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Small bowel obstruction in an adult patient with situs ambiguous and mid gut malrotation

Shwe Phyo Han, Jonathan Grassby

ABSTRACT

Introduction: Situs ambiguous or heterotaxy syndrome is defined as the abnormal positioning of internal viscera relative to the normal. Diagnosis in adult is extremely rare as 90–99% of the patients have severe cardiac abnormalities and die by the age of five years.
Case Report: A 32-year-old male was presented to hospital with sudden onset abdominal pain, abdominal distension, vomiting and absolute constipation for one day. There was no other medical problems. Examination was also unremarkable. Abdomen was distended and generally tender. Per rectal examination showed empty rectum. Blood tests were unremarkable. Computed tomography scan of abdomen and pelvis showed closed loop mid to distal small bowel obstruction with small bowel wall thickening. Malrotation of the bowel was noted. It also showed that stomach and spleen were on the right side of the body. Emergency laparotomy showed small bowel volvulus and spleen, stomach, duodenojejunal flexure and small bowel were in the right side of the abdomen and colon was in the left side of the abdomen. Ileocecal valve was noted in the left side of the abdomen. Adhesiolysis and derotation of the affected small bowel were performed. Appendicectomy was performed due to its location in left lower quadrant. The patient was discharged from the hospital four days after the operation.
Conclusion: There is no case report on adult situs ambiguous presented with acute intestinal obstruction before. We report this case for extremely rare occurrence of situs ambiguous with mid gut malrotation presented with small bowel obstruction in the adult age group.

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Keywords: Abdominal pain, Mid gut malrotation, Situs ambiguous, Small bowel obstruction, Vomiting

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INTRODUCTION

Situs ambiguous or heterotaxy syndrome is defined as the abnormal positioning of internal viscera relative to the normal [1]. It is due to a primary defect in lateralization around day-28 of gestation, leading to a deviation from the normal position of viscera [1]. The incidence of situs ambiguous is approximately 1 in 40,000 live births [2]. Diagnosis in adult is extremely rare as 90–99% of the patients have severe cardiac abnormalities and die by the age of five years [2, 3].

CASE REPORT

A 32-year-old male was presented to Dubbo base hospital with sudden onset abdominal pain, abdominal distension, vomiting and absolute constipation for one day. There are no other medical problems. There was no previous abdominal operation. On examination, he was afebrile and observations were within normal limits. Abdomen is distended and soft. There was generalized tenderness in the abdomen. Per rectal examination showed empty rectum and no mass or lesion was noted. Blood tests were unremarkable.

Computed tomography scan of abdomen and pelvis with oral and IV contrast were performed. It showed closed loop mid to distal small bowel obstruction with small bowel wall thickening (Figure 1). Malrotation of the bowel was noted. It also showed that stomach and spleen was on the right side of the body (Figure 2 and Figure 3).

Emergency laparotomy was performed. Small bowel volvulus was noted. Spleen, stomach, duodenojejunal flexure and small bowel were in the right side of the abdomen and colon is in the left side of the abdomen. Ileocecal valve was noted in the left side of the abdomen. Adhesiolysis and derotation of the affected small bowel were performed. Appendicectomy was performed due to its location in left lower quadrant.

There was no complication after the operation and the patient was discharged from the hospital four days after the operation.

DISCUSSION

Situs ambiguous can be divided into two main types. They are left isomerism (situs ambiguous with polysplenia) and right isomerism (situs ambiguous with asplenia). Associated anomalies include congenital heart disease, IVC interruption with azygos or hemiazygos continuation (left sided isomerism), bilateral bilobed lungs (left sided isomerism), bilateral trilobed lungs (right sided isomerism), polysplenia (left sided isomerism), asplenia (right sided isomerism), midline liver and right sided stomach (left sided isomerism) [1, 2].

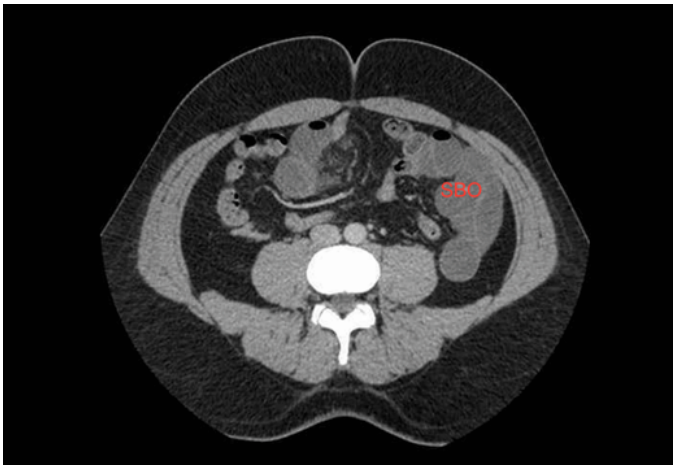

Figure 1: Computed tomography scan of abdomen and pelvis showing closed loop mid to distal small bowel obstruction with small bowel wall thickening.

