

Endoscopic Retrograde Cholangiopancreatography of Pancreatitis Due to Microlithiasis in a Patient with Situs Inversus Totalis: A Case Report and Literature Review

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ABSTRACT

Situs inversus is a rare congenital disorder characterized by transposition of the major thoracic organs and all the visceral organs in the abdomen to the opposite side of the normal place in the body. Herein we present a 73-year-old woman with a history of situs inversus totalis who was referred with the complaints of abdominal pain caused by pancreatitis and large common bile duct due to choledochal sludge. Laboratory tests of serum amylase, aspartate aminotransferase, alanine aminotransferase, and alkaline phosphatase were 960 IU/L, 207 IU/L, 119 IU/L, and 263 IU/L, respectively. The radiographic evaluation confirmed situs inversus totalis, enlarged liver, and dilatation of the intra- and extrahepatic bile duct. The distal end of the common bile duct (CBD) was smoothly tapered. Endosonography also confirmed CBD microlithiasis. Endoscopic retrograde cholangiopancreatography (ERCP) was done to remove microlithiasis. Our Challenge was on ERCP techniques in the position of the patient and endoscopy team. We believe ERCP can be safely performed in usual position with minor modification.

Keywords: Endoscopic retrograde cholangiopancreatography, Situs inversus, Pancreatitis, Microlithiasis

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INTRODUCTION

Situs inversus is a rare congenital disorder inherited via an autosomal recessive gene(1). Its first description was in 1701, which led to a reversal of right-left polarity. Situs inversus is recognized as congenital aberrations resulted from abnormalities of deletions on chromosomes 7 and 8(2). The disease 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. It is estimated to occur in 1 in 5000 to 1 in 20,000 births according to different regions(3).

Subdivisions of Situs inversus are related to the position of heart apex;

The posture of the apex in the left side is called levocardiapex and in the right side dextrocardiapex (4). Although cardiac disorders are more common in the general population of situs inversus patients. Cardiopulmonary Bypass (CPB) is found in some patients with this congenital malformation. In addition, Patent Ductus Arteriosus (PDA) ligation may occur before CPB. In this regard mitral valve regurgitation, PDA and another malformation and critical heart disease occurred in this patients.

Some previous studies about gastrointestinal interventions in these patients indicate that ERCP

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Table 1: Laboratory findings in blood parameters

Blood test Parameter	Result	Normal Range
Total white blood cell	22,120 K/mm ³	3.8 – 11 K/mm ³
total bilirubin	3.63 mg/dL	up to 1.4 mg/dL
direct bilirubin	0.57 mg/dL	up to 0.4 mg/dL
amylase	960 IU/L	111 – 296 IU/L
aspartate aminotransferase (AST/ASAT)	207 IU/L	6 -34 IU/L
alanine aminotransferase (ALT/ALAT)	119 IU/L	5 -56 IU/L
alkaline phosphatase (ALP)	263 IU/L	42 -98 IU/L
γ-glutamyl transpeptidase	2,132 IU/L	5 – 78 IU/L
partial thromboplastin time	35 Sec	25 – 43 Sec
prothrombin time	12.8 Sec	12.5 Sec
prothrombin activity	93%	98 - 100
I.N.R	1.1	2-3
PT Control	12 Sec	12 Sec

can be performed successfully despite difficulties with routine maneuvers. So endoscope can be rotated clockwise to 180 degrees with some torsion and shortening(3).

The liver and gallbladder are located on the left, while the stomach and the spleen are on the right. The normal development requires a 270-degree counterclockwise rotation to yield the normal anatomy. In situs inversus totalis, the 270-degree rotation is in the clockwise direction(7,8). Conventionally, reversal of the position of the endoscopist in relation to the patient is advocated to overcome the anatomical difficulty(9).

Here we report a very rare case of situs inversus totalis with pancreatitis and large common bile duct caused by choledochal sludge.

