Case report

Endoscopic management of limy bile syndrome presenting with obstructive jaundice

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SUMMARY

Limy bile syndrome is a rare entity in which there is an excessive precipitation of calcium salts, mainly calcium carbonate in the gallbladder (GB) and to a rare extent in the common bile duct (CBD), making it radiopaque in plain radiographs. Laparoscopic cholecystectomy is safe and effective in patients with limy bile confined to the GB. However, for patients with an extension to the CBD, bile duct exploration with a T-tube placement, endoscopic naso-biliary drainage or endoscopic sphincterotomy (EST) may be warranted. We report a case of limy bile syndrome associated with obstructive jaundice in a young female patient. She was managed successfully with EST followed by laparoscopic cholecystectomy.

BACKGROUND

Limy bile syndrome is a rare disorder in which the gallbladder (GB) and rarely the common bile duct (CBD) are filled with a precipitated calcium salt-rich bile, resulting in opacification of the GB on plain radiographs.^{1 2} The prevalence of this syndrome among patients undergone cholecystectomy for cholelithiasis ranges between 0.1% and 1.7%.^{3 4} There is a female predilection (3:1), and can occur in all ages, however it tends to occur after the age

Figure 2 Abdominal ultrasonography demonstrating a distended gallbladder (GB) with hyperechoic stone measuring 4.4×1.9 cm.

of 40.⁴ Since Churchman described this rare entity in 1911, more than 300 cases have been reported in the literature.¹ However, approximately 7% of case were reported to have limy bile in the GB and the CBD.¹ Although there is no clear consensus for the management of patients with limy bile in the CBD associated with obstructive jaundice, an endoscopic approach followed by cholecystectomy has been reported to be effective.¹

CASE PRESENTATION

A 39-year-old female patient who is not known to have any chronic medical illness presented to the emergency department with a history of epigastric pain, associated with nausea and repeated vomiting for 5 days. She had a history of jaundice and dark urine. She had a history of jaundice in the past and was noticeable during her last pregnancy which was two and half years ago, however she was not investigated. Physical examination revealed a scleral icterus. Her vital signs were as follows: blood pressure 131/78 mmHg, pulse 87 beats/min, temperature 36.8°C, respiratory rate 18 breaths/min and oxygen saturation 99% on room air. Local examination revealed a soft and lax abdomen with no guarding or tenderness. Murphy's sign was negative.

INVESTIGATIONS

Laboratory examination revealed an elevated liver enzymes with a total bilirubin level of 138 $\mu\text{mol/L}$ (3–17 $\mu\text{mol/L}$), direct bilirubin 110 $\mu\text{mol/L}$ (0–3 $\mu\text{mol/L}$), alkaline phosphatase 178 U/L (50–136 U/L), gamma-glutamyl transferase 177 U/L

Figure 1 Abdomen X-ray showing a large well-defined opacity noted in the right upper quadrant.

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Figure 3 Magnetic resonance cholangiopancreatography showing elongated gallbladder with a stone in the fundus, measuring 4.3×2.3 cm. An elongated filling defect is seen in the distal common bile duct measuring 2.3×8 mm, with proximal mild dilatation reaching 7 mm (coronal view).

Figure 4 Gross appearance of a thick paste-like material was found filling the gallbladder.

(9–48 U/L), alanine aminotransferase 260 U/L (12–78 U/L) and aspartate aminotransferase 151 U/L (15–37 U/L). Pancreatic enzymes were within normal limits (lipase 148 U/L (73–393)). Abdominal X-ray revealed a large well-defined opacity noted in the right upper quadrant (figure 1). Abdominal ultrasonography demonstrated a distended GB with hyperechoic stone measuring 4.4×1.9 cm, residing in the non-dependent portion of the GB. The intrahepatic ducts and CBD were not dilated. No sonographic signs of cholecystitis were noted (figure 2).

Magnetic resonance cholangiopancreatography was obtained on the second day of admission and showed an elongated GB measuring 11 cm with a stone in the fundus, measuring 4.3×2.3 cm. An elongated filling defect was seen in the distal CBD measuring 2.3×8 mm in its maximal length, with proximal mild dilatation reaching 7 mm (figure 3).

