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Case Report

Choledochal cyst type VI: a rare case report

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ABSTRACT

Choledochal cyst or biliary cyst is a congenital anomaly affecting the biliary tree. It involves the dilatation of the biliary tree that could affect the extra hepatic and/or the intrahepatic segments. We present a 40 year old female with constant right upper abdominal pain, diagnosed with choledochal cyst. She underwent laparoscopic cholecystectomy with choledochal cyst (CDC) excision. She was uneventful postoperatively and then discharged.

Keywords: CDC, MRCP, Laparoscopic cholecystectomy

INTRODUCTION

Choledochal cysts (CDC), are rare congenital dilations involving the extrahepatic biliary tree with or without dilation of the intrahepatic bile ducts.[1] It is a rare condition with incidence in western population of 1 in 100000-150000, and remarkably high in Asian population, with 1 in 1000 births.[2-4]

They are commonly classified into five types. A new type, type VI, causing dilation of the cystic duct between the neck of the gall bladder and the common hepatic duct (CHD) has also been described by some authors which is the rarest of all these subtypes. These are commonly seen in middle aged females and are mostly symptomatic. Most of the time they need magnetic resonance cholangiopancreatography (MRCP) for accurate diagnosis. Management for these lack any definite criteria but range from simple cholecystectomy to complete excision of the entire bile duct and biliary reconstruction, due to chances of malignant transformation in these cysts. Hence, these rare cysts, must wander a surgeon's mind while handling suspicious cystic lesions in the biliary tract. One such rare case and its management of CDC in middle aged female is discussed here.

CASE REPORT

A 40-year-old female presented with complaints of pain in periumbilical and right upper quadrant of abdomen which aggravated by fatty meals associated with nausea for 2 months of duration. No vomiting, fever, itching, anorexia, altered sleep pattern, change in stool form, yellowish discoloration of skin. No H/O OCPs intake.

Clinical examination of abdomen was normal and routine laboratory investigations were within normal limits. Ultrasound of the abdomen revealed a fusiform dilatation of mid part of cystic duct with maximum diameter 3.8 cm. There was no intrahepatic biliary radicle dilatation (IHBRD). MRCP revealed a fusiform dilatation of mid part of cystic duct with maximum diameter 2.8 cm revealing CDC and pancreatic cyst communicating with pancreatic duct was also found. CECT abdomen revealed choledocal cyst type VI. Serum CEA was done keeping in view of the pancreatic cyst and the results were within normal limits.

Laparoscopic cholecystectomy was done and gall bladder along with CDC arising from cystic duct were excised and sent for histopathological examination.

The postoperative course was uneventful and patient was discharged on postoperative day (POD) 5 after surgery. On histopathological examination the specimen revealed chronic cholecystitis with CDC.