CASE REPORT

A 73-year-old woman with a history of situs inversus totalis referred to our hospital with the complaints of abdominal pain. General appearance was acutely ill and there was tenderness on the left upper quadrant of the abdomen. The intensity of symptoms increased before hospital admission. The symptoms were accompanied by mild fever, nausea, and vomiting. Physical signs were: temperature: 38.5°C, blood pressure: 140/90 mmHg, icterus, abdominal tenderness in left hypochondrium, non-palpable liver and spleen, and few regional ascites.

Laboratory findings

Total white blood cell: 22,120/mm³, total bilirubin:

3.63 mg/dL, direct bilirubin: 0.57 mg/dL, amylase 960 IU/L, aspartate aminotransferase: 207 IU/L, alanine aminotransferase: 119 IU/L, alkaline phosphatase: 263 IU/L, γ-glutamyl transpeptidase: 2,132 IU/L, partial thromboplastin time: 35 Sec, prothrombin time: 12.8 Sec, prothrombin activity: 93%, and international normalised ratio: 1 (table. 1).

Echocardiographic Report

There was a history of occasional retrosternal chest pain, with no radiation and angina. On examination, she was pale, had a regular pulse, with average volume. Her jugular venous pulse (JVP) was normal.

Also, there were an aortic valve and ventricular valve concordance. Normal left ventricular size with mild left ventricular systolic dysfunction was reported, and left ventricular ejection fraction was 45%. The heart septum is hyperkinesia. The left ventricular have mild diastolic dysfunction. Left atrium had an enlargement. Normal mitral valve with no sign of stenosis was detected; however mild mitral valve regurgitation was reported.

Echocardiography showed tick aortic valve with the suspicion of bicuspid aortic valve disease. Moderate aortic insufficiency with no sign of aortic stenosis was detected. Mild insufficiency and regurgitation were detected in the pulmonary and tricuspid valves. Systolic pulmonary artery pressure was 30 mmHg.

Imaging

Abdominal computed tomography (CT)

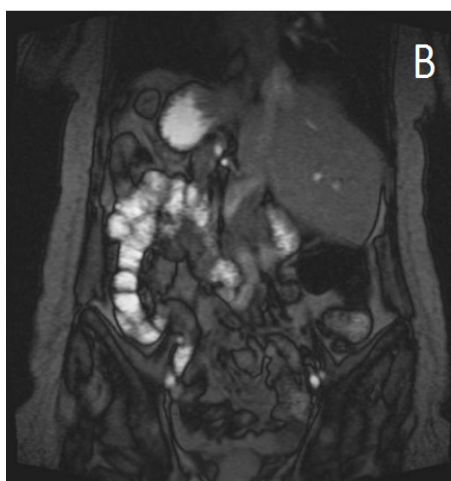
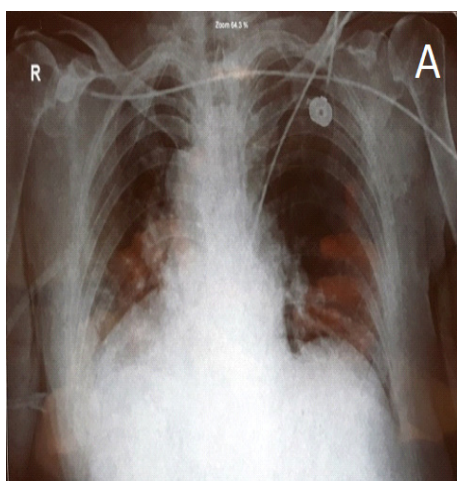


Fig.1: (A): Radiography of thoracic part of the patient (B): An abdominal computed tomography revealed transposition of the visceral organs from the right to left side and microlithiasis in the dilated common bile duct.

