



“A RARE CASE OF ORIENTAL CHOLANGITIS IN AN INDIGENOUS WOMAN IN NORTHEAST INDIA”

Radio-Diagnosis

Dr. Pintu Biswas*	Senior Resident, Department Of Radiology, Agartala Government Medical College & Gb Pant Hospital, Agartala, West Tripura *Corresponding Author
Dr Sandip Nath	Post Graduate Trainee, Department Of Radiology, Agartala Government Medical College & Gb Pant Hospital, Agartala, West Tripura
Dr. Arun Reang	Post Graduate Trainee, Department Of Radiology, Agartala Government Medical College & Gb Pant Hospital, Agartala, West Tripura

ABSTRACT

Oriental cholangitis also known as recurrent pyogenic cholangitis is a complex disease that is characterized by intrahepatic pigmented stones and recurrent attacks of cholangitis. This is a condition found almost exclusively in individuals who lived in Southeast Asia. We report a case of an indigenous woman diagnosed with oriental cholangitis after presenting with a 7–8 year history of recurrent fevers, jaundice and upper abdominal pain. A detailed evaluation led to the diagnosis of oriental cholangitis with improvement after biliary decompression and antibiotics.

KEYWORDS

Oriental cholangitis, jaundice, hepatolithiasis, biliary stricture

INTRODUCTION

Oriental cholangitis, also known as recurrent pyogenic cholangitis (RPC) is a complex disease that is characterized by intrahepatic pigmented stones and recurrent attacks of cholangitis [1]. This is a condition found almost exclusively in individuals who lived in Southeast Asia [2]. This condition was first described by Digby in 1930, who observed eight Chinese patients with recurrent bacterial cholangitis associated with hepatolithiasis. Since then various other names have been used such as 'Hong Kong disease', 'biliary obstruction syndrome of the Chinese' and 'recurrent pyogenic cholangitis (RPC)' [3]. It is strongly associated with biliary infestation by such as *Ascaris lumbricoides* & *Clonorchis sinensis*. In the Indian subcontinent ascariasis is highly endemic in Kashmir (70%), Bangladesh (82%), central and south west India (20%-49%) though is seen exclusively in Kashmir as reported by Khuroo et al [4].

Case Report

A 36 years old female from a remote rural place of Tripura in Northeast India presented in emergency department with pain in upper abdomen for 3 days, fever for 1 day & yellowish discoloration of sclera for 15 to 20 days. After taking an extensive history of patient's illness, it has been found that, patient is having intermittent pain abdomen, nausea, bloating, intermittent fever & jaundice for last 7 to 8 years. For that she got symptomatic relief in local hospitals. She had a history of open cholecystectomy 16 years back for cholelithiasis.

Upon admission, physical exam was otherwise unremarkable except for tenderness in upper abdomen, fever of 101.5 °F, jaundice, and mild pedal edema. In laboratory studies antinuclear antibody test, anti-mitochondrial antibody test & rheumatoid factor was negative. Hemoglobin was 10.5 gm/dl, White blood cell (WBC) count was 8.3k /micro liter with eosinophilia of 9%. Liver tests were consistent with obstructive pathology with total bilirubin of 19.4 mg/dL, direct bilirubin of 12.8, alkaline phosphatase of 247 U/L, aspartate aminotransferase of 199 U/L, alanine aminotransferase of 421 U/L, and gamma-glutamyl transferase of 700 U/L. Serum urea was 76 mg/dl & creatinine was 2.1 mg/dl.

After these initial work ups patient was sent to Radio-diagnosis department when we started evaluating the case. Initial X-ray of abdomen was unremarkable other than some linear radiopaque lesions consistent with cholecystectomy clips.

In ultrasonography of abdomen there were dilatation of intra hepatic bile ducts with multiple hyperechoic lesions associated with poorly formed posterior acoustic shadow seen along the distribution of right & left hepatic ducts (Figure 1A). Common bile duct (CBD) was dilated. No calculus could be found in CBD, though the distal part of it could not be visualized satisfactorily due to duodenal gas shadow. Gall bladder (GB) was not visualized, but GB fossa also could not be clearly visualized due to some unusual acoustic shadow.

After that, magnetic resonance cholangio pancreatography (MRCP)

was advised. In MRCP, T1 VIBE images shows multiple signal void lesions in intrahepatic biliary radicles with dilated bile ducts. The largest signal void calculus measured approx. 0.86x0.52 cm. In T2 HASTE images shows peri-portal ill-defined hyper intense signal seen reflecting associated portal triad inflammatory changes. Multiple intra hepatic biliary strictures seen with interrupted segments of dilatation involving the right & left anterior ducts reaching up to capsule (Figure 1B & 2A).

The common bile duct was dilated measuring approx. 1.48 cm in diameter with no signal void within.

In all the sequences of MRCP, the GB fossa – again was not clearly visualized due to superimposition of multiple signal void foci & dilated IHBR. The rest of the liver was unremarkable. The spleen, pancreas & bilateral kidneys were unremarkable.

To assess the GB fossa, then non-contrast computed tomography (NCCT) of abdomen was performed, as intravenous contrast could not be given since serum creatinine was raised.

In NCCT abdomen, there was moderate intrahepatic biliary ductal dilatation in bilateral hepatic lobes with narrowing of the intrahepatic bile ducts towards the hilum. Gall bladder was not seen- consistent with previous cholecystectomy. GB fossa was unremarkable. Multiple rounded hyper dense lesions were seen within the distended intrahepatic bile ducts, resembling biliary calculi (Figure 2B). The common bile duct was dilated. Other organs were unremarkable.

