CASE REPORT

Acute acalculous cholecystitis of an intrahepatic gallbladder causing Mirizzi's syndrome

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SUMMARY

We present the case of a young female with symptoms of biliary colic and a biochemical profile consistent with biliary obstruction. Imaging was suspicious for Mirizzi's syndrome. Intraoperatively, the patient was found to have a complete intrahepatic gallbladder causing common hepatic duct compression with final pathology confirming acute cholecystitis. We review the embryological development of the gallbladder as well as clinical presentation of Mirizzi's syndrome. Special consideration for clinical workup and surgical management is discussed.

BACKGROUND

Biliary anatomy is highly variable. An ectopic gallbladder is a congenital abnormality in which the gallbladder is not located in its usual gallbladder fossa attached to the liver. This occurs in approximately 0.1%–0.7% of people.¹ The most common ectopic locations are beneath the left lobe of the liver, transversely oriented, intrahepatic and retroplaced (retrohepatic or retroperitoneal).² In extremely rare cases, the gallbladder has been positioned within the falciform ligament, suprahepatic below the diaphragm, attached to the abdominal wall and behind the pancreas.² An intrahepatic gallbladder is one partially or completely embedded within the liver.³ The degree to which the gallbladder is embedded within the liver parenchyma

Figure 2 A coronal oblique endoscopic retrograde cholangiopancreatography image shows narrowing of the proximal common bile duct (CBD) with associated upstream biliary dilatation. The completely opacified gallbladder (GB) has an abnormal ovoid shape and is of small volume.

can affect the surgical approach and warrants close attention to defining biliary anatomy both preoperatively and intraoperatively.

Mirizzi's syndrome describes the external compression of the common hepatic duct (CHD) by an obstructed and inflamed gallbladder.⁴ This compression can lead to fistulisation and requires meticulous surgical dissection with *thorough* delineation of biliary anatomy.⁵ We present a case of biliary obstruction with preoperative imaging suggesting Mirizzi's syndrome. Intraoperatively, this was confirmed but unlike most cases where Mirizzi's syndrome is secondary to cholelithiasis, this patient had acute acalculous cholecystitis of an intrahepatic gallbladder causing the CHD obstruction.

CASE PRESENTATION

A 36-year-old woman presented with 1 day of right upper quadrant (RUQ) and epigastric abdominal pain, radiating to the back. There were no other gastrointestinal symptoms. She describes similar episodes in the past decade. Her medical history is significant for gastro-oesophageal reflux for which she takes esomeprazole. She underwent a splenectomy for idiopathic thrombocytopenic

Figure 1 An axial greyscale ultrasound image shows an intrahepatic gallbladder (GB). The GB is surrounded by liver parenchyma over the majority of its circumference, has an abnormal ovoid shape and is of small volume. The GB has a normal wall thickness and no intraluminal gallstones are visualised. Posteriorly, the GB causes compression and stretching of the common bile duct (CBD).

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Figure 3 A coronal oblique endoscopic retrograde cholangiopancreatography image shows placement of a plastic biliary stent performed to relieve the obstruction on the proximal common bile duct. The degree of intrahepatic biliary dilatation has improved and contrast excretion from the biliary system into the duodenum is noted. GB, gall bladder.

purpura. She is a non-smoker, who occasionally consumes alcohol.

Physical examination showed her vital signs to be stable. She was afebrile and with no clinical evidence of jaundice. The RUQ

Figure 4 An axial T2-weighted MRI image of the upper abdomen performed following endoscopic retrograde cholangiopancreatography (ERCP) show the intrahepatic gallbladder (GB). Consistent with the ultrasound findings in figure 1, the GB is noted to be surrounded by liver parenchyma over the majority of its circumference, has an abnormal ovoid shape and is of small volume. A tiny antidependent signal void within the lumen of the GB at the 12 o'clock position is consistent with pneumobilia post-ERCP.

Figure 5 A coronal T2 weighted MRI image of the abdomen demonstrates the intrahepatic gallbladder (GB) as well as the biliary stent. As outlined by the stent, the proximal common bile duct is stretched by the adjacent GB where it abuts its left upper surface from 12 to 3 o'clock.

and epigastrium were tender to palpation without rebound or guarding.

