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Hepatolithiasis : A review of literature and report of three cases

KEYWORDS: Hepatolithiasis, gall bladder, bile duct

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ABSTRACT *Hepatolithiasis is defined as gallstones present in all bile ducts peripheral to the confluence of the right and left hepatic ducts, irrespective of the coexistence of gallstones in other parts of the biliary tract, such as the extrahepatic bile duct and/or the gallbladder. Hepatolithiasis or intrahepatic calculi are prevalent in East Asia, including Japan, but occurs much less frequently in Western countries. The disease is characterized by its intractable nature and frequent recurrence, requiring multiple operative interventions, in distinct contrast to gallbladder cholesterol or black pigment stones. Here, we present a report of three cases with hepatolithiasis, highlighting the morphological features of the same.*

Introduction :
Hepatolithiasis, is characterized by the presence of stones within the intrahepatic bile ducts proximal to the right and left hepatic ducts. Hepatolithiasis is rare in Western countries, and the incidence in East Asian countries, such as Taiwan, China, Hong Kong, South Korea, and Japan is higher. [1,2] Hepatolithiasis is benign in nature, but the prognosis is poor due to an association with recurrent cholangitis, biliary strictures, liver abscesses, and atrophy or cirrhosis of the affected liver. [3] It is also a known risk factor for intrahepatic cholangiocarcinoma.[4]

In this article we present a report of three cases highlighting the morphological features of each.

Case reports :
Case 1 : A 43 year old female patient came with complaints of pain abdomen for past two years and on and off yellowish discolouration of eyes and urine since 1 year. On examination tenderness in the right hypochondrium was present. On investigation, Hemoglobin was 12 gm/dl, other hematological parameters were also within normal limits.

Biochemical parameters showed serum bilirubin of 0.19 mg/dl, aspartate aminotransferase (AST) of 21 IU/L, alanine aminotransferase (ALT) of 14 IU /L and serum alkaline phosphatase 72 IU/L. Serology for hepatitis-B virus and hepatitis-C virus were negative.

CT scan of abdomen showed left lobar intrahepatic biliary dilatation with calculi involving segment II and III – post obstructive lobar atrophy.

The patient underwent a left lateral hepatectomy with cholecystectomy with Intraoperative cholangiogram. Intraoperatively, there was atrophied left lobe of liver with multiple intraductal calculi. Intraoperative cholangiogram showed no filling defects.

Grossly, the specimen measured 6.5x6x2 cms. Cut surface showed multiple dilated ducts filled with multiple black coloured calculi (Fig 1). Histopathologically, the liver parenchyma was atrophied and showed periductal and periportal lymphocytic infiltrate (Fig 2). Bile ducts were dilated and lumen showed cross section of pigment stones. The gall bladder showed features of chronic cholecystitis.

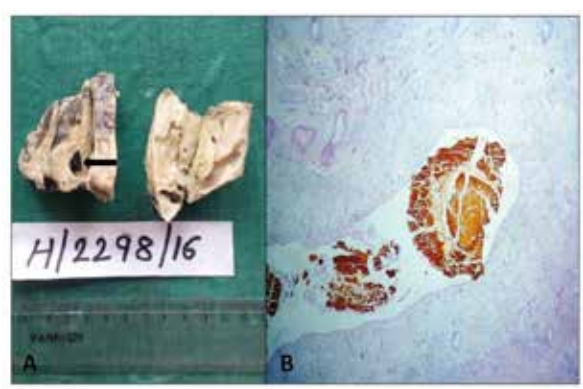

Figure 1 : A) Gross specimen of hepatic segmentectomy showing dilated ducts filled with calculi B) Atrophied liver parenchyma showing cross section of pigment stones in the duct lumen. H&E stain 100x

Case 2: A 50 year old female was admitted with complaints of pain abdomen and vomiting since 1 week. On examination, vitals were stable, icterus was present. Right hypochondrial tenderness was present. Ultrasound of the abdomen showed multiple CBD calculi. MRI showed choledocholithiasis with biliary obstruction, bilobar intrahepatic biliary dilatation and left intrahepatic biliary ductal calculi causing obstruction with post obstructive lobar atrophy. The patient underwent left hepatectomy with hepaticojejunostomy.

Grossly, the gall bladder showed denuded mucosa and multiple pigment stones. Liver showed few dilated ducts

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and black coloured calculi. Histopathologically, the hepatic parenchyma was largely unremarkable and showed dilated bile ducts surrounded by fibrosis and chronic inflammation. Dilated ducts showed dark brown calculi. The gall bladder showed features of chronic cholecystitis. (Fig 2)