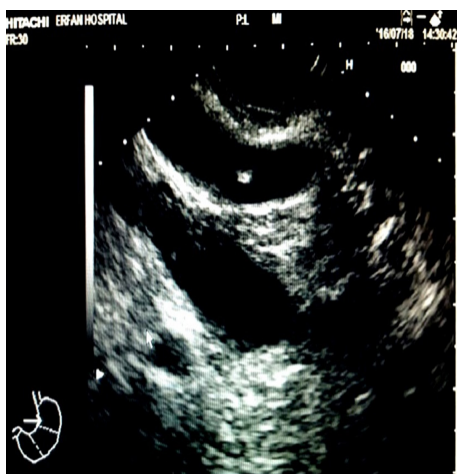


Fig.2: EUS indicate Gallbladder (GB) had hydrops with some small layered gall stone.

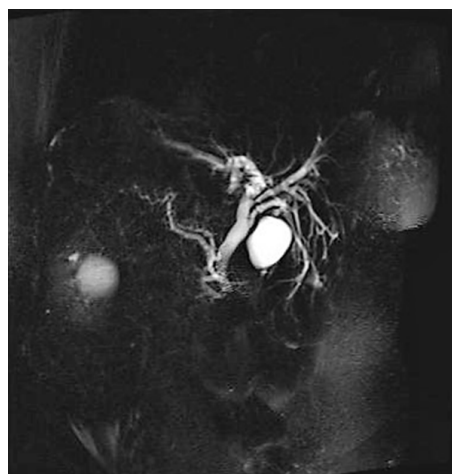


Fig.3: After biliary cannulation using a catheter with a straight end at the 7 O'clock direction of major papilla, a guidewire was placed across the ampullary orifice. Following endoscopic papillary balloon dilatation using a controlled radial expansion balloon (10 mm), a common bile duct microlithiasis was successfully retrieved using a basket.

without contrast (figure 1), magnetic resonance cholangiopancreatography (MRCP), and endosonographic (EUS) (figure 2) confirmed situs inversus totalis. The liver was normal, and dilatation of the intra- and extrahepatic bile duct was detected. The distal end of common bile duct (CBD) had smooth trapper and sludge. Gallbladder (GB) had hydrops with some small layered gall stone. Also, few regional ascites were seen in EUS (figure 2).

EUS indicate Gallbladder (GB) had hydrops with some small layered gall stone.

Mild bilateral pleural effusion and lung bases parenchymal bands (or subsegmental atelectasis) and some umbilical hernia were also noted by CT (figure 3). MRCP showed a tapered stricture of the common bile duct with upstream dilation of the bile ducts.

MRCP reported the hepatic bile ducts and pancreatic duct were dilated (CBD=11mm, PD=4). The GB was also distended and had a smooth border and homogenous content (figure 1).

EUS showed that CBD was dilated (11 mm) and had some microlithiasis. There was also a GB stone



Fig.4: Endoscopic retrograde cholangiopancreatography (ERCP) was performed with a cap-assisted forward-viewing endoscope in the patient.



Fig.5: A cholangiogram revealed transposition of the pancreatic duct oriented to the right side and the gallbladder and dilated common bile duct with a floating microlithiasis to the left side.

(10 mm). The pancreas was heterogeneous and coarse, probably due to recent pancreatitis. Any mass or tumoral lesion was reported in any types of imaging (figure 2).

Endoscopic Retrograde Cholangiopancreatography (ERCP)

After sedation with diazepam and nalbuphine, ERCP was performed with a side-viewing endoscope (Olympus TJF-H-180, Tokyo, Japan). The procedure was started while the patient was in the usual prone position. Duodenoscope was turned 180° clockwise with long loop cross the stomach and the scope was introduced to a 2nd part of the duodenum (D2). In the second part of the duodenum, the endoscope was rotated clockwise to 30 degrees without torsion under fluoroscopic guidance (figure 5). In this regard with regular sphincterotome we cannot perform cannulation, Moreover, after exchanging the sphincterotome to standard rotatable sphincterotome (Autotome™ Cannulating Sphincterotome; Boston Scientific, Cork, Ireland) deep cannulation was done.

Papilla major was also prominent after completed sphincterotomy. CBD microlithiasis was successfully retrieved using a balloon (Extractor™, Boston Scientific, Cork, Ireland) also, some microlithiasis were extracted. Cholangiography showed dilatation of the biliary ducts without filling defects and tumoral lesions (figure 4).