INVESTIGATIONS

Laboratory investigations were significant for elevation in white cell count $15.8 \times 10^9/L$, neutrophils $11.3 \times 10^9/L$, monocytes $1.4 \times 10^9/L$, total bilirubin $21 \mu\text{mol/L}$, alanine aminotransferase 453 U/L and gamma-glutamyltransferase 135 U/L . Ultrasonography showed the gallbladder to have an unusual rounded shape (figure 1). No cholelithiasis, abnormal wall thickness or pericholecystic fluid were seen. Common bile duct (CBD) calibre was normal (3 mm) with mildly dilated intrahepatic bile ducts. After 2 days, the patient was transferred for endoscopic retrograde cholangiopancreatography (ERCP). Cholangiography showed a normal CBD and smooth CHD stricture (figure 2). A sphincterotomy was carried out followed by balloon sweeps of the CBD without evidence of sludge or stones. The extraction balloon could not be advanced beyond the CHD stricture, which was dilated using a Hurricane balloon to 4 mm and a 7 French, 9 cm plastic biliary stent was placed (figure 3). The gallbladder filled with contrast during the procedure and was located adjacent to the stricture, raising the concern for Mirizzi's syndrome. In order to better delineate the CHD stricture, MRI of the abdomen was performed. This showed the gallbladder to have an abnormal rounded configuration but normal wall thickness (figure 4). The gallbladder was exerting mass effect over the proximal CBD, which stretched and draped over the gallbladder anteriomedially. The biliary stent was seen in good position (figure 5). The cystic duct could not be clearly visualised.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included Mirizzi's syndrome, acalculous cholecystitis and benign CHD stricture.

TREATMENT

The patient was taken for an open cholecystectomy. Intraoperatively, the gallbladder was found to be completely intrahepatic and intimately associated with the structures of the porta

Figure 6 Coronal oblique endoscopic retrograde cholangiopancreatography image performed 2 months postcholecystectomy and following prior removal of the biliary stent. The common bile duct (CBD) is normal calibre and the previously noted narrowing at the proximal portion of the CBD has resolved.

hepatitis. The visible portion of gallbladder appeared gangrenous. Meticulous dissection confirmed Mirizzi's syndrome without fistula. The plastic common bile duct stent was easily palpable and assisted in clarifying biliary anatomy. By opening the gallbladder, the depth of intrahepatic extension was appreciated and the gallbladder dissected out of the liver parenchyma.

OUTCOME AND FOLLOW-UP

The patient recovered well after surgery and was discharged home on postoperative day 5. Final pathology of the gallbladder showed acute cholecystitis with benign-associated metaplasia. There was no evidence of cholelithiasis. The patient returned 2 months after her surgery for ERCP. Her previous pain had resolved. The biliary stent was removed and cholangiography showed a normal CBD (figure 6).

DISCUSSION

The gallbladder develops from the distal foregut as part of the hepatic diverticulum in the third week of gestation.⁶ The hepatic diverticulum has two parts: the *pars hepatica*, which develops into liver, common hepatic duct and intrahepatic biliary tree, as well as the *pars cystica*, which forms the gallbladder and cystic duct.⁶ In humans, the gallbladder is initially intrahepatic and moves into its extrahepatic location during the second month of

gestation.⁶ It is therefore thought that an intrahepatic gallbladder signifies developmental positional arrest.⁶ Intrahepatic gallbladders tend to have impaired emptying and as such are at increased risk of pathology.⁷ An intrahepatic gallbladder can alter surgical approach and requires thorough delineation of biliary anatomy. In cases, such as we present here, with the entire gallbladder being intrahepatic, an open approach may be required over a laparoscopic technique. This case was further complicated by Mirizzi's syndrome with compression of the CHD. Preoperative imaging should include an ultrasound as well as cross-sectional imaging such as CT or magnetic resonance cholangiopancreatography. ERCP aided in diagnosis of Mirizzi's syndrome and allowed placement of a plastic biliary stent, which assists intraoperatively with delineation of biliary anatomy and can be useful if indeed a fistula is found.

Learning points

- ▶ Suspect intrahepatic gallbladder location when the gallbladder has abnormal shape or is located high on endoscopic retrograde cholangiopancreatography (ERCP).
- ▶ If abnormal biliary anatomy is suspected on ultrasound, delineate biliary anatomy with cross-sectional imaging such as CT or MRI preoperatively.
- ▶ Consider ERCP and placement of a biliary stent especially when Mirizzi's syndrome is suspected.
- ▶ Intraoperatively, careful dissection of Calot's triangle may be aided by a preoperatively placed biliary stent.

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