Therapeutic ERCP in Patient with Situs Inversus Totalis and Ampullary Diverticulum

Lubna Kamani1, Raj Kumar1, Shahid Mahmood1, Sana Jafri1 and Faisal Siddiqui2

ABSTRACT

Situs Inversus Totalis (SIT) is a rare entity with complete transposition of all viscera, making endoscopic and surgical procedures challenging and complicated. We describe a rare case of a 55 years old man with SIT and ampullary diverticulum presenting with cholangitis and deranged liver function tests due to common bile duct stones. Therapeutic Endoscopic Retrograde Cholangiopancreatography (ERCP) was planned to relieve biliary obstruction and removal of stones. Procedure was started after informed consent in usual left semi-prone position but rotation of scope to 180 degrees and shortening under fluoroscopic guidance was done to attain and maintain desirable ampullary position and cannulation was done with standard sphincterotome followed by sphincterotomy and sphincteroplasty. ERCP was performed successfully despite difficulties of dual pathology and the patient made uneventful recovery.

Key Words: Situs inversus. Ampullary diverticulum. Choledocholithiasis.

INTRODUCTION

Situs Inversus Totalis (SIT) is a rare entity with complete transposition of all viscera, estimated to occur in the range of 1:10,000 to 1:20,000 with a male/female ratio of 3:2.1 The clinical presentation of acute cholangitis secondary to bile duct stone is very rare in patients with SIT.2-4 Duodenal diverticulas are present in up to 25% of patients but rarely cause symptoms. Endoscopic retrograde cholangiopancreatography (ERCP) is the main treatment modality in patients with biliary obstruction but it becomes challenging in SIT patients due to altered anatomy.

We report a case of therapeutic ERCP in patient with dual anatomic pathology, SIT and ampullary diverticulum.

CASE REPORT

A 55-year-old male with known SIT, presented with left upper quadrant abdominal pain. The pain was colicky and associated with nausea, vomiting, fever and jaundice. Physical examination revealed tenderness in the left upper abdominal quadrant. Laboratory studies showed white cell count of 15,500/l; total bilirubin level of 3.5 mg/dl; ALT level of 132 U/l; alkaline phosphatase level of 395 U/dl and gamma-GT level of 1674 U/dl. All other laboratory parameters were in the normal range. Abdominal computed tomography confirmed situs inversus viscerum and dilatation of the intra- and extrahepatic bile duct with common duct stones largest being 0.8 cm in size (Figure 1).

After conscious sedation with diazepam and nalbuphine, ERCP was performed with a side-viewing endoscope (Olympus TJF-H-180). The procedure was started after informed consent in usual left semi-prone position but rotation of scope to 180 degrees and shortening under fluoroscopic guidance was done to attain and maintain desirable ampullary position and cannulation was done with standard sphincterotome followed by sphincterotomy and sphincteroplasty. ERCP was performed successfully despite difficulties of dual pathology and the patient made uneventful recovery.

DISCUSSION

Endoscopic sphincterotomy and stone extraction are standard procedures for the removal of bile duct stones. Therapeutic ERCP for bile duct stones has a success rate of 96 - 100%.5 The cause of SIT is unknown. More than one genetic mutation was implicated in etiopathogenesis.6 SIT increases the technical difficulties and the possibilities of complications of therapeutic interventions. Pathak et al. reported a case of ERCP performed in SIT,7 keeping the patient in the prone position and the endoscopist at the left side, but that procedure was done with rotation of the instrument. In this case, we succeeded to pass the stomach and to reach the duodenum with 180 degrees turn and the correct...
position was maintained throughout the procedure. We also found ampullary diverticulum making the procedure more challenging. To authors’ knowledge there is no data about the concurrence of SIT and ampullary diverticulum in patient with bile duct stones. This case is the first one in literature in terms of dual anatomical pathology.

In patients with SIT and ampullary diverticulum, ERCP and generous sphincterotomy is not only challenging but sometimes technically difficult because of altered CBD anatomy. Sphincteroplasty is safe and effective in removing large biliary stones.8 In expert hands, ERCP can be performed with some changes in scope positions with the help of fluoroscopy.

REFERENCES


