An *Ambiguus* Case of Acute Cholecystitis

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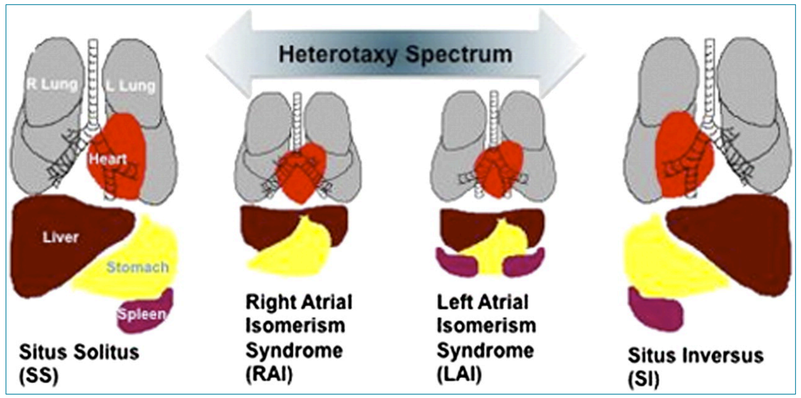
Introduction

- *Situs ambiguus* is a condition in which the internal thoraco-abdominal organs are abnormally arranged somewhere between *situs solitarus* and the complete mirror image (*situs inversus*). It is also called *Heterotaxy syndrome* – derived from the Greek terms *hetero*, meaning "different," and *taxy*, meaning "arrangement." The syndrome is often associated with cardiac and bronchopulmonary structural alterations, but it can also cause alterations in abdominal visceral organs, especially in association with the IVC. The affected individuals commonly have either asplenia or polysplenia. They can also have abnormal development of hepatic, biliary, pancreatic, intestinal, and gastric structures. [2,5] The prevalence is estimated to be 1 in 10,000 worldwide, and accounts for approximately 3% of all congenital heart defects. The condition appears to be more common in Asian populations, and occurs more frequently in children born to black or Hispanic mothers than in children born to white mothers.[6,7,8] 40% die within 2 years, and the majority die by 5 years old.



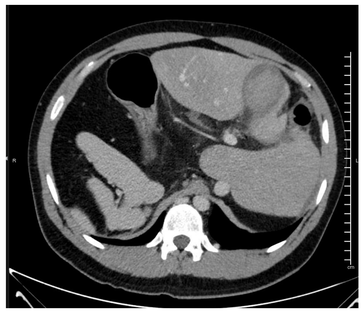
Case Presentation

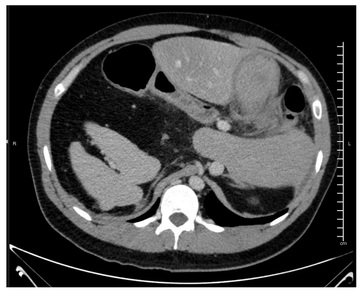
- A 33-year-old male, otherwise healthy, presented with chest tightness with onset of one day. The pain was located in the mid epigastric region, described as sharp, non radiating. It was worsened by deep inspiration, and laying in the left lateral decubitus. The patient examination was positive for mid epigastric tenderness to palpation, left upper quadrant tenderness to palpation; with no guarding or rebound appreciated. Upon a comprehensive workup, the patient was found to have situs ambiguus and acute cholecystitis. General surgery was consulted for cholecystectomy. The patient was discharged the day after presentation after undergoing a laparoscopic cholecystectomy without complications.

Diagnostics

(Fig. 1-2) Ultrasound abdomen significant for cholelithiasis with thickened gallbladder wall and suspected situs ambgiuus

(Fig. 3-4) CT abdomen and pelvis with contrast significant for inflammatory stranding/distended gallbladder concerning for acute cholecystitis





(Fig. 5-6) Sagittal and Coronal view of thickened gallbladder

Differential Diagnoses

- Acute Coronary Syndrome
- Pulmonary Embolism
- Cholecystitis
- Pancreatitis
- Cholangitis
- Pneumonia
- Indigestion
- Functional Dyspepsia

Discussion

- The patient was found to have heterotaxy syndrome with **left isomerism including bilobed lungs, right-sided polysplenia, left-sided liver azygous continuation of the IVC.**

- This case was a prime example where providers must be aware of anchoring bias. The patient's presentation was atypical for biliary disease in terms of demographics and presenting complaints. This clinical case requires the physician to prudently deliberate over pretest probabilities for various investigations.

- Although situs ambiguus is an uncommon condition, it is commonly associated with a host of complications. Those who survive later into life commonly present with acute thromboembolic, pulmonary, and GI pathologies. These patients are functionally asplenic and require expedited management due to risk of severe infection and sepsis.

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