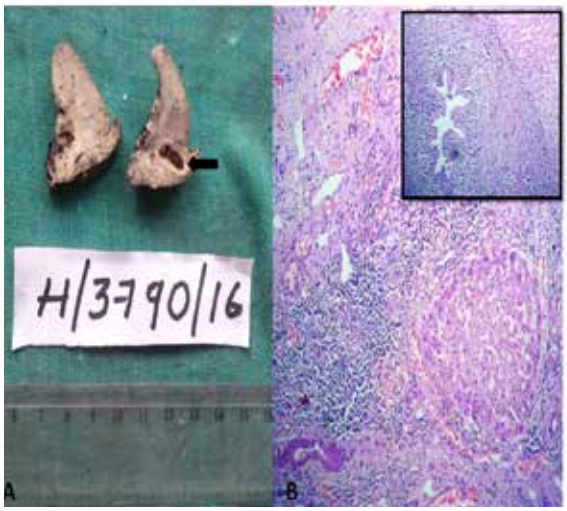

Figure 2 : A) Gross specimen of liver showing dilated duct with luminal calculi B) Hepatic parenchyma showing inflammation and fibrosis , Inset shows periductular inflammation. H&E stain 400x

Case 3: A 70 year old male came with complaints of pain abdomen for past 1 month. On examination vitals were stable. Right hypochondrial tenderness was present. On investigation, hematological parameters were within normal limits. Total bilirubin was 0.32mg/dl, Serum AST 47 IU/L, Serum ALT 42 IU/L. CT showed choledocholithiasis and left lobar intrahepatic biliary ductal calculi. MRCP showed distended gall bladder with calculi and intrahepatic biliary dilatation (Fig 3). The patient underwent left lateral segmentectomy with cholecystectomy and intra operative cholangiogram. Intraoperatively, multiple adhesions were noted. Atrophied segment 2 and 3 of liver with multiple hepatic duct calculi. Gall bladder wall was thickened and large calculi was present in the gall bladder neck. Grossly, the gall bladder wall was thickened and showed pale brown nodularities. The segment of liver showed dilated ducts filled with black coloured stones. Histopathologically, the gall bladder showed features of acute on chronic cholecystitis. The liver exhibited parenchymal fibrosis with numerous dilated biliary ducts. Neural hyperplasia and Monckebergs medial calcification of blood vessels were noted. (Fig 4)

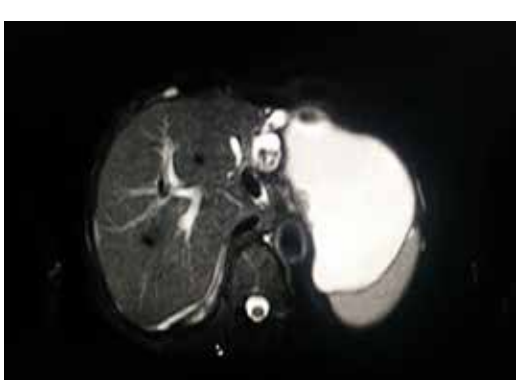

Figure 3 : MRCP showing distended gall bladder with calculi and intrahepatic biliary dilatation.

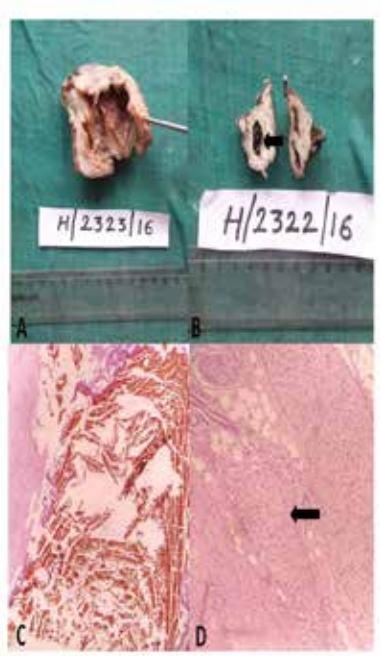

Figure 4 : A) Specimen of gall bladder showing markedly thickened wall and denuded mucosa B) Liver showing dilated ducts and dark brown to black calculi C) Dilated duct showing luminal calculi H&E 400x D) Atrophied hepatic parenchyma exhibiting neural hyperplasia (arrow) H&E 400x

Discussion : Hepatolithiasis is most common in East Asian countries where the prevalence has been reported to be 20% of all patients undergoing surgery for gall stone disease. In the west, it is generally thought to be secondary to gall bladder stones or primarily associated with sclerosing cholangitis, benign biliary strictures, choledochal cysts or malignant biliary tumours [5]. In the East it is regarded as a separate entity altogether. Majority of cases are associated with recurrent pyogenic cholangitis in regions with parasitic infestations like 'Ascaris lumbricoides' and 'Clonorchis sinesis.' Co-existence of parasitic infestations is common and appears to be incidental rather than causative. [6] The main morphologic feature of stone-containing bile ducts in hepatolithiasis is chronic proliferative cholangitis and peribiliary glands proliferation, in which the epithelial lining becomes hyperplastic. [1] Long standing cases of hepatolithiasis exhibit post obstructive hepatic atrophy as seen in two of our cases, along with periductular fibrosis and inflammation. In our patients the exact etiology of hepatolithiasis is unclear, however, the most probable possibility could be the extrahepatic bile duct calculi which would have played a role in development of intrahepatic stones.

Conclusion : Although the etiopathogenesis of hepatolithiasis is complex, a diligent histopathological evaluation can help in narrowing down the etiological factors and also in detecting pre malignant and malignant lesions associated with hepatolithiasis.

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