Acute pancreatitis in a patient with situs inversus/polysplenia syndrome: A case report and review of literature

Srinivas Kavuturu, MD1,2, William Roper, MD2, S. P. Stawicki, MD3

1 Department of Surgery, Bronx Lebanon Medical Center, Bronx, NY, USA
2 Department of Emergency Medicine, University of Pennsylvania School of Medicine, Philadelphia, PA, USA
3 OPUS 12 Foundation, Columbus, OH, USA

ABSTRACT
A 45-year-old man was admitted and evaluated for epigastric pain. Computerized Tomography (CT) scan revealed acute pancreatitis as well as incidental situs inversus and polysplenia. Patient did not have any congenital anomalies that are usually associated with polysplenia syndrome. No anatomical anomalies were found in the duodenum, the pancreas or biliary tree to account for pancreatitis. Based on history of alcohol abuse, this case was treated as alcohol induced pancreatitis. Complete resolution of pancreatitis was seen. This case is unique in that it describes acute pancreatitis in the setting of heterotaxy (situs inversus)/polysplenia syndrome.


Correspondence to: Srinivas Kavuturu, MD, Department of Surgery, Bronx Lebanon Medical Center, 1650 Grand Concourse, Bronx, NY 10456 USA.

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INTRODUCTION
Situs inversus is a rare congenital anomaly with an approximate incidence of 1 in 10,000 live births.1-4 The normal position of the organs is known as situs solitus. Situs inversus is a condition in which the usual positions of the organs are reversed from left to right as a mirror image of the normal anatomy. Situs ambiguous covers everything in between these two anatomic configurations on a continuum of asymmetric thoracic and abdominal organ formation. The term heterotaxy has been used to encompass these syndromes, which involve abnormal symmetry and malposition of the thoracic and abdominal organs and vasculature, including complex congenital heart and extracardiac abnormalities.5-8 Perhaps the best known example of heterotaxy is the Kartagener syndrome, a combination of situs inversus, bronchiectasis, and male infertility attributed to abnormal ciliary motility.9

Polysplenia is seen in almost 50% of patients with heterotaxy syndrome. Various gastrointestinal tract malformations have been reported to occur in the syndrome as well, including failure of the head of pancreas to form, duodenal duplications or atresia, intestinal malrotation, and biliary atresia.

We present a case of an adult patient with situs inversus, polysplenia, and pancreatitis without any duodenal or biliary malformations directly contributory to pancreatitis. Only one case of recurrent pancreatitis in polysplenic patient has been previously reported. The authors of that report attributed the pancreatitis to a duodenal diverticulum that was found to be obstructing the pancreatic duct.10

Figure 1. Computed tomographic scan of the abdomen showing pancreatitis with situs inversus.
DISCUSSION

Heterotaxy results from failure of the developing embryo to establish normal left-right asymmetry. This results in malposition of thoraco-abdominal organs and vasculature, complex congenital heart and extracardiac abnormalities. The term situs is applied to specify the left-right anatomic orientation of various asymmetric body structures. There are two types of unambiguous situs: (a) situs solitus – the normal; and (b) situs inversus – a mirror image of the normal.1

In patients with heterotaxy syndrome, the spleen is almost always affected, although the reason is not clearly understood. Three types of splenic anomalies have been described: (a) the spleen may be absent; (b) the spleen may be composed of a cluster of smaller splenules, a large spleen may be accompanied by several smaller splenules, or it may be multilobed; and (c) the spleen may be of normal size but located in the right upper quadrant of the abdomen. In a study of 109 autopsies of visceral heterotaxy with congenital heart disease, 58 patients (53%) had asplenia, 46 (42%) had polysplenia, and 5 (5%) had a single right-sided spleen. The polysplenia and asplenia syndromes, each have their own associated characteristic anomalies.2-4

The combination of ultrasound and CT imaging can diagnose most of the intra-abdominal anomalies contributory to pancreatitis. Magnetic resonance imaging (MRI) with MR cholangiopancreatography (MRCP) may be a useful as adjunctive diagnostic tools. Endoscopic Retrograde cholangio-pancreatography (ERCP) may be valuable as both diagnostic and therapeutic procedure in selected cases. An upper gastrointestinal series can help diagnose duodenal diverticulum, bowel atresia, and malrotation. An electrocardiogram should be performed to rule out cardiac conduction disturbances and other cardiac rhythm disorders. Echocardiography is the main diagnostic tool for diagnosing complex cardiovascular anomalies. Cardiac catheterization may be necessary in some patients.

CONCLUSIONS

Any patient with heterotaxy/polysplenia syndrome with pancreatitis should be aggressively investigated for structural and biochemical abnormalities that could potentially contribute to pancreatitis. The finding of a surgically correctable cause of pancreatitis can lead to curative surgery. Alcohol and gallstones should still be regarded as more common causes of pancreatitis in heterotaxy/polysplenia syndrome.

REFERENCES


