right lobe of liver” and air in left side of biliary channel (Pneumobilia) - suggestive of SOL in liver (Liver abscess). ERCP, computed tomography (CT) scan of abdomen (Fig-2) and MRI of Gallbladder and Cholangiogram showed multiple cystic dilated bile ducts within the liver occupying the both lobes but more in the right lobe. No feature of hepatic fibrosis was seen and both the kidneys were normal. All other biochemical investigations including random blood sugar & Alfa-Fetoprotein

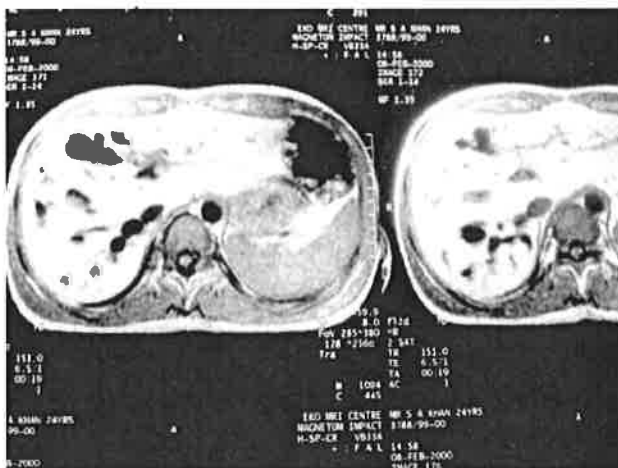


Fig-2: CT scan of liver showing Cystic dilatation with pneumobilia.

He was then diagnosed as a case of Caroli's disease with liver abscess. He was treated with Inj. Ceftriaxone 1 gm iv bid for 14 days and Inj Metronidazole 500mg iv tds for 10

days followed by cefixime 400mg bid for another 14 days and 3 units of packed cell transfusion. Aspiration from liver abscess could not be done due to extensive involvement and toxic condition of the patient. With above conservative treatment, his temperature was subsided, upper abdominal pain and pedal edema was resolved with remarkable clinical improvement. He was then discharged home with advice of follow up in every month.

Discussion:

There are two forms of Caroli's disease, one associated with congenital hepatic fibrosis and a simpler form occurring alone⁵. The former, called Caroli's syndrome is associated with portal hypertension. The later, known as Caroli's disease, may be associated with autosomal recessive polycystic kidney disease⁶ or rarely with autosomal dominant polycystic kidney disease⁷. Caroli's disease has also been reported in patient with choledochal cysts for which reason some authorities classify it as a type of choledochal cyst⁸. Caroli's disease may be localized to one lobe of liver or may be diffuse. It results from an arrest in ductal plate remodeling at the level of the larger intra-hepatic bile ducts³. Caroli's disease usually presents with intermittent abdominal pain and hepatomegaly⁹. Cholangitis, cholelithiasis, biliary abscess, septicemia, liver cirrhosis and cholangiocarcinoma are all its potential complications. Malignant complication (cholangiocarcinoma) occurs in approximately 7% of cases¹ and is due to prolonged exposure of the ductal epithelium to high concentration of unconjugated secondary bile acids¹⁰.

The patient discussed in this report is a pure form of Caroli disease with no evidence of periportal fibrosis and renal cystic disease. Our case also had history of 4 episodes recurrent cholangitis since 1996. This was also complicated with recurrent stone formation (cholelithiasis & choledocholithiasis) for that he had undergone ERCP& endoscopic extraction of CBD stone with papillotomy in 1996 and cholecystectomy in 2001.

The diagnosis of Caroli's disease rests on demonstrating that the cystic lesions are in continuity with the biliary tree. It can be done by imaging studies such as abdominal

USG, CT scan, and isotope scan, ERCP, PTC and MRCP. These studies demonstrate irregular cystic dilatation of the large proximal intrahepatic bile ducts with normal ducts in between. Current Ultrasonography of our case also showed cystic dilatation of biliary tree with extensive pneumobilia with an SOL suggestive of Liver abscess. Previous MRI and Cholangiogram also supported our current findings. Similar findings were observed in some other studies^{11,12}. In one case report, Ultrasonography showed multiple cystic lesions in the right lobe of liver and Color Doppler study showed evidence of multiple intrahepatic dilated biliary radicals and CT scan showed bizarre, saccular dilatation of intrahepatic bile ducts involving almost all subsegments of Bismuth and Corinauds in varying degree, most severely affecting right superior segment¹¹.

The treatment of Caroli's disease depends on the clinical features and the location of the biliary abnormalities. Cholangitis is treated with appropriate antibiotics. In case of intrahepatic cholelithiasis litholytic therapy with ursodeoxy cholic acid (UDCA) is indicated¹³. When the ductal abnormalities are localized to one lobe, lobectomy relieves symptoms and appears to remove the risk of malignancy. In case of diffuse involvements of both lobes of liver, treatment options include conservative management, endoscopic therapy (sphincterotomy for clearance of intra-hepatic stone), internal biliary bypass procedures and in carefully selected cases liver transplantation³. Those who can not be operated radically should have regular clinical follow up with ultrasound and liver biopsy if needed. Further family studies are needed in all cases to exclude the autosomal dominant mode of inheritance⁴. We also managed the case conservatively as there was diffuse involvement and family screening showed no relevant disease in his family.

Conclusion: Abdominal pain is a common presentation in Caroli's disease and though it is a rare congenital anomaly one should keep in mind the possibility of Caroli's disease in its differential diagnosis especially in patients who present with intermittent fever and hepatomegaly along

with abdominal pain and occasional jaundice.

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Conflicts of interests: None

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