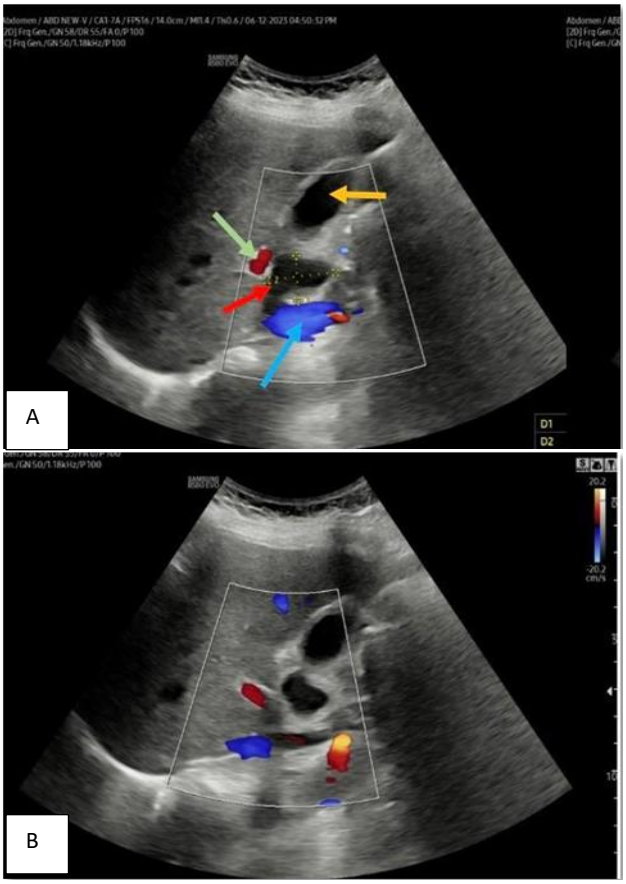

Figure 1 (A and B): USG images.
USG images in coronal plane shows fusiform dilation of cystic duct adjacent to the bladder. Red arrow indicates fusiform dilation of cystic duct. Blue indicates portal vein. Yellow arrow indicates normal gall bladder, green arrow indicates hepatic artery.

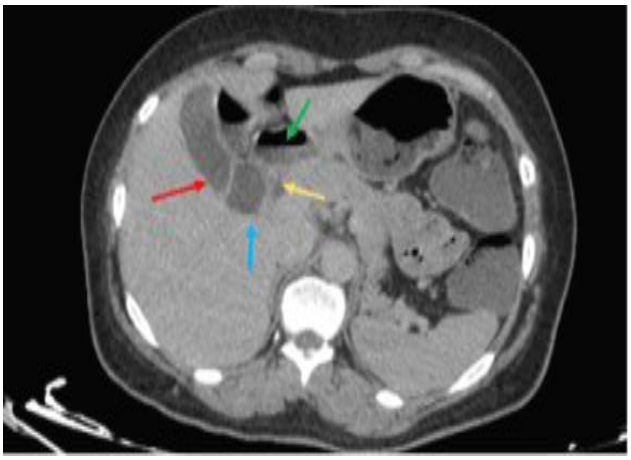

Figure 2: CECT abdomen image of axial plain CT.
Red arrow indicates normal gall bladder, blue arrow indicates dilated mid part of cystic duct, yellow arrow indicates normal CBD. Green arrow indicates second part of duodenum.

