Case Report

Caroli’s disease: A case report
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Abstract
Caroli’s disease (congenital dilatation of the intrahepatic ducts) is a rare condition which is usually diagnosed postoperatively. Cholangitis, hepatic abscess, cholangiocarcinoma are its potential complications. A case of Caroli’s disease in a 20 years old lady with extensive stone formation is reported here.

Key words
Caroli’s disease, choledocholithiasis, hepaticolithiasis

Introduction
Caroli’s disease first described by Jacques Caroli and his associates in 1958, is a rare congenital condition characterized by non-obstructive saccular or fusiform, multifocal, segmental, cystic dilatation of intrahepatic bile ducts. Mode of inheritance is usually in autosomal recessive fashion affecting both sexes equally. This disease usually presents with recurrent cholangitis and hepatomegaly.

Case report
A 20 yrs. old unmarried muslim female from poor socio-economic condition came to the Noakhali 250 Bedded General Hospital, with complaints of fever, abdominal pain and distension, yellow discoloration of sclera and urine for 8 days. Fever was high grade, continuous associated with chills and rigor. Abdominal pain was severely aching, maximum in right hypochondrium, no radiation, no relation with meal or change of posture. The patient noticed that she had been suffering from this sort of illness for several times. Her father died of geriatric illness and there is no H/O such illness among her siblings.

Clinical examination revealed she was febrile (Temp = 1030F) icteric, mildly anaemic, her liver was palpable (5 cm from costal margin in mid clavicular plane), tender, firm, smooth surfaced and well defined margin; mild ascites was present. After admission into hospital, laboratory investigations showed anaemia (Hb- 8 gm%), neutrophilic leucocytosis (TLC 13.8 x 103/mL, polymorph 83 %), raised ESR (55 mm at the end of 1 hour). Liver function tests showed S. Bilirubin= 6.7 mmol/L, SGPT= 56 U/ml, Alkaline Phosphatase = 516 U/ml, and raised Prothrombin time (18 seconds, control= 12 seconds). Serum Total Protein = 6.2 gm/dl, Serum Albumin = 3.10 g/dl. USG showed hepaticolithiasis in both lobes and choledocholithiasis with features suggestive of chronic cholecystitis and chronic hepatitis. CT scan showed dilated common bile duct and intrahepatic ducts containing multiple calculi within dilatations. Diagnosis was made cholangitis due to Caroli’s disease and treated conservatively with antibiotics, analgesics and anti ulcerant. After 14 days she improved clinically and was discharged with advice of follow up with ERCP and oral cholangiogram.

Figure 1: USG showing calculi in dilated CBD, Rt. Intrahepatic duct & Lt. Intrahepatic duct

Figure 2: CT Scan showing A. Calculi in dilated Rt. Intrahepatic duct; B. Calculi in dilated Lt. Intrahepatic duct; C. Calculi in dilated CBD

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In first follow up visit in OPD, oral cholangiogram showed non functioning gall bladder, ERCP showed dilated intrahepatic ducts containing multiple calculi with cholangitis (papillotomy done). Her clinical condition was found much better and she was counselled with provision of some supportive treatment (multivitamins, antiulcerant) as well.

**Discussion**

Carolii's disease occurs in two forms: (1) Pure form (Caroli disease) characterized by ectasias of intrahepatic bile ducts often limited to one hepatic lobe (mainly the left) remarkably sparing liver parenchyma and (2) Combined form (Caroli syndrome) in which ectasia of intrahepatic bile ducts is associated with chronic hepatic fibrosis, choledochal cyst and renal cystic disease. The whole liver usually affected and extra hepatic biliary dilatation occurs in about one quarter of patients. Kidney lesions include renal tubular ectasia (medullary sponge kidney, cortical cyst), lesions of adult recessive polycystic kidney disease. Clinical presentation of patients with Carolii's disease is heterogenous because symptoms may be absent for years, may occure at a very early age, or infrequently throughout life. Patients may experience recurrent episodes of abdominal pain, fever and intermittent obstructive jaundice caused by cholangitis and stone formation. Complications include biliary abscess, septicaemia and liver chirrhosis.

Malignant complication (cholangiocarcinoma) found in approximately 7% of cases. Infectious pathogens may become resistant to antibiotic treatment and sepsis frequently leads to death or secondary biliary cirrhosis.

The diagnosis of Carolii's disease involves recognition of the symptoms of liver disfunction and imaging studies. Imaging studies include abdominal sonography, CT scan, Endoscopic Retrograde Cholangiography (ERC), Percutaneous Transhepatic Cholangiography (PTC) and Magnetic Resonance Cholangiography (MRC).

Because of the disease's rarity and rather unspecific symptoms, diagnosis often delayed. Treatment should be focused on conservative or interventional (Endoscopic sphincterotomy) attempts first. Cholangitis is treated with appropriate antibiotics. In case of intrahepatic cholelithiasis, litholytic therapy with urso deoxy cholic acid is indicated. Indications for surgical treatment includes failure of conservative treatment, suspected malignancy or symptoms associated with chronic hepatic fibrosis. In patients with Carolii's disease confined to one lobe, hemi-hepatectomy (Rt./Lt.) is the choice. In contrast diffuse Carolii's disease needs an extended resection (multisectionectomy). For patients with diffuse involvement of both lobes in association with cirrhosis or associated hepatic fibrosis, liver transplantation is the option.

**Conclusion**

Although Carolii's disease is a rare congenital anomaly, it should be included in differential diagnosis when presenting with abdominal pain and hepatomegaly and sonographic study suggests multiple cystic lesion of the liver and multiple calculi within intrahepatic and extrahepatic biliary channel.

**References**

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