



Flipped and Twisted - Situs Inversus Totalis with Cecal Volvulus

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Abstract

Situs inversus totalis is an uncommon congenital anomaly resulting in 'flipping' of intra-abdominal and intrathoracic organs. It occurs in only 0.01% to 0.02% of live births and poses a diagnostic challenge for abdominal emergencies. We describe a rare case of intestinal obstruction in an adult patient of situs inversus, due to cecal volvulus twisted on its long mesentery with a spleniculus acting as pivot. Literature review regarding etiology, evaluation and management of situs inversus and cecal volvulus is presented.

Keywords: Situs inversus totalis; Cecal volvulus; Intestinal obstruction

Introduction

Situs inversus totalis is an autosomal recessive congenital disorder known to present with cardiac anomalies and intestinal obstruction due to intestinal malrotation in infants and children [1]. Presentation in adults is rare and usually incidental. The anomalous anatomical mapping associated with volvulus presents as a diagnostic predicament. We report a case of a young male with situs inversus totalis who presented with intestinal obstruction due to cecal volvulus.

Case Presentation

A 25 year old male presented with colicky pain in the hypogastric region for two days. He had three episodes of vomiting and absolute constipation over the last day. Pain was mild to moderate in severity and gradually spread to the whole abdomen. He was a diagnosed case of dextrocardia incidentally discovered during his pre-employment fitness check.

On examination, he had sinus bradycardia (pulse = 50/min), was normotensive and his respiratory rate was 32/min. His abdomen showed asymmetry with a distended visible loop of bowel in the left hemiabdomen. There was generalized abdominal tenderness and a distended bowel loop was palpable on the left side. Inferior margin of the liver was also palpable on left side. Occasional bowel sounds could be heard on auscultation and digital rectal examination revealed an empty rectum. Heart sounds including apex beat were audible on the right side of chest. Nasogastric tube and Foley catheter were passed, and intravenous fluid resuscitation started.

Hematological workup revealed a raised leukocyte count of 18.2/mm³ and hemoglobin of 11.2 gm/dl. Liver function tests and renal functions were within normal range. Chest radiograph showed dextrocardia and stomach bubble with nasogastric tube could be seen on right side. Erect view of plain abdominal X-ray revealed a massively dilated bowel loop with haustrations in the left field suggesting large bowel volvulus (Figure 1).

Ultrasonography showed situs inversus with liver on left, spleen and stomach on right, and multiple spleniculi in perisplenic region. Contrast enhanced CT of abdomen and pelvis showed situs inversus totalis with a large dilated loop of cecum in the left hemiabdomen. Rest of the bowel was collapsed. Spleen was seen on right side with spleniculi in perisplenic region and mesentery of the bowel. Anomalous vein running parallel to inferior vena cava was noted draining the left kidney (Figure 1).

Immediate midline laparotomy was performed under general anesthesia. A massively dilated cecum and ascending colon with diameter of 12 cm (Figure 2) was seen twisted over a long mobile mesentery. A 3 cm × 1.5 cm rounded lesion near the base of mesentery was seen acting as the pivot for rotation of the cecum (Figure 3). This lesion was suspected to be a spleniculus.

Volvulus was detorted (Figure 3) the whole bowel was run through to confirm a completely

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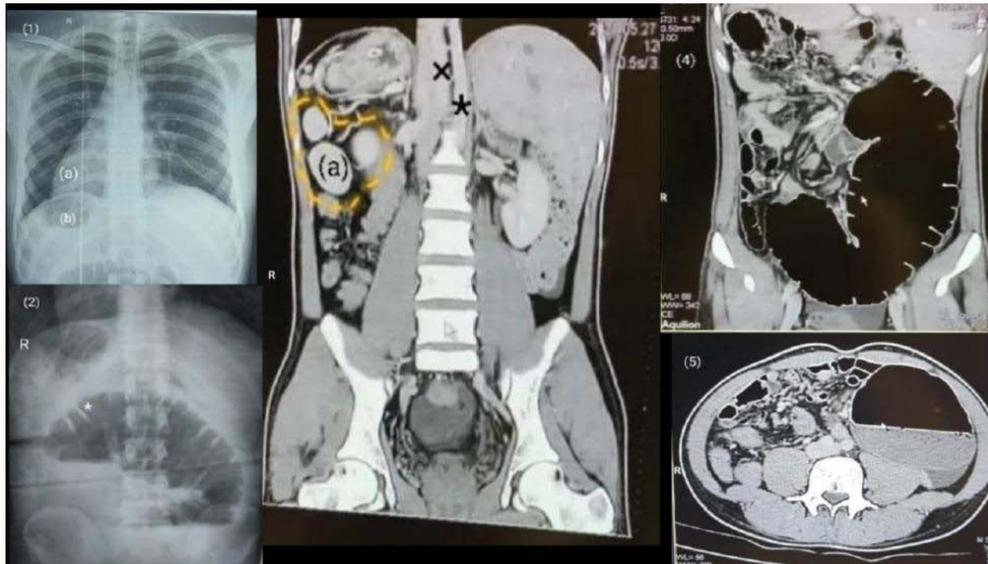