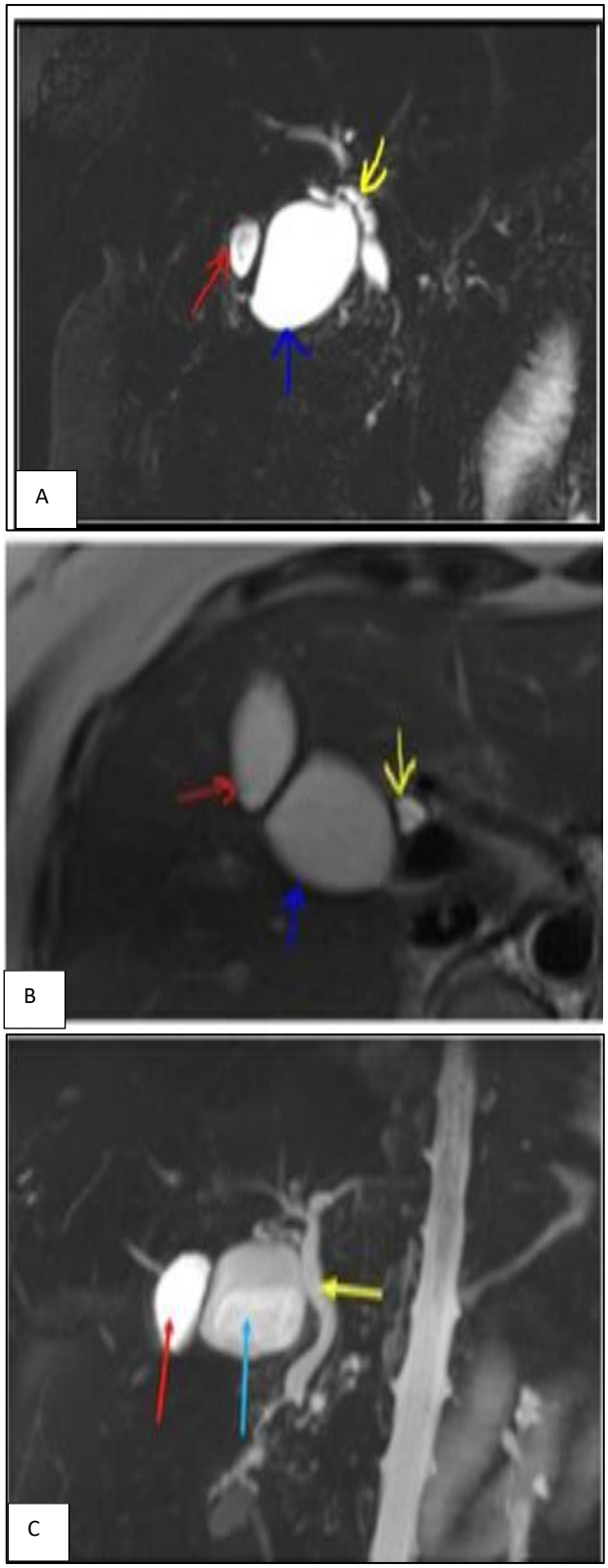

Figure 3 (A-C): MRI images.
T2 weighted coronal images and transverse, dilated of mid part of cystic duct is noted. On another T2 weighted coronal images dilation of mid part of cystic is noted along with clear visualization of biliary system. Red arrow indicates normal gall bladder and blue arrow indicates dilated mid part of cystic duct and yellow arrow indicates normal CBD.

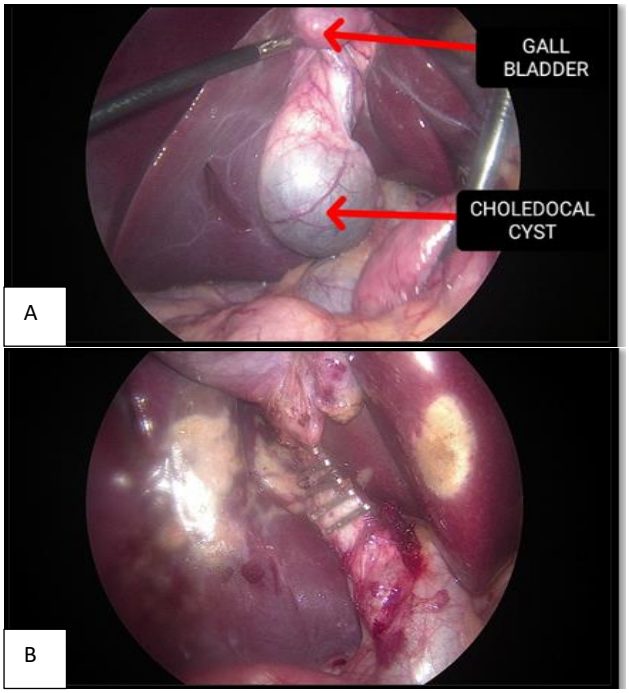

Figure 4 (A and B): Intraoperative laparoscopic image of CD.

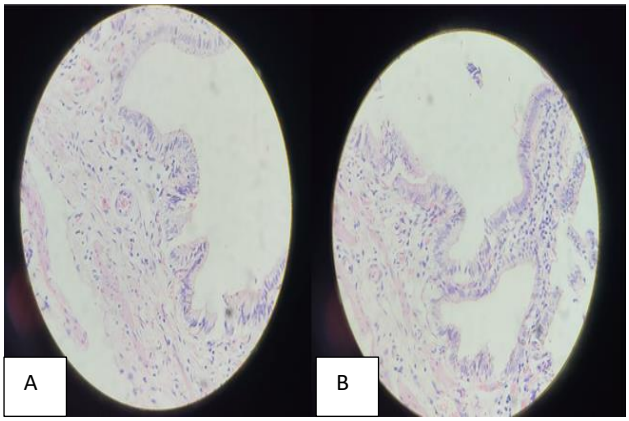

Figure 5 (A and B): Histological slides of cyst showing cyst lining to be same as that of gall bladder with no dysplastic changes.