DISCUSSION

Situs inversus totalis is a term used to describe

a condition in which organs or organ systems are transposed from their normal sites to locations on the opposite side of the body. The incidence of this disease is approximately 1:20000 (10), and it is a genetic predisposition with the inheritance of autosomal recessive mode. More than one genetic mutation including gene mutations that cause ciliopathy and cystic renal diseases were implicated in the etiopathogenesis of the disease (11,12). Situs inversus totalis is associated with various gastrointestinal abnormalities, intestinal malrotation, and also acute appendicitis. Liver transplantation has been successfully performed in a patient with juvenile biliary atresia and Situs inversus totalis (10,13).

Some reports indicate that etiology of acute pancreatitis remains idiopathic in 30% of the patients. Following a single idiopathic attack of pancreatitis and a negative EUS examination, relapse is infrequent (14). However, this evidence suggests that EUS may be a useful, minimally invasive tool for the diagnostic evaluation of all pancreatitis (14,15). In addition, MRCP, a non-invasive and complication free imaging modality is able to establish the cause of acute pancreatitis in patients in whom the diagnosis of idiopathic pancreatitis has been made following standard investigations (16). In this case, we used both techniques to find the cause of pancreatitis.

During ERCP to avoid the technical difficulty caused by anatomical right-left reversal, it has been recommended that the endoscopist stands on the right side of the bed and patient is positioned in a right

lateral decubitus position. Performing a technically demanding procedure from an unaccustomed position is often uncomfortable(10). The cutting direction of sphincterotome in Y-shaped endoscopic sphincterotomy (EST) is sometimes technically difficult to control, particularly in patients with specific and difficult anatomy including situs inversus. In contrast to EST, Endoscopic biliary drainage (EBD) is easy to handle in the above condition and it is a safe procedure in less experienced hands (17-19).

Moreover, in patients with altered anatomy (e.g., periampullary diverticulum, Billroth II anatomy), performing an "adequate" biliary endoscopic sphincterotomy is technically difficult or impossible and is associated with a higher risk of complications such as bleeding and perforation (20).

In previous studies, Use of large-balloon dilation in ERCP recommended in a patient with the difficult anatomy of the biliary system (21-24).

Considering many previous studies we used large balloon dilatation (EPBD) with controlled radial expansion (12-15 mm). A recent study suggested that reversal of the position of the endoscopist in relation to the patient is advocated to overcome the anatomical difficulty (10,25). Other studies suggest that mirror image of normal position can help during ERCP(26). In contrast to some previous findings, our experience in this study suggested that normal position of endoscopist in relation to the patient is more predominant.

In the first report of using ERCP by Venu and colleagues in 1985, after intubation, their patient was moved into several positions. In this way, the papilla could be identified and cannulation could be achieved with the endoscope in a straightened position. Some authors have reported similar cases in which ERCP was performed in other positions. In 1988, Nordback and co-workers successfully completed ERCP in patients with situs inversus and the authors found that the best position for cannulation was achieved when the endoscopist turned his right side towards the patient (27). CBD stones were observed and extracted with a Dormia basket after an endoscopic papillotomy was made toward the direction of "1 O'clock". In 1997, Chowdhury and colleagues reported that a patient with situs inversus was placed in the usual left lateral position and the endoscopist was at the left side of the table at the beginning of ERCP (9). The endoscope was shortened by a 90° twist to the left and withdrawn. Subsequently, standard EST was performed successfully. Fiocca and co-workers[10]

that ERCP was carried out successfully despite situs inversus requiring maintenance of the patient in a prone position with the endoscopist at the right side of the table. García Fernández and colleagues performed ERCP in a patient with complete situs inversus using a "mirror image" technique (28). Some studies reported in 2010 and 2014 indicated that a patient with situs inversus was placed in the usual prone position and the duodenoscope was turned 180° clockwise in the stomach. EST and removal of CBD stones were then performed (3,26,29). The case reported by Kamani and co-workers was similar to ours because this previous case also showed the same therapeutic ERCP pattern as patients with situs inversus and an ampullary diverticulum.