DISCUSSION

Oriental cholangitis is characterized by recurrent bacterial cholangitis and the presence of calculi within the intrahepatic bile ducts & biliary strictures. The incidence of the disease among men and women is comparable. The stones form within the intrahepatic biliary system and are mainly composed of calcium bilirubinate or brown pigment, in contrast to the cholesterol stones more commonly seen in patients with other gallstone-related diseases, which is the vast majority of patients in the west [3].

The gallbladder is usually remains disease free in most of the cases. It commonly presents with the classical Charcot triad which includes right upper quadrant pain, fever, and jaundice. It is strongly associated with malnutrition, low protein intake, and biliary infestation by trematodes such as *Ascaris lumbricoides* or nematodes such as *Clonorchis sinensis*. *Clonorchis sinensis* is a liver fluke which is having a lifespan of 20 years and can thus manifest itself many years after initial infestation. It is said to cause a disruption in the bile duct's epithelial barrier leading to a cascade of transient portal bacteremia with gut flora (e.g. *E. coli*, *Klebsiella*, *Pseudomonas*, and *Proteus*) which leads to secondary stone formation and increased biliary pressure.

Some studies have shown that debris of ova and parasites in the ductal

system may serve as a nidus for calcium bilirubinate stone formation [2]. There is involvement in both the left and right hepatic lobes and the common bile duct. The left hepatic duct is usually more severely and frequently affected presumably due to its more horizontal configuration which may affect bile drainage.

Ultrasound is the first preferred initial test to demonstrate biliary dilation, hepatolithiasis and liver abscess. Endoscopic Retrograde cholangio pancreatography (ERCP) is both diagnostic and therapeutic and allows for the delineation of the biliary tree and definitive decompressive procedures. But as it is invasive procedure it is not always possible to perform. MRCP can be done where ERCP is difficult or impossible which is particularly useful in the evaluation of proximal ductal obstruction in cases with severe biliary stenosis. The peripheral ducts, multiple intrahepatic biliary strictures are better depicted on MRCP.

Other diagnostic clues pointing to RPC include thickening and hyper enhancement of bile duct walls, stones within both intrahepatic and extrahepatic ducts, and lobar and segmental liver parenchymal atrophy.

There is evidence that the incidence of the disease is declining. The improved economic situation and living standards associated with westernization of diet are thought to be involved in the changing epidemiology in Asian countries [3].

If left untreated, RPC is associated with a high morbidity and mortality as it progresses to biliary cirrhosis and liver failure [5]. It is a well-known risk factor for cholangiocarcinoma [6].

Initial management should be directed at treatment of cholangitis with antibiotics and biliary drainage. A multidisciplinary approach is mandatory for the correct diagnosis and choice of proper management. In patients with dilated intrahepatic and extrahepatic bile ducts with intrahepatic stones, ERCP is usually the preferred interventional procedure.

Percutaneous transhepatic cholangiography and drainage is an important alternative or additional therapeutic procedure. In recent times this intervention procedure is in limelight. In specific cases, particularly with localized or unilateral involvement, segmental resection or hemi hepatectomy with or without hepatico-jejunostomy may be indicated and even be the therapy of choice.

CONCLUSION

Though cases of oriental cholangitis is most common in south east Asia, several sporadic cases also encountered in many regions of India. RPC is unique in its pathophysiology due to its de novo intrahepatic stone formation with sparing of the gallbladder. Management of this disease is a multidisciplinary challenge especially when there is a delay in diagnosis due to the extent and inaccessibility of strictures. So a careful history taking, routine blood tests & a thorough radiological evaluation can detect those cases very early.

Once prompt treatment and follow-up are initiated, patients will have early recovery with reduced burden of unnecessary hospitalizations. In our case the patient got discharged at 19th day of hospitalization after prompt antibiotic therapy & interventional radiological procedure.

Representative Images Of This Article:

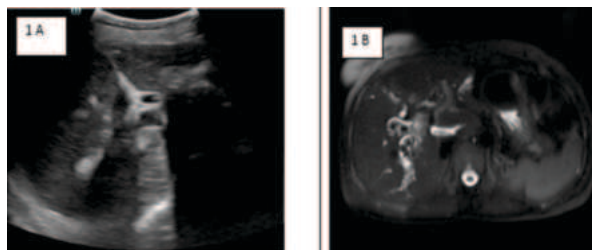


Figure 1- A & B. A) USG of abdomen showing evidence of multiple hyperechoic lesions with poorly formed posterior acoustic shadows seen in dilated intra hepatic biliary radicles. **B)** Axial T2 HASTE: Multiple signal void lesions in intrahepatic biliary radicles with dilated bile ducts; also peri portal ill-defined hyper intense signal seen reflecting associated inflammatory changes.

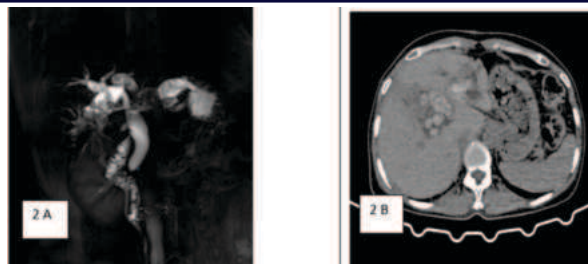


Figure 2. A & B. A) T2 HASTE FS COR: Multiple intra hepatic biliary strictures seen with interrupted segments of dilatation involving bilateral IHBRs with multiple calculi. **B)** NCCT of abdomen: showing dilated intra hepatic biliary radicles with multiple hyper dense calculi with in.

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