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Gallbladder agenesis with a primary choledochal stone in a patient with situs inversus totalis

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 68
Final Diagnosis: Gallbladder agenesis with situs inversus totalis
Symptoms: Epigastric pain • jaundice
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

Background: Situs inversus totalis is an inherited condition characterized by the mirror-image transposition of thoracic and abdominal organs. Gallbladder agenesis, which has normal bile ducts, is a rare congenital condition that occurs in 13 to 65 people out of 100 000. A common bile duct (CBD) stone or choledocholithiasis in patients with gallbladder agenesis is even rarer.

Case Report: We report the case of a 68-year-old woman who presented with epigastric pain and jaundice. She was not known to have situs inversus totalis. Abdominal ultrasound showed a large stone in the CBD, which could not be extracted by endoscopic retrograde cholangiopancreatography (ERCP), necessitating exploration. The gallbladder and cystic duct were found to be absent. Incisional exploration of the CBD was performed, and a large stone was removed. A choledochoscope was used to identify the remnants and exclude the presence of ectopic gallbladder, and a T-tube was placed into the CBD.

Conclusions: Gallbladder agenesis in a patient with situs inversus totalis is extremely rare, with no single reported case identified in the literature. In addition, our case showed a rare complication of ERCP – a failure to extract the CBD stone – and illustrates a way to overcome this complication.

MeSH Keywords: Choledocholithiasis • Gallbladder • Situs Inversus

Full-text PDF: http://www.amjcaserep.com/download/index/idArt/890523

Background

Situs inversus totalis is a rare congenital disorder inherited via an autosomal recessive gene. It is estimated to occur in 1 in 5000 to 1 in 20 000 births according to region [1]. It is characterized by transposition of the major thoracic organs and all the visceral organs in the abdomen to the opposite side of the normal position in the body. Normal development requires a 270 degree counterclockwise rotation. In situs inversus totalis, the 270 degree rotation is clockwise [2]. Agenesis of the gallbladder and cystic duct is a rare congenital condition that results from the failure of the cystic bud to develop in the 4th week of intrauterine life, and it has a reported incidence of 13–65 per 100 000 population [3]. It was first described in 1701 [4]. Choledocholithiasis has been reported to occur in 18% to 50% of patients with gallbladder agenesis, and all have jaundice as the first presentation [5–9].

Here we report a very rare case of a patient with situs inversus totalis with a large choledochal stone and gallbladder agenesis identified during surgery.

Case Report

Our case was a 68-year-old woman with a history of diabetes and dyslipidemia who was on oral treatments. The patient was referred to our hospital from the primary care center for epigastric pain with obstructive jaundice because our hospital serves as the referral center for the central region. During evaluation in the emergency department, she reported a history of on-and-off abdominal pain in the epigastric region during the previous month, which was aggravated by fatty meals. The pain was non-radiating and not associated with an increased body temperature or change of urine or stool color. Physical examination results were normal except for mild jaundice and audible first and second heart sounds, mostly in the right-sided chest. The abdomen was soft and lax, with mild epigastric tenderness and no organomegaly. On laboratory workup, she was found to have WBC of 12.6, hemoglobin of 11.8, platelet count of 471×10^9, total bilirubin of 29.4 µmol/L, direct bilirubin of 19.1 µmol/L, alkaline phosphatase of 720 U/L, GTP of 1151 U/L, and amylase of 47 U/L. Chest X-ray showed dextrocardia (Figure 1). Abdominal ultrasound showed intrahepatic biliary duct dilatation with a maximum diameter of 0.7 cm. The gallbladder was collapsed and contained a stone. The common bile duct was dilated to 1.14 cm and the stone measured 0.7 cm (Figure 2).

The patient was admitted, started on IV antibiotics, and scheduled for endoscopic retrograde cholangiopancreatography (ERCP) on day 2 of hospitalization. ERCP was performed and showed a large CBD stone measuring more than 3 cm

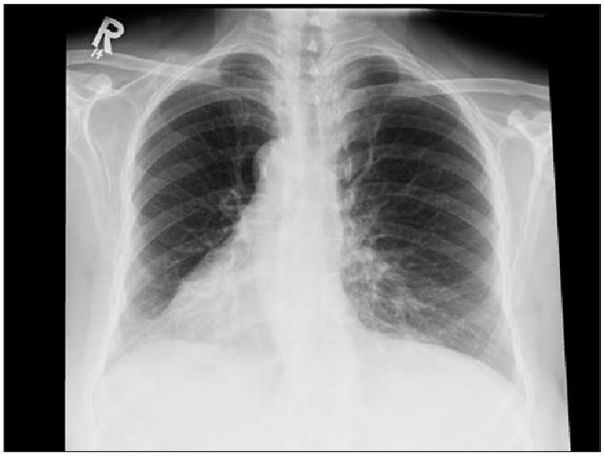

Figure 1. Preoperative chest X-ray, demonstrating dextrocardia.

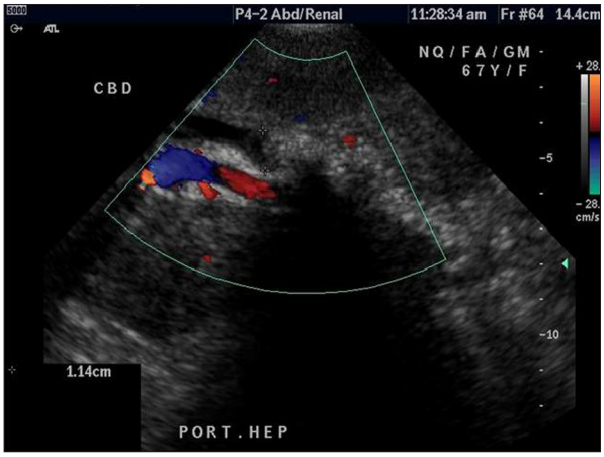

Figure 2. Ultrasound image of the porta hepatis, showing a markedly dilated common bile duct.

