

Original Article

Successful Stone Extraction From The Common Bile Duct By ERCP In A Patient With Situs Inversus Totalis

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Abstract

ERCP is an invasive procedure for removing common bile duct and pancreatic stones, and the instrument is specially designed for the normal anatomy of intra-abdominal organs. We describe a case in which CBD stones were removed by ERCP in a patient with total situs inversus. ERCP is a challenging procedure, most of the time, even with normal anatomical structures, and is more difficult or impossible with total situs inversus. CBD was assessed as very difficult by an experienced gastroenterologist, and the stones were successfully removed. ERCP was performed by placing the patient in the prone position with the gastroenterologist standing on the normal left side of the table. The duodenoscope was first rotated 180° counterclockwise in the stomach and then shortened by rotating the scope again to 180° clockwise in the duodenum. For the second technique, we assessed the second portion of the duodenum by following the lesser curvature while slowly turning the endoscope clockwise. Papillotomy was performed in a complicated manner, and a stent was successfully placed in the CBD with a free flow of bile.

Keywords: Situs inversus totalis, Endoscopic retrograde cholangiopancreatography (ERCP), Choledocholithiasis.

Introduction

Situs inversus totalis is an autosomal recessive and very rare anatomical problem in which all internal organs are in a mirror image of their normal position; its incidence is approximately 1 in 20,000. The anatomy of the major organs on the left and right sides of the abdomen is reversed; therefore, diagnostic or therapeutic ERCP with sphincterotomy is very difficult to perform in patients with situs inversus compared to patients with normal anatomy. Normally, the patient lies in the left lateral position during ERCP, and the endoscopist performs the procedure standing on the right side of the patient (viewing from the foot end). Several techniques have been tried in patients with situs inversus to perform the procedure, such as the twist method, mirror image method, 180-degree clockwise turn, 360-degree turn, and changing the patient's position to the left lateral decubitus position.⁴ However, our experience has shown that it is possible to perform ERCP in patients with situs inversus without much change in position, although it is performed with a very difficult manoeuvre in the supine position.

Case Presentation

A 65-year-old lady presented with chronic right hypochondrial and epigastric pain for the last 10 days. On examination, she was deeply jaundiced with generalized pruritus. She complained of severe epigastric pain along with tenderness, but Murphy's sign was negative. Serum aspartate transaminase levels were elevated to 181 IU/l, serum alanine transaminase levels were raised to 82 IU/l, and total bilirubin levels were raised to 14.32 mg/dl. The alkaline phosphatase levels were 499 IU/L, and the gamma-glutamyl transferase level was 915 IU/L. CT scan abdomen with contrast showed grossly dilated common bile duct with dilated intrahepatic channels and obvious multiple stones in the common bile duct, and there is malposition of all intra-abdominal organs and vessels called situs inversus totalis. Although a patient with situs inversus totalis was seen on a contrast-enhanced CT scan of the abdomen, before doing ERCP, we did an endoscopic examination of the stomach and duodenum, which showed a reversed anatomy of the entire gastrointestinal system. The patient was put in the normal left lateral position, and the scope was introduced in a normal way with the endoscopist standing on the right side of the patient from the foot end.

The duodenoscope was once introduced into the stomach, rotated 180 degrees counterclockwise, and the pyloric opening was looked for. The scope was passed with difficulty in the duodenum, and after entering the 2nd part, the papilla was identified and rotated clockwise 180 degrees, and short routing was done by pulling the scope after fixing its wheels, leading to adequate visualization of the duodenal papilla. The papilla was found to be bulky, suggesting some impacted stone (Figure 1). As the orifice was not oriented in the normal way according to the scope camera and vision, it was much more difficult to gain proper deep cannulation of the papilla and approach the CBD. Once the guide wire was passed and confirmed to be in the common bile duct, contrast was given, which showed multiple filling defects in the CBD suggestive of stones. The bulky papilla made it relatively easy to do the sphincterotomy. The filling defects were proven to be stones by giving further contrast, and although it was very difficult due to positioning, we tried twice with a balloon sweep to remove the stones, but failed. Our team then unanimously decided to pass the stent, and a plastic 10 French 7 cm stent was placed in the CBD after removing the papillatome, keeping the guide wire in the CBD, and passing the stent over the guide wire. Free flow was seen, and no gross bleeding was noted. The pancreatic duct was neither cannulated nor visualized. After the procedure, the patient was admitted to the ward for a couple

Contributions:

SMC GM - Conception, Design
SMC GM- Acquisition, Analysis, Interpretation
HU - Drafting
KF KHK HU TH - Critical Review

All authors approved the final version to be published & agreed to be accountable for all aspects of the work.

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of days to monitor for any complications. The patient did well with marked improvement in the liver tests and was discharged with regular follow-up in the Gastroenterology OPD.

