CASE REPORT

Caroli's Disease: A Case Report

Abu Hena Mostofa Kamal[1], Mohammad Ali Sarker[2], Kabir Hossain Molla[3], Kudrat E Khuda[4]

Introduction:

Segmental, saccular or fusiform non-obstructive dilatation of intrahepatic biliary channels is referred to as Caroli's disease (CD). Two forms of this rare congenital disorder are described. The 'pure'form which involves only the large intrahepatic bile ducts is less common. The more common 'complex'form, which is associated with congenital hepatic fibrosis (CHF) is designated as Caroli's syndrome (CS)[1]. The inheritance of Caroli's disease is uncertain but in majority of cases it is transmitted in autosomal recessive fashion[2,3]. CD is associated with renal involvement is about 60%, and it includes medullary sponge kidney disease or polycystic kidney disease[4]. Review of the literature suggests that there are no large series of this disorder[5].

Although the disease present from birth, it usually remains asymptomatic during the first 20 years, and may also remain so throughout the life. However when becomes symptomatic, a significant number of patients present significant loss of quality in their life and the clinical course of the disease frequently worsen due to the repeated episodes of cholangitis with the presence of intrahepatic calculi, intrahepatic abscesses and sepsis6.

Case Report :

Mrs. Ayesha Akhter of 32 was admitted in holy family red crescent medical college hospital with the complaints of recurrent pain in right upper abdomen for last 2 months. The pain was colicky in nature with radiation to the back. It was subsided after taking some antispasmodics but increased repeatedly. During those attacks the pain was not associated with other complaints. But for last few days pain was severe and associated with vomiting and fever. Vomitus was not blood stained and there was no diarrhea. The patient non-diabetic, non-hypertensive, and non-asthmatic.

She has a history of admission in this hospital about 10 months ago with the complaints of severe abdominal, fever with jaundice and diagnosed as a case of acute pancreatitis with intrahepatic lithiasis.With conservative management she was recovered that time.

On examination the patient was febrile but there was no jaundice, anaemia or any stigmata of chronic liver disease . She was hemodinamically stable, abdomen was soft, right hypochondrium was tendered, liver spleen not palpable and shifting dullness was absent. Other systemic examination findings were normal.

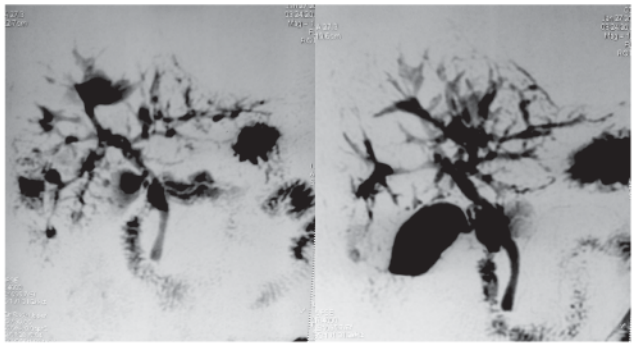
Investigations revealed HB 10.9 gm/dl, ESR 95 mm in 1st hour, WBC count 11,100/cmm, N 62.6%,

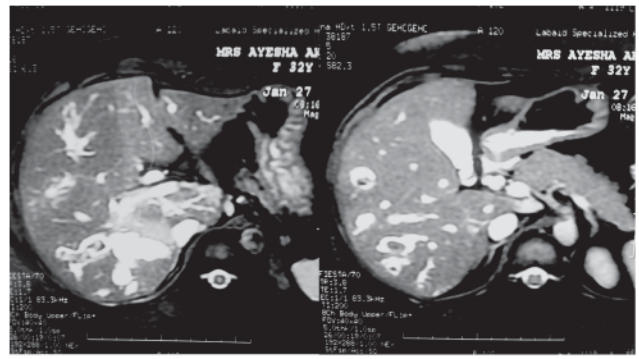
1. Professor, Department of Medicine, Holy Family Red Crescent Medical College, Dhaka.
2. Professor, Department of Radiology and Imaging, Holy Family Red Crescent Medical College, Dhaka.
3. Assistant Professor Department of Medicine, Holy Family Red Crescent Medical College, Dhaka.
4. Resident Physician, Department of Medicine, Holy Family Red Crescent Medical College, Dhaka.

L-29.7%, RBS-8.4mmol/L, S. bilirubin-0.3mg/dl, SGPT- 17.0 IU/L, SGOT- 22.0 IU/L, S. alk. Phosphatase- 138.0 IU/L, Total protein-5.97gm/L, S. albumin-4.09gm/dl, Globulin-1.90gm/dl. Serum electrolyte and prothrombin time were within normal limit. Lipid profile-total cholesterol-132.7mg/dl, TG- 160.0 mg/dl, HDL- 19.6 mg/dl, LDL- 84.5mg/dl. Ultrasonogram (USG)- Features of pancreatitis with prominent intrahepatic biliary trees, liver was slightly enlarged, no splenomegaly or ascites. CT-scan of hepatobiliary system- gross segmental dilated intrahepatic biliary ducts in postero-superior aspect of liver, suggesting Caroli's disease. MRCP findings compatible with Caroli's disease(localized - postero-superior aspect).

Treatment- The patient was treated conservatively with i.v fluid, ing. Ceftriaxone , ing. Metronidazole, analgesic- tramadol, paracetamol. With these medications the patient was recovered and discharged with advice for follow up at OPD after one month.

At follow up after 1 month she was asymptomatic and referred to a hepatobiliary surgeon for partial hepatectomy.





Figures 1-4 : MRCP images. Showing multifocal saccular dilatation of intrahepatic biliary ducts.

Discussion :

Caroli's disease is a rare form of congenital dilatation of the intra-hepatic bile ducts. Although its cause is unclear , Jorgensen and others have suggested involvement of ductal plate malformation during development, the circular ductal plates surrounding the portal vein are remodeled and leaving a network of small bile ducts around the portal veins. In ductal plate formation, resorption of the circular plates remains incomplete, resulting in a semicircular or cylindrical lumen that contains a portal radicle[7].

Diagnosis of Caroli's disease is usually done radiologically-Ultrasonogram and CT scans may be regarded as useful initial investigations but many other patients require further investigations like MRCP, ERCP or PTC to confirm the diagnosis[8]. USG didn't help to confirm the diagnosis but CT scan and MRCP confirmed the diagnosis.

Caroli's disease may present at any age although it is most commonly diagnosed between the second and fourth decades of life[9]. as was our case. CD rarely present after the age of 55 years. The common presentations of this disease are features of cholangitis[2] and our patient also presented with features of cholangitis. This

patient previously presented once with acute pancreatitis about 10 months ago. Though pancreatitis is a rare presentation of CD but it may happen[10].

Treatment of Caroli's disease depends on the location and extension of the disease. Partial hepatectomy is the preferred option for localized disease and orthotopic liver transplantation is the ultimate treatment for diffuse hepatic involvement[11]. Our patient was suitable for partial hepatectomy as it was a localized case and we referred her to a hepato-biliary surgeon.

In conclusion, CD is an inherited disorder which may cause severe life threatening cholangitis or cholangio-carcinoma or hepatobiliary degeneration[12]. Caroli's disease may be misdiagnosed due to lack of experience and unavailability of many of the diverse modern imaging techniques. For this reason, despite its rare incidence; Caroli's disease should not be ignored in the differential diagnosis of recurrent cholangitis.

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