Lipschutz's and study indicate for first time the laparoscopic cholecystectomy and ERCP in the situs inversus patient. However, in recently evidences present procedure was 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 (30,31).

In 2003 Yamaguchi reported repeated attempted for stone removal during ERCP by semi prone position (32). In this study we showed the management of biliary microlithiasis by radial expansion balloon after sphincterotomy in Situs inversus totalis by (prone) normal position. No complications such as bleeding, pancreatitis (after ERCP), or perforation was detected.

REFERENCES

1. Arnold G, Bixler D, Girod D. Probable autosomal recessive inheritance of polysplenia, situs inversus and cardiac defects in an Amish family. *Am J Med Genet* 1983;16:35-42.
2. Warwicker P, Goodship TH, Donne RL, Pirson Y, Nicholls A, Ward RM, et al. Genetic studies into inherited and sporadic hemolytic uremic syndrome. *Kidney Int* 1998;53:836-44.
3. Kamani L, Kumar R, Mahmood S, Jafri S, Siddiqui F. Therapeutic ERCP in a patient with situs inversus totalis and ampullary diverticulum. *J Coll Physicians Surg Pak* 2014;24:365-6.
4. Macafee D, Armstrong D, Hall R, Dhingsa R, Zaitoun A, Lobo D. Pancreaticoduodenectomy with a "twist": the challenges of pancreatic resection in the presence of

