Successful Endoscopic Treatment of a Pancreatic Pseudocyst in a Patient with Situs Inversus Totalis and Upper GI Duplication

Patient: Female, 64-year-old

Final Diagnosis: Situs inversus totalis and doubling of the esophagus, stomach, and first part of the duodenum

Symptoms: Epigastric abdominal pain

Clinical Procedure: Cystogastrostomy with stenting • esophagogastroduodenoscopy

Specialty: Anatomy • Gastroenterology and Hepatology

Objectives:

Congenital defects/diseases

Background:
Duplication of the gastrointestinal tract is a rare congenital malformation that can develop in any part of the digestive tract. These duplications may be asymptomatic into adult age. Situs inversus totalis is a rare congenital anomaly characterized by a mirror transposition of thoracic and abdominal organs. We present a case of a pancreatic pseudocyst in a patient with a combination of situs inversus totalis and doubling of the esophagus, stomach, and first part of the duodenum.

Case Report:
A 64-year-old woman presented with epigastric pain. Abdominal computed tomography revealed a pancreatic pseudocyst and a previously identified duplication of the esophagus, stomach, and duodenum with situs inversus totalis. The patient underwent esophagogastroduodenoscopy (EGD) with endoscopic ultrasonography for pseudocyst drainage. During EGD, a bifurcation of the esophagus was found. Duplication of the esophagus, stomach, and first part of the duodenum was evident on further advancement. A week later, there was repeated filling of the pseudocyst with a liquid component, and the patient underwent cystogastrostomy with stenting. Five months after discharge, the stent was removed without complications.

Conclusions:
Duplication of the gastrointestinal tract and situs inversus totalis are very rare congenital malformations that require early diagnosis. While situs inversus totalis does not represent any medical disadvantage, physicians should be aware of abnormal anatomy before procedures to prepare specialists for this in case of the need for special techniques. Endoscopic treatment of pancreatic pseudocysts is safe and effective even in such rare cases. The use of endoscopic methods also minimizes intervention and decreases the length of the patients' stays in the hospital.

Keywords:
Congenital Abnormalities • Duodenum • Endoscopy, Gastrointestinal • Esophagus • Pancreatic Pseudocyst • Stomach

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Background

Duplication of the gastrointestinal tract is a rare congenital malformation that can develop in any part of the digestive tract. It can manifest itself in the newborn period in the form of obstruction in the abdominal cavity or may be asymptomatic. This condition differs from other intra-abdominal cystic lesions by the presence of a normal mucous membrane of the gastrointestinal tract [1]. Duplications can be noncommunicating or communicating with the normal gastrointestinal lumen, with the latter having the possibility of causing more clinical symptoms, such as constipation or obstruction, due to the connection with the normal tract [2]. The incidence of duplication of the gastrointestinal tract is estimated to be 1 in 4500, more often in men [3]. Although the exact cause of the doubling of the digestive tract is unknown, it is believed that it occurs between the fourth and eighth weeks of pregnancy as a result of partial or abortive twinning, split notochord theory, diverticular and canalization defects, or intrarterine vascular accidents [4,5].

Heterotaxy is defined as abnormal arrangement of intrathoracic organs across the left-right axis of the body. Different types of heterotaxy can be present, which include dextrocardia when involving the heart and other intrathoracic organs, and situs solitus when involving only the heart [6]. Situs inversus totalis is a rare congenital anomaly characterized by a complete mirror transposition of the thoracic and abdominal organs [7]. The incidence of situs inversus itself ranges from 1:6500 to 1:25000, but the real frequency is difficult to estimate as many of these patients are asymptomatic. A number of genes expressing asymmetry may be linked to the development of this process [8].

Pancreatic pseudocysts are fluid-filled cavities arising from the pancreas, often with fibrous or inflammatory tissue and secondary to acute or chronic pancreatic pathologies [9]. Endoscopic drainage is the mainstay of treatment and is completed through a transpapillary or transmural approach, sometimes with cystogastrostomy stenting [10]. Patients with the aforementioned anatomical abnormalities can initially seem difficult to treat in these instances, but this may not always be the case. We present a case unlike any we were able to find in the literature, of a patient with asymptomatic situs inversus totalis, with duplication of the esophagus, stomach, and first part of the duodenum. The duplication was incidentally found through a case report.

Case Report

A 64-year-old woman presented to O.O. Shalimov National Institute of Surgery and Transplantology with a chief complaint of pain in the epigastrium for 1 week without any other associated symptoms. She denied any history of smoking or alcohol use. The patient’s past medical history consisted of chronic gallstone disease, for which cholecystectomy was planned in the future. During a recent hospitalization for COVID-19, a computed tomography (CT) scan incidentally diagnosed situs inversus totalis and duplication of the esophagus, stomach, and first part of the duodenum. The patient had no surgical history. On this admission, CT of the abdomen confirmed the anatomic abnormalities, and acute pancreatitis was suspected (Figure 1). Necrosis of a large part of the pancreatic parenchyma along with formation of a pseudocyst with free liquid in the paragastric parapancreatic tissues was evident. The patient’s gallstone disease was also noted in the scan, which was likely the cause of the pancreatitis. Lab results were significant for marked leukocytosis and increased amylase; CA 19.9 was negative.