(Figure 3). The stone could not be extracted by endoscopy; therefore, stenting of the CBD was performed.

CBD exploration was carried out, and the gallbladder was found to be missing, with only a dilated CBD containing a large stone, which was removed after crushing. A choledochoscope was introduced inside the duct to visualize the presence of any remnant and ensure that there was no ectopic gallbladder connecting to the CBD. Subsequently, a T-tube was placed. A T-tube cholangiogram was performed, which showed no filling defects or leak postoperatively (Figure 4).

The postoperative period was uneventful with no complications and the patient was discharged.

Discussion

Situs inversus totalis may affect intraabdominal viscera and the intrathoracic organs [1]. The first known case of this condition

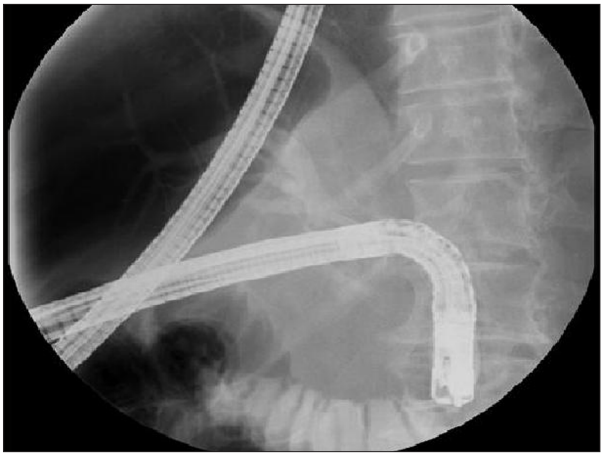

Figure 3. Endoscopic retrograde cholangiopancreatography, confirms the presence of a large stone of the common bile duct.

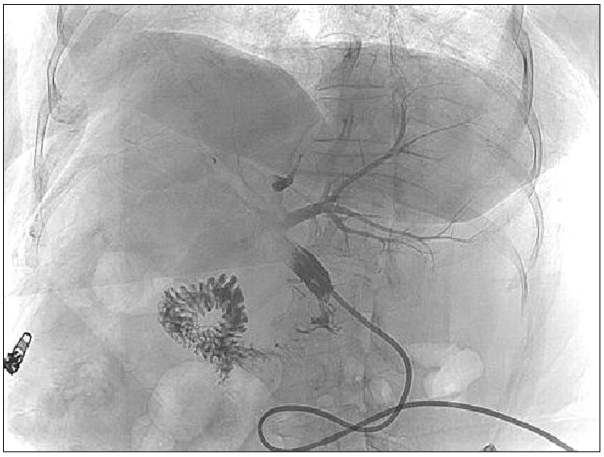

Figure 4. T-tube cholangiogram, placed on the left side of the abdomen.

was reported in 1600 [10]. Approximately one-third of patients with situs inversus and symptomatic gallstones present with epigastric pain, and approximately 10% of patients present with right-sided pain [11].

Hepatobiliary system development begins at around the third week of gestation, when the hepatic diverticulum forms as an outgrowth of the endoderm at the distal end of the foregut. As the diverticulum grows, its connection with the intestines narrows and becomes the extrahepatic bile ducts. A small ventral outgrowth develops and vacuolizes, eventually forming the gallbladder and cystic duct. Failure of this outgrowth results in agenesis of the gallbladder and a cystic duct, without extrahepatic biliary atresia [12]. Agenesis of the gallbladder can present as 1 of 3 categories [3]. Category 1 is associated with multiple fetal anomalies leading to death in the perinatal period from causes related to non-biliary abnormalities. Category 2 is an asymptomatic group in which the anomaly is discovered at either autopsy or laparotomy for unrelated diagnosis. Category 3 is a symptomatic group that undergoes surgery for symptoms of hepatobiliary disease, but no gallbladder is found at surgery. According to this classification, our patient belongs in category 3.

It has been suggested that gallbladder agenesis mirrors the post-cholecystectomy condition [13], which predisposes patients to biliary dyskinesia. Furthermore, this biliary dyskinesia, which occurs from a greater retrograde ampullary sphincteric musculature contraction compared with normal subjects, promotes dilation of the CBD and biliary stasis [14]. This condition predisposes patients to the development of biliary calculi. The preoperative diagnosis of gallbladder agenesis has been considered to be difficult, and most diagnoses are made at laparotomy or during attempted laparoscopic cholecystectomy [15]. The usual ultrasonographic report in cases that were subsequently confirmed to be gallbladder agenesis is a shrunken gallbladder containing gallstones. In case of false-positive ultrasonography of gallbladder agenesis, it has been suggested that either a loop of gas-containing bowel located in the gallbladder fossa or periportal tissue and subhepatic peritoneal folds mimic a shrunken gallbladder containing gallstones [16]. If the diagnosis of gallbladder agenesis is made preoperatively, patients who have choledocholithiasis should undergo ERCP stone extraction. However, in case of failed stone extraction with ERCP, choledocholithotomy followed by T-tube drainage or the placement of an endoscopic retrograde biliary drainage tube performed after primary closure of the CBD is a standard treatment [17].

No single case was identified in an extensive literature review using MEDLINE and other search engines with both English and non-English languages [18].

Conclusions

Gallbladder agenesis in a patient with situs inversus totalis is extremely rare, with no single reported case identified in the literature. In addition, our case showed a rare complication of ERCP – a failure to extract the CBD stone – and illustrates a way to overcome this complication.

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