- situs inversus totalis and situs ambiguus. *Eur J Surg Oncol* 2007;33:524-7.
5. Grey DP, Cooley DA. Dextrocardia with situs inversus totalis: cardiovascular surgery in three patients with concomitant coronary artery disease. *Cardiovasc Dis* 1981;8:527-30.
 6. Cobiella J, Muñoz C, Arís A. Complex cardiac surgery in a patient with dextrocardia and polysplenia. *Rev Esp Cardiol* 2005;58:1236-8.
 7. Topuz O, Sozen S, Agachan AF, Tukenmez M, Vurdem UE. Laparoscopic cholecystectomy in a patient with situs inversus: Advantage of 30° scope. *Pak J Med Sci* 2011;27:929-31.
 8. Song JY, Rana N, Rotman CA. Laparoscopic appendectomy in a female patient with situs inversus: case report and literature review. *JSLs* 2004;8:175-7.
 9. Chowdhury A, Chatterjee B, Das U, Dutta P, Dhali G, Banerjee P. ERCP in situs inversus: do we need to turn the other way? *Indian J Gastroenterol* 1997;16:155-6.
 10. Mohanprasad V, Patil P, Prasad M, Shanker GK. ERCP in Situs Inversus can be Performed in Usual Left Lateral Position. *J Dig Endosc* 2012;3:42-4.
 11. Katsuhara K, Kawamoto S, Wakabayashi T, Belsky JL. Situs inversus totalis and Kartagener's syndrome in a Japanese population. *Chest J* 1972;61:56-61.
 12. Bajwa SJS, Kulshrestha A, Kaur J, Gupta S, Singh A, Parmar SS. The challenging aspects and successful anaesthetic management in a case of situs inversus totalis. *Indian J Anaesth* 2012;56:295.
 13. Tutiven JL, Sundararaman LV, Sarah G. Anatomy and physiology Brain, fontanelles, cranial sutures, and spinal cord. *Essent of Pediatr Anesth* 2014.
 14. Wilcox CM, Seay T, Kim H, Varadarajulu S. Prospective Endoscopic Ultrasound-Based Approach to the Evaluation of Idiopathic Pancreatitis: Causes, Response to Therapy, and Long-term Outcome. *Am J Gastroenterol* 2016;111:1339-48.
 15. Choudhary NS, Bansal RK, Shah V, Nasa M, Puri R, Thandassery R, et al. Prospective evaluation of yield of endoscopic ultrasonography in the etiological diagnosis of "idiopathic" acute pancreatitis. *J Dig Endosc* 2016;7:133.
 16. GN Y, Amin WG, Shaheen FA, Zargar S, Javid G. The efficacy of magnetic resonance cholangiopancreatography in assessing the etiology of acute idiopathic pancreatitis. *Int J Hepat Pancre Dis* 2014;4:32-9.
 17. Rabenstein T, Ruppert T, Schneider HT, Hahn EG, Ell C. Benefits and risks of needle-knife papillotomy. *Gastrointest Endosc* 1997;46:207-11.
 18. Sherman S, Hawes RH, Nisi R, Lehman GA. Endoscopic sphincterotomy-induced hemorrhage: treatment with multipolar electrocoagulation. *Gastrointest Endosc* 1992;38:123-6.
 19. Gholson CF, Favrot D, Vickers B, Dies D, Wilder W. Delayed hemorrhage following endoscopic retrograde sphincterotomy for choledocholithiasis. *Dig Dis Sci* 1996;41:831-4.
 20. Mugica F, Urdapilleta G, Castiella A, Berbiela A, Alzate F, Zapata E, et al. Selective sphincteroplasty of the papilla in cases at risk due to atypical anatomy. *World J Gastroenterol* 2007;13:3106-11.
 21. Maydeo A, Bhandari S. Balloon sphincteroplasty for removing difficult bile duct stones. *Endoscopy* 2007;39:958-61.
 22. Yoo B, Kim J, Jung J, Hwang J, Kwon H, Kim H, et al. Large balloon sphincteroplasty along with or without sphincterotomy in patients with large extrahepatic bile duct stones-multi center study. *Gastrointest Endosc* 2007;65:AB97.
 23. Attasaranya S, Cheon YK, Mchenry L, Sherman S, Schmidt S, Watkins JL, et al. Large-diameter papillary balloon dilation to aid in endoscopic bile duct stone removal: a multicenter series. *Gastrointest Endosc* 2007;65:AB214.
 24. Espinel J, Pinedo E, Olcoz J. [Large hydrostatic balloon for choledocolithiasis]. *Rev Esp Enferm Dig* 2007;99:33-8.
 25. Diverticulectomy Le. 13th International Congress of the European Association for Endoscopic Surgery and other Interventional Techniques (EAES) Venice Lido, Italy, 1-4 June 2005 Video presentations. *Surg Endosc* 2006;20:S225-S50.
 26. Lee JH, Kang DH, Park JH, Kim MD, Yoon KT, Choi CW, et al. Endoscopic removal of a bile-duct stone using sphincterotomy and a large-balloon dilator in a patient with situs inversus totalis. *Gut Liver* 2010;4:110-3.
 27. Nordback I, Airo I. ERCP and endoscopic papillotomy in complete abdominal situs inversus. *Gastrointest Endosc* 1988;34:150.
 28. García-Fernández F, Infantes J, Torres Y, Mendoza F, Alcazar F. ERCP in complete situs inversus viscerum using a "mirror image" technique. *Endoscopy* 2010;42:E316-E7.
 29. De la Serna-Higuera C, Perez-Miranda M, Flores-Cruz G, Gil-Simón P, Caro-Patón A. Endoscopic retrograde cholangiopancreatography in situs inversus partialis. *Endoscopy* 2010;42:E98.
 30. McDermott J, Caushaj P. ERCP and laparoscopic cholecystectomy for cholangitis in a 66-year-old male with situs inversus. *Surg Endosc* 1994;8:1227-9.
 31. Lipschutz JH, Canal DF, Hawes RH, Ruffolo TA, Besold M, Lehman GA. Laparoscopic cholecystectomy and ERCP with sphincterotomy in an elderly patient with situs inversus. *Am J Gastroenterol* 1992;87:218-20.
 32. Yamaguchi Y, Sugiyama M, Sato Y, Mine Y, Yamato T, Ishida H, et al. Annular pancreas complicated by carcinoma of the bile duct: diagnosis by MR cholangiopancreatography and endoscopic ultrasonography. *Abdom Imaging* 2003;28:381-3.