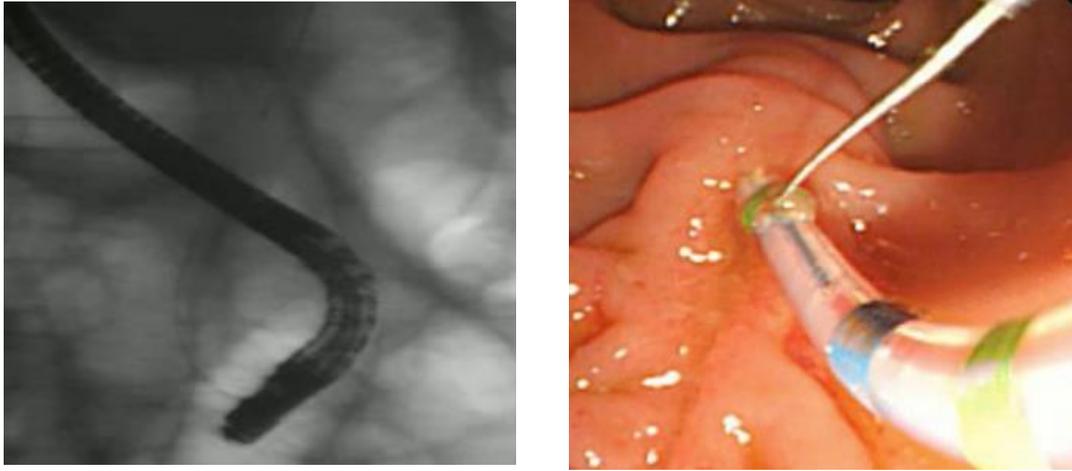


Figure 1:

Discussion

Endoscopic retrograde cholangiopancreatography (ERCP) is widely used worldwide in patients with diseases of the biliary tract and pancreatic abnormalities. Even in patients with normal anatomy, ERCP procedures are challenging due to various presentations of hepato-biliary disease. However, in the presence of anatomical abnormalities, such as total situs inversus, even a skilled endoscopist can encounter technical difficulties while performing this procedure. In the literature, there are only a few reports on the successful performance of ERCP in patients with total situs inversus. Several techniques have been tried for ERCP in these patients, including rotating the endoscope 180° counterclockwise in the stomach and, after entering the duodenum, rotating the endoscope 180° clockwise. A similar technique involves rotating the duodenoscope 180° clockwise in the stomach and using a rotating sphincterotome for cannulation. These methods were performed where the position of the patient, endoscopist, endoscope machine, and monitor were the same as usual, although requiring a skilled endoscopist with abundant experience. In addition, if loop formation occurs during the procedure, the endoscopist may find it difficult to achieve or maintain the position of the scope at the ampulla. Another technique is the “mirror image” method, in which the patient is placed on the table in the right pron position as there is reversal of internal organs and the endoscopist performs this procedure from the left side of the table seen from the foot end; although in this technique shortening of the scope using a counterclockwise rotation is very inconvenient because the endoscopists need and are trained to manipulate the endoscope with the right hand. Moreover, the position of the patient, endoscopist, endoscopic machine, and monitor must all be changed. In another method, “inverted normal” ERCP can be performed where the patient is placed in the prone position, and the endoscopist stands on the right side of the table. The ERCP scope is advanced in the duodenum by following the lesser curvature without rotating the endoscope. Another technique is endoscopic papillary balloon dilation (EPBD) when there is axis malalignment; dilation can be easily achieved once deep cannulation is achieved. EPBD performed with or without endoscopic sphincterotomy (EST) is a useful alternative to endoscopic sphincterotomy alone for the removal of stones from the common bile duct. More complicated ERCP procedures have been reported in the literature in cases of total situs inversus, such as repeated mechanical lithotripsy or Spyglass-guided laser lithotripsy, and common bile duct stone removal in cases of B-II gastrectomy. In cases where there is failure of the procedure in these difficult cases, laparoscopic surgery or percutaneous drainage with or without the rendezvous method has also been reported. Complications of ERCP in patients with situs inversus are rare, according to published cases. Eitler K et al. reported that one case of bleeding from porto-biliopathy during stone removal was successfully treated by a self-expandable metal stent and balloon compression. Our postoperative bleeding event occurred in a patient with decompensated cirrhosis, as the cutting direction was at the opposite side (1–3 o'clock) compared to the usual condition (10–12 o'clock). Whether the vessel distribution around the papilla in a situs inversus condition is also inverted is still unknown.

Conclusions

Situs inversus totalis is a rare congenital condition in which there is visceral left–right asymmetry, which presents with a major challenge to the endoscopist in doing the ERCP procedure. Our case study also found a higher cannulation difficulty and failure under SIT, even in the hands of experts in doing the ERCP procedure. Various modified positions of patients are needed for easier cannulation and a lower chance of complications, although more literature review is required in this type of difficult case. There is also a need for more detailed documentation of the procedural parameters and complications, which will help in developing further standards of care in such cases.

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