The patient underwent esophagogastroduodenoscopy (EGD) with endoscopic ultrasonography (EUS) for drainage of the pseudocyst. During EGD, a bifurcation of the esophagus was found 26 cm distal from the incisors (Figure 2). The endoscope was advanced into the anterior lumen with easy passage into the associated stomach and duodenum. The associated posterior stomach was a deformed tubular structure, and 2 polyps, both 0.2-0.3 cm in diameter, were identified in the antral portion. The lumen narrowed on advancement into the duodenum but expanded once the scope was postbulbar. During inversion of the scope at this point, the opening to the anterior duodenal section was seen (Figure 3). A schematic of the patient’s upper gastrointestinal tract anatomy is shown in Figure 4, which was developed using the CT and endoscopy results.
The EUS of the pancreas showed a pseudocyst with a size of 80×78 mm. The anterior stomach was used due to proximity to the pancreas. Ultrasound-guided drainage of the pseudocyst was completed; 200 mL of fluid was collected, and the pseudocyst was completely drained with hemostasis following puncture and drainage. Fluid cytology revealed no dysplasia and CA 19-9 in aspirate was within normal limits.

On follow-up a week later, the patient had similar epigastric pain without other symptoms. An endoscopic ultrasound at this time revealed repeat fluid accumulation in the pseudocyst, and a decision was made to undergo endoscopic cystogastrostomy with stenting due to the refractory nature of the cyst. The patient’s abnormal anatomy did not prove to increase the difficulty of stent placement, and the same stomach that was used for original drainage was used for this procedure. A double pigtail stent was successfully placed at the middle third of the stomach closest to the cyst, endoscopically, and subsequent drainage was effective. An endoscopic method was selected due to the success with prior endoscopy with drainage. The postoperative period was uneventful and the patient was discharged from the hospital. During follow-up 5 months after discharge, the patient had no complaints and the stent was removed without complications.

**Discussion**

Situs inversus totalis and duplication of the gastrointestinal tract are both isolated, rare congenital abnormalities. Although they can cause symptoms early in life, it is possible for each condition to go undetected until other pathology is present. In our patient’s case, throughout her life, she never presented with any gastrointestinal-related symptoms unrelated to her gallstone disease that would have alluded to her anatomical abnormalities. For unknown reasons, her distinct abnormalities represent a type that seems to not cause obstruction or...
other pathologic gastrointestinal manifestations. Additionally, the patient’s family history was unremarkable.

Situs inversus can be suspected after careful examination, but widespread availability of medical imaging and routine screening programs allow confirmation of the findings and search for additional details and pathology. Standard diagnostic methods, such as x-ray or ultrasound, are usually the first choice and allow for visualization of typical signs, such as dextrocardia, inversion of the location of the liver and spleen, and sometimes even polysplenia [11-13]. More advanced diagnostic methods such as MRI and CT can be used for a more detailed study of this anomaly and other pathological findings, of which CT was used in our case. To perform EGD in these patients, a “mirror technique” is required, where the patient is placed in the right lateral position and the equipment placed mirror-like in the endoscopic operating room [14,15]. There are few reports on the use of EUS for diagnostic and therapeutic procedures in patients with situs inversus totalis, and during the procedure the position of the patient and the echoendoscope were changed [16]. A successful EUS-guided transgastric biliary drainage is described in the literature as well, performed through a solid mass of the pancreatic isthmus involving the common bile duct, duodenum, portal vein, and mesenteric axis causing duodenal stenosis [17]. In the case we present, the procedure was successfully completed in this fashion as well.

Duplication of the gastrointestinal tract is another pathology that can be diagnosed by imaging. According to the opinion of most authors, the main method of diagnosis remains CT and MRI, as they provide the most reliable information about the localization of the duplication of the organ and its relationship with other adjacent anatomical structures [18]. CT proved to be useful for the detection of our patient’s duplication, and the subsequent EGD confirmed the findings. Almost 67% of cases of stomach duplication are detected in the first year of life [19]. This is in contrast to our patient, in which the duplication was only evident when she required imaging for her respiratory illness. Treatment of patients with duplication of the gastrointestinal tract is surgical in symptomatic cases. The majority of duplications occur solely in the ileum, and duplications involving the stomach are very rare, accounting for only 2-8% of gastrointestinal duplications [20]. There are cases of doubling of the stomach causing a gastric tumor, but there are no guidelines or recommendations for prophylactic surgical treatment. The majority of the duplications described in the literature are cystic in nature and complete duplication of any part of the gastrointestinal tract is extremely rare; a review of the literature only yielded 6 case reports. In our case, the patient’s duplication of 3 sections of the gastrointestinal tract, along with situs inversus totalis, represents a truly unique case. The treatment of an acute pancreatic pseudocyst even with such rare and complicated anatomy with endoscopy illustrates the safety and effectiveness of the technique.

Conclusions

Situs inversus totalis and duplication of the gastrointestinal tract are both very rare congenital malformations that can go unnoticed in patients due to the abnormalities not always causing symptoms. Situs inversus totalis does not represent any medical disadvantage, but physicians should be aware of this anomaly in advance of diagnostic and therapeutic procedures or surgeries to prepare specialists for abnormal anatomy. Examples include the mirror technique in endoscopy and surgical manipulations that require more flexibility and creativity from the surgical team. Similarly, physicians should also be aware of duplication of the gastrointestinal tract in their patients, especially in the acute setting when requiring intervention. We found no cases of the combination of situs inversus totalis with doubling of the esophagus, stomach, and first part of the duodenum in the studied literature. Our case illustrates how endoscopic treatment of pancreatic pseudocysts is safe and effective even in such unusual cases.

Location of Case

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Declaration of Figures’ Authenticity

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