CASE REPORT

A large choledochocystolithiasis mimicking Mirizzi syndrome

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SUMMARY

An 18-year-old man presented with spontaneous severe epigastric pain, progressing and radiating to back since 3 days. It was associated with epigastric tenderness, bilious vomiting and jaundice. He had been intermittently experiencing these symptoms for the last 1 year. No known comorbid. Ultrasound showed a poorly visualised heterogeneous focus at porta hepatis; considering poor visualisation, this might represent an enlarged calcified lymph node or cystic duct calculus causing extrinsic compression or a large sludge ball within the common bile duct (CBD), leading to dilatation of common hepatic duct and intrahepatic biliary system. Subsequent magnetic resonance cholangiopancreatography revealed a focal saccular dilatation of middle part of CBD, a type I-B choledochal cyst, large heterogeneous focus seen within it representing choledochocystolithiasis. Later, CT was performed for further characterisation of surrounding anatomy and pathology, which confused the appearance of choledochocystolithiasis for Mirizzi syndrome. Later, surgery and histopathology confirmed type I-B choledochocystolithiasis and chronic cholecystitis.

BACKGROUND

Choledochal cyst (CC) is an abnormal congenital dilatation of the extrahepatic or intrahepatic biliary tree¹ and is usually diagnosed during the childhood. There are around 20% cases which remain symptom free and, hence, undiagnosed until adulthood.² According to Todani's classification, the cyst formation within common bile duct (CBD) is classified as type I CC.^{3,4} Adult CCs frequently present with associated complications such as cholangitis, cholangiocarcinoma, traumatic perforation, abscess formation, portal hypertension and choledochocystolithiasis (CDC).^{4,5} CDC clinically presenting with signs of acute cholecystitis and obstructive jaundice is a rare entity. We are reporting a case of a young man with CDC who presented with clinical signs and symptoms of acute cholecystitis, but, on imaging, CDC appeared to be as Mirizzi syndrome (MS).

CASE PRESENTATION

An 18-year-old man was admitted to the emergency department (ED) with complains of colicky, spontaneous, severe epigastric pain, progressing and radiating to back since 3 days. It was associated with epigastric tenderness, bilious vomiting and

jaundice. He had been intermittently experiencing these symptoms for the last 1 year. The pain was mostly present after a meal with associated nausea. His further medical history was unremarkable with no known comorbid, previous surgical history, history of drug intake or any abdominal trauma. On physical examination, he was jaundiced with yellow sclera and pale skin. He was tender to palpation in right upper quadrant and epigastric region with mild guarding. The Murphy's sign was positive at the time of examination. The patient's body temperature at the time of presentation was 37.2°C.

INVESTIGATIONS

Routine laboratory testing was performed in ER. The liver enzymes were significantly raised: total bilirubin 6.7 mg/dL (0.1–1.2), direct bilirubin 5.1 mg/dL (0–0.2), alkaline phosphatase 449 IU/L (54–369), γ -glutamyltransferase 610 IU/L (<55), aspartate aminotransferase 27 U/L (<35), alanine aminotransferase 223 IU/L (<45), lactate dehydrogenase 270 U/L (<250), lipase 115 U/L (6–51) and amylase 61 U/L (<100). The patient was hyponatraemic (Na: 131) and hypochloreaemic (Cl: 96) with leucocytosis [white blood cell: 14.4 (4–10)] at the time of presentation.

An ultrasound examination was performed (figure 1) which showed dilated cystic duct (CD) with sludge within it. A poorly visualised heterogeneous focus at porta hepatis was also visualised, likely representing a sludge ball within extrahepatic biliary system. Only small proximal portion of CBD was visualised, and the rest could not be separately delineated. Considering poor visualisation, this might represent an enlarged calcified lymph node or CD calculus causing extrinsic compression leading to dilatation of CD, common hepatic duct (CHD) and intrahepatic biliary system. Subsequently, a magnetic resonance cholangiopancreatography (MRCP) was performed, which revealed (figure 2) a focal saccular dilatation of middle part of CBD (arrow heads), a type I-B CC.⁶ A large heterogeneous focus seen within it represents CDC, and it was measuring 4.3×4.2 cm. Distal CBD was also visualised. There was gross dilatation of CHD and intrahepatic ducts. The CD was also dilated and junction of CD and CBD was not visualised. Later, CT was performed for further characterisation of surrounding anatomy and pathology. It was obtained with the rationale to rule out extent and if there was any related complications of acute