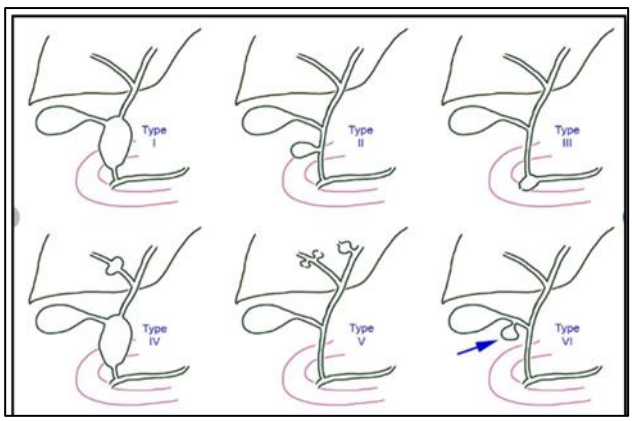

Figure 6: Types of CDC.

DISCUSSION

CDC commonly cause dilations in the extrahepatic parts of biliary apparatus. Widely used classification was given by Todani et al into five types where type I CDC being commonest (60%) causing fusiform dilation of the common biliary duct apparatus.[5] Type II involves saccular diverticulum of the CBD, type III involves perivaterian part of CBD, type IV involves multiple focal dilations of the bile ducts which are further subdivided into 4a-extrahepatic with intrahepatic involvement and 4b-extra hepatic involvement only. Type V i.e. Caroli's disease involves the intrahepatic bile ducts only.

Serena Serradel et al modified the widely accepted and used classification of Alonso-Lej which was previously modified by Todani et al including cystic duct cysts as a separate entity.[5,6] It was described by Bode and Aust in 1983, but given the rarity of these lesions, only a few cases have been described till now in literature.[7]

Most of these lesions in cystic duct are symptomatic, most common symptom being epigastric and/or right upper quadrant pain aggravated by a fatty meal (as seen in this case). We don't know exact etiology of these cysts; type VI CDC is thought to occur due to ectasia at the cystic duct caused by an abnormal pancreaticobiliary duct junction (APBDJ) as in this case pancreatic cyst communicating with pancreatic duct was found on MRI.[7-10]

Initial investigation for these cases is abdominal ultrasonography which acts as screening for abnormal pathology. MRCP is ideal option to give a clear idea of the entire biliary system including the course of the cystic duct, presence or absence of ABBDJ, GB thickening, presence of gall stones, IHBRD, and CBD involvement. Next invasive option is endoscopic retrograde cholangiopancreatography (ERCP) which provides with the same information and detail regarding the biliary system as MRCP. ERCP and Tc-99m hydroxy imino diacetic acid (HIDA) scan can be used for diagnosis but are not commonly used due to their invasiveness.

Typical radiologic abnormalities that are specific to type VI CDC includes dilatation and squaring of the cystic duct, acute angulation of the CHD, and cystic duct junction with a distinct plane present between the dilated cystic duct and CHD, a normal or wide (Mirrizi's syndrome) opening of the cystic duct to the CBD, a normal CBD, and associated APBDJ.[11] Most common differential diagnosis is a type II or type I CDC due to similarities in appearance of cyst in close proximity to CBD.

Type VI choledocal cyst is is a rare anomaly involving cystic duct. Cases are reported here cystic duct alone is involved alone and, in another type, common bile duct with cystic duct is involved.[12] We need to know that these distinct type of choledochal cyst exists and a new

standard classification is needed. It is important to classify type 6 into two types A and B owing to their different management.

CONCLUSION

The current increasing use of MRCP to diagnose hepatobiliary problems will result in an increased number of such cystic duct dilations. Being in laparoscopic era, such cysts will also be seen intraoperatively while operating on cases of acute cholecystitis or symptomatic biliary cholelithiasis. Hence, knowledge of type VI CDC, its diagnosis by MRCP, and treatment options are the urgent need of the hour for effective treatment and management of this rare entity.

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