TREATMENT

The patient was admitted to the hospital. She was kept nothing per mouth (NPO), resuscitated with IV fluids and kept for observation. Her symptoms improved markedly during the hospital course, and her hepatobiliary enzymes were improving slowly. Decision was made to take the patient for an endoscopic retrograde cholangiopancreatography (ERCP) before proceeding with a laparoscopic cholecystectomy. An ERCP and sphincterotomy was performed with no complication. A moderate amount of limy bile was noted at the papilla after sweeping the duct. However, no filling defects were appreciated and no stent was applied. The next day, she underwent a laparoscopic cholecystectomy. The resected GB was incised and a thick paste-like material was found filling the GB (figure 4).

OUTCOME AND FOLLOW-UP

The patient had an uneventful postoperative course and was discharged home on day 1 postoperative. On pathology, the specimen showed thick inspissated bile and the GB showed signs of chronic cholecystitis. The patient was followed in the outpatient department for 4 weeks. She denied any symptoms of jaundice or abdominal pain, and her hepatobiliary enzymes remained within normal range.

DISCUSSION

Limy bile syndrome is an unusual biliary pathology in which the GB and to a lesser extent the biliary tree is filled with a thick paste-like material.¹ As the term indicates, it is associated with precipitation of calcium carbonate within the GB.¹ Since Churchman first described this unusual entity in 1911,² till now the pathogenesis is still not fully understood. Many theories have been proposed in the literature. Partial or complete obstruction of the cystic duct with a subsequent biliary stasis is believed to be a key factor in the development of this syndrome. As a result, precipitation of calcium carbonate, which is present normally within the bile occurs.^{3,4} Inflammation of the GB wall, and alkalinisation of the bile, which usually occurs secondary to obstruction and inflammation, may lead to precipitation and crystallisation of calcium carbonate.^{3,4} Hyperparathyroidism and abnormal calcium metabolism with a resultant high serum calcium have been proposed to play a role in the development of this disorder.³

These cases usually behave like cholelithiasis and have a wide range of consequences. They can be asymptomatic and found incidentally in a plain radiographs, or they may present with symptoms of biliary colic.⁴ Complications such as cholecystitis, and cholangitis can occur.^{1,5} Gilani *et al* reported a case of

Mirizzi syndrome in patient with limy bile.⁶ Nevertheless, these complications seem to be associated with concomitant presence of cholelithiasis.⁷

The presence of limy bile in the CBD is rare with a few reported cases. It usually occurs when a stone in the cystic duct or the GB neck gets dislodged leading to migration of limy bile to the CBD, and the duodenum, with resultant obstructive jaundice and rarely pancreatitis.^{8,9} The presence of limy bile within the intrahepatic biliary tree is extremely rare. It can be attributed to a reflux from the GB or an intrahepatic formation.¹⁰

Precipitation of the radiopaque calcium salts in limy bile leads to opacification of the GB on a plain X-ray, thereby it is considered an important tool for the diagnosis.⁴ It may resemble a normally opacified GB following cholecystogram and has to be excluded before making the diagnosis.⁴ Although ultrasonography is the initial modality of choice for evaluating patients with biliary disease, it is less specific in these circumstances.⁸ Unenhanced CT scan to localise limy bile within the biliary tree have been used.¹⁰ It is considered a helpful tool to exclude other calcifying GB pathologies including porcelain GB.⁴ The diagnosis of limy bile extending to the CBD can be readily made when an opacification identified in both the GB and the CBD.⁸ Exclamation mark sign can be appreciated when a stone coexists ahead of limy bile within the CBD.¹¹

Cholecystectomy is the mainstay treatment for limy bile syndrome, however for patient with commitment limy bile in the GB and the CBD cholecystectomy with CBD exploration and T-tube placement may be warranted.⁸ Endoscopic naso-biliary drainage (ENBD) and ERCP with endoscopic sphincterotomy as

an effective alternative approach.¹ Few reports are found in the literature of spontaneous disappearance of limy bile. It can be attribute to several factors including dissolution within the GB, migration through a fistula or by passage through the normal biliary tract.¹¹ Schwartz and Feuchtwanger successfully demonstrated the gradual passage of limy bile from the GB into the biliary ducts till its complete resolution.¹²

There is no evidence in the current literature correlates between the presence of limy bile with development of GB cancer or cholangiocarcinoma.¹³

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Learning points

- Limy bile syndrome is a rare entity, with a similar clinical spectrum of cholelithiasis.
- The ultimate decision of the best management approach for patients with limy bile syndrome presenting with obstructive jaundice should be made based on the surgeon experience, available expertise, as well as patient-specific factors including the patient's general condition, and the size of the coexisting stone.
- Endoscopic sphincterotomy followed by laparoscopic cholecystectomy is considered an effective approach in selected patients.

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