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Figure 1 Ultrasound (US) examination of liver gall bladder (GB). (A) US (transverse section) showing dilated cystic duct with sludge in its lumen (*), GB appears unremarkable. (B) US (longitudinal section) shows a poorly visualised heterogeneous focus with posterior acoustic shadowing at porta hepatis (white arrows), likely representing a sludge ball within extrahepatic biliary system. Rest could not be separately delineated.

pancreatitis and to rule out malignancy. CT scan (figure 3) findings confused the appearance of CDC for an impacted large calculus probably at the junction of CD and CBD representing MS (arrow heads). It was assumed that it caused severe extrinsic compression of CBD and resulted in biliary system dilatation. No abnormal enhancement of biliary ductal system or lesion was seen.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included acute on chronic cholecystitis and acute pancreatitis secondary to either choledocholithiasis, CDC or MS.

TREATMENT

Patient was treated with analgesics to reduce pain; empirically treated with antibiotics including injection metronidazole and injection ceftriaxone; antiemetic, injection metoclopramide; and with injection omeprazole at the time of presentation in emergency department. Subsequently, a midline laparotomy was performed in supine position and surgical exploration of the biliary tract was done. Inflamed gall bladder (GB) was identified which was strongly adherent to surrounding extrahepatic biliary tree. The dissection was started near the hilum of the liver, CHD and CBD were lifted and a large type I-B CC was identified with a narrowed distal CBD. Cholecystectomy and excision of CC was performed. The resected GB was having a tan white, smooth, serosal surface with haemorrhagic areas. The mucosal surface was velvety. No stones were identified in the lumen of the GB. The CC appeared as tubular structure with a narrowed one end and a dilated blind end forming a cyst. The specimen measured approximately 6×4 cm. The proximal and distal margins were sent for frozen section, which turned out negative for any malignancy. A large, dark brown calculus was identified

within the CC, measuring approximately 4 cm with irregular surface. Some residual sludge was also noted within the CC. Significant sludge was seen coming out of CHD. However, no calculus was identified, on per-operative rigid choledochoscopy, in CHD. Subsequently, a Roux-en-Y hepaticojejunostomy was performed to restore biliary drainage system. Primary closure was done and two surgical drains were left in surgical bed. No complication was observed during and after the procedure. He was shifted back to ward after surgery, where he remained haemodynamically and vitally stable. His diet was gradually progressed, which he tolerated well. He was ambulated and his drains were removed at fourth postoperative day.

OUTCOME AND FOLLOW-UP

Histological examination showed that the GB wall was thickened, measuring 5.5 mm. The lamina propria and submucosa shows mild to moderate chronic inflammation with congested thick walled vessels. Muscularis propria showed thickening. There was no evidence of intestinal metaplasia, dysplasia or malignancy. The CC wall thickness was measuring 2 mm, with inflamed, partly denuded and ulcerated mucosa. Underlying wall shows moderate patchy chronic inflammation and congested vessels with areas of haemorrhage. There is no evidence of malignancy. The patient showed good recovery with resolution of pain, vomiting and signs of obstructive jaundice. The follow-up ultrasound after 1 week showed resolution of intrahepatic biliary dilatation (figure 4). Overall, the postoperative follow-up was unremarkable.

DISCUSSION

The anomalous cystic dilatation of extrahepatic biliary system is considered as CC.^{6,7} The ‘anomalous pancreatic duct–biliary junction theory’ is mostly accepted in explaining the development

Figure 2 Magnetic resonance cholangiopancreatography examination. (A) T2 sequence: axial section, (B) T2 sequence: coronal section and (C) volume coronal image. All showing dilated cystic duct (white arrow), gall bladder (GB) is shrunken with thick wall favouring chronic cholecystitis. A focal saccular dilatation of middle part of common bile duct (CBD) (arrow heads), a type I-B choledochal cyst, with a large choledochocystolithiasis (*) causing extrahepatic and intrahepatic biliary dilatation. Distal CBD (red arrow) appears unremarkable.

Figure 3 Biphase contrast-enhanced CT examination. (A) Axial section: venous phase, (B) coronal section: venous phase and (C) coronal section: arterial phase. There is suspicion of a large impacted calculus (*) probably at the junction of cystic duct (CD) and common bile duct (CBD) representing Mirizzi syndrome (white arrow heads) rather than a choledochocystolithiasis. Mid and distal CBD (black arrow) appears unremarkable. This possible impacted large calculus is causing compression over extrahepatic system and resulting in wall thickening and dilatation of CD (white arrow), common hepatic duct and intrahepatic ducts. Gall bladder (GB) is shrunken with thick wall favouring chronic cholecystitis.