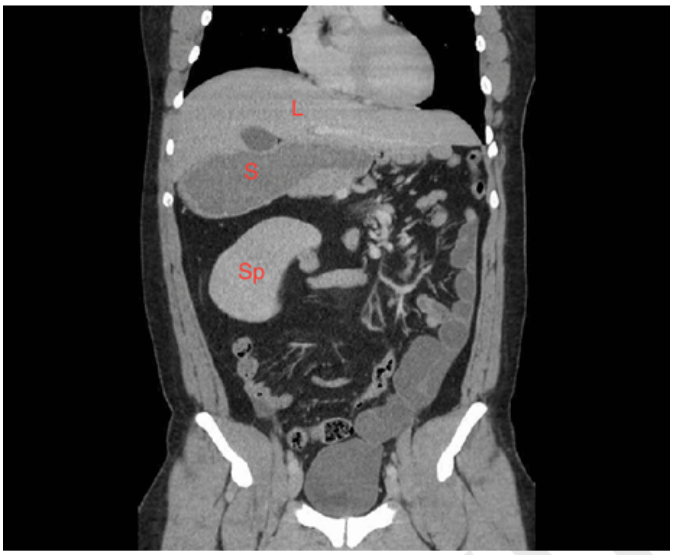

Figure 2: Coronal section of computed tomography scan showing stomach and spleen on the right side of the body.

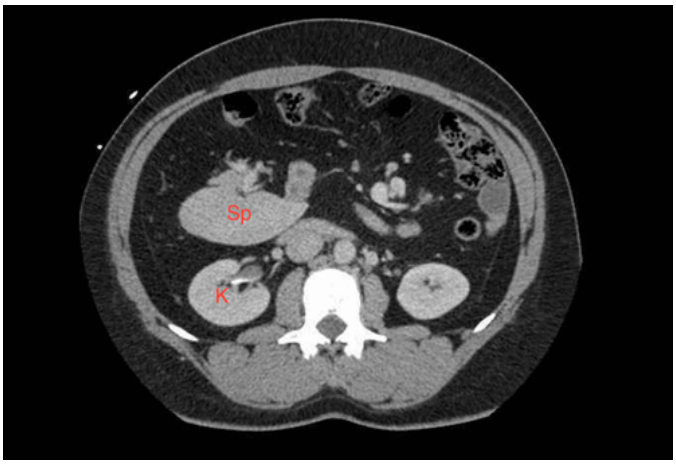

Figure 3: Cross-sectional computed tomography scan showing spleen on the right side of the body.

The number of spleens ranges from one to ten in left sided isomerism and spleens may be located either bilaterally or if unilateral, ipsilateral to the stomach because spleen arises from the dorsal mesogastrium. Patients with situs ambiguous can present in different ways due to different anatomical abnormalities that may be present. Most of the patients will not fit into the two categories described above and many patients will fall somewhere in between. Therefore, individual anatomical abnormalities should be documented rather than using the terms left and right sided isomerism. Thorough imagine studies are required to diagnose the anatomical abnormalities [1].

Congenital heart disease is the major cause of morbidity and mortality in situs patients. Patients with right sided isomerism have very high incidence of

congenital heart disease. Patients with situs ambiguous with asplenia have increased risks of sepsis secondary to encapsulated bacteria. Vaccination against these organisms is recommended [1].

Situs ambiguous may not cause any symptoms but they can present with confusing symptoms when they have common surgical problems such as appendicitis and cholecystitis [1].

Mid gut malrotation including the cases of non-rotation is seen in 1 in 500 births. It is associated with situs ambiguous in 70% of the cases. It arises from failure of counter clockwise rotation around the superior mesenteric artery [1]. Intestinal malrotation in adult is rare with an occurrence between 0.0001–0.19%. Presentation with chronic abdominal pain, vomiting, diarrhea, abdominal distension and early satiety is common. Acute presentations with intestinal obstruction due to small bowel volvulus are less common [4].

CONCLUSION

There are few cases of situs ambiguous reported which were incidentally found during operation and abdominal imaging. There is no case report on adult situs ambiguous presented with acute intestinal obstruction before. We report this case for extremely rare occurrence of situs ambiguous with mid gut malrotation presented with small bowel obstruction in the adult age group.

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Author Contributions

Shwe Phyo Han – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Jonathan Grassby – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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