of CC. This explains that an anomalous biliary and pancreatic ductal junction results in chronic reflux of pancreatic enzymes into the biliary tree. This can result in the weakening of CBD wall followed by the dilatation.^{8,9} According to Todani's classification, the cyst within CBD is due to a focal dilatation of any of its part, and it is then classified as type I-B CC.^{3,4} The stasis of bile within the CC is considered to result in calculi formation. The consistency of calculi can range from inspissated bile to earthy soft to stony hard calculi. Adult CC is usually asymptomatic⁸ and CDC is more frequent condition in adults with CC than in children.¹⁰

Ultrasound can be considered as a reliable method for initial assessment and diagnosis.^{6,11} However, in our case, it could not reveal an accurate situation due to a large CC and calculi resulting in ultrasonic beam attenuation and poor visualisation of adjacent structures and extrahepatic biliary dilatation. The full extent of the cyst, its relation to the biliary tree and the liver can be very well depicted by a non-invasive diagnostic modality, an MRCP, like in our case; it revealed the biliary anatomy with accuracy. However, it was a large CDC which caused biliary system dilatation and compression over the junction of CD and CBD which resulted in mild CD dilatation, rather than an MS resulting in this dilatation.¹² CT imaging is helpful in demonstrating the associated anatomy, helps to rule out any associated abdominal pathology or any neoplastic lesion within the hepatobiliary system. However, it is not helpful in isolation to accurately delineate complex biliary ductal system than MRCP or endoscopic retrograde cholangiopancreatography (ERCP).¹¹

Figure 4 Follow-up ultrasound examination of liver. The transverse section (left side) showing right hepatic lobe with resolution of intrahepatic biliary dilatation. The longitudinal section (right side) shows normal appearing segment VI of liver and right kidney.

In cases of small CDC, ERCP can be of critical importance in retrieving calculi with a less invasive approach than surgery.⁹ However, ERCP was not attempted in our case due to a large calculus and remote chances for therapeutic success. However, ERCP is an overall good option for establishing diagnosis and management of type III CC. It is useful for endoscopic sphincterotomy and long-term follow-up.¹³ The most appropriate treatment option for type I CDC is CC excision followed by bilioenteric anastomosis. Complete choledochal cystectomy and Roux-en-Y hepaticojejunostomy is a recommended treatment for type I CC.¹³ Frattaroli *et al* also reported a kind of similar case of type I CC with a multiple CDC mimicking MS.¹⁴ They opted for laparoscopic management and ultimately converted the surgery for laparotomy and used the same approach of cholecystectomy, choledochal cystectomy and Roux-en-Y hepaticojejunostomy. This treatment approach helps in resolution of obstructive jaundice, avoids chances of developing biliary system inflammation, including cholecystitis, cholangitis and pericholangitis and eradicates the chances of developing extrahepatic biliary malignancy.¹⁵ This case also signifies that single stage excision of large CDC is possible.^{16,17} More treatment options are described for other types of CC. Choledochoduodenostomy is a preferred treatment option for type II CC; however, simple cyst excision with T-tube drainage is also performed for its management.^{18,19} Similarly, Hwang *et al* implemented the same management plan in treating type II CDC masquerading as MS.²⁰

The other treatment options for CDC may include medical treatment with ursodeoxycholic acid (UA) and lithotripsy for calculi in biliary system. UA can be used palliatively in poor candidates for surgery. However, its role as a first-line treatment option is not established.^{6,21} Percutaneous or endoscopic lithotripsy can be effectively used in cases of Caroli's disease with recurrent CDC and hepatolithiasis; these cases can also present with recurrent cholangitis.²¹

Patient's perspective

The team of doctors at Aga Khan University Hospital helped me to recover completely. I was amazed when the doctor asked for a permission to report my case and revealed that it was a rare disease with atypical presentation. I had never imagined before that moment that I would become part of a scientific literature.

Learning points

- ▶ Type I-B CC is a rare entity and may remain silent for a long period of time.
- ▶ Choledochocystolithiasis is a rare complication of type I-B CC and requires meticulous evaluation for proper diagnosis.
- ▶ Magnetic resonance cholangiopancreatography is superior to ultrasound to accurately delineate hepatobiliary ductal system and its congenital anomalies.
- ▶ Choledochocystolithiasis just distal to the CD insertion can result in compression over extrahepatic biliary system and mimic Mirizzi syndrome.

Contributors MBHC diagnosed the case with MA on MRCP. OP was a treating physician and MRK treated the patient and performed surgery. MBHC selected the images and wrote the manuscript with the help of MA. MRK and OP endorsed the manuscript and images. All the authors contributed to the intellectual context and approved the final version.

Competing interests None declared.

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