Figure 1: Radiological investigations: (1) Chest PA view showing (a) dextrocardia, (b) stomach bubble on right; (2): Abdomen AP view (erect) with a central dilated loop of bowel having maintained haustrations*; (3) (a) spleniculi in perisplenic area on right, * marks aberrant vessel parallel to Inferior vena cava (x) draining the left kidney; (4) coronal view, (5) axial view of contrast enhanced CT showing massively dilated large bowel loop (arrow) on the left displacing the rest of the bowel to right.

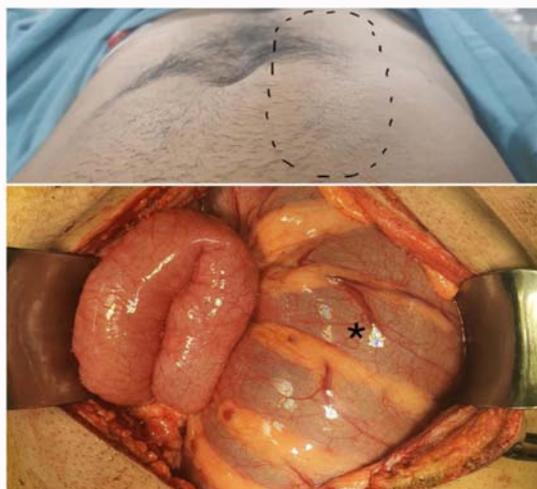


Figure 2: Preoperative and perioperative findings; the image on top shows preoperative abdomen with marked asymmetry and a large visible bowel loop (dotted line); the image below shows intraoperative cecal volvulus* in the corresponding area.

free mesentery of both small and large bowel. A few soft adhesions between loops of large bowel were lysed. Right hemicolectomy with ileotransverse anastomosis was performed to prevent prolonged postoperative ileus secondary to massively dilated part of gut. Postoperative course of the patient was unremarkable, and he was discharged on the third postoperative day.

Discussion

Situs inversus totalis is a rare presentation ranging between 1:5000 and 1:10000 live births [2]. There are variations in presentation with major difference being in the position of the heart (levo/dextrocardia), and cases of situs ambiguous where there is incomplete flipping of abdominal or thoracic organs. When compared with situs solitus, more complex malrotations of gut may be associated with abdominal

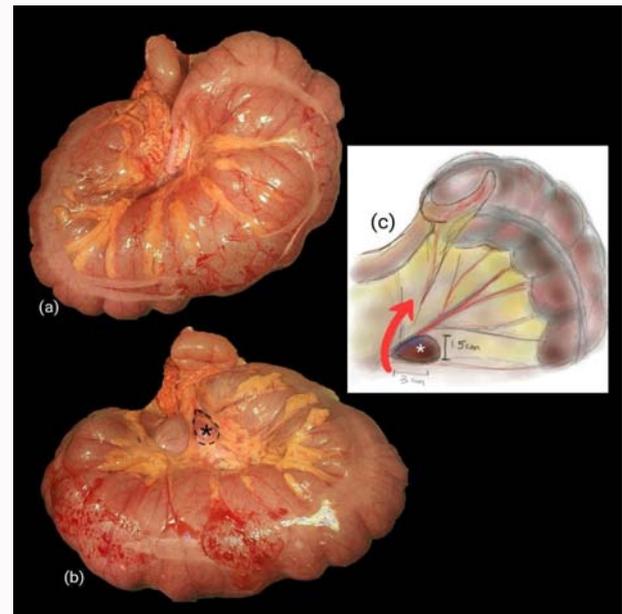


Figure 3: Cecum. (a) prior to detorsion; (b) after detorsion (c) pictorial depiction of the cecum after detorsion showing direction of rotation of volvulus over the long mesentery with a lesion* likely spleniculus acting as pivot.

anomalies like polysplenia, asplenia, and folded pancreas, midline liver and vascular anomalies including disruption of the inferior vena cava. Presentation is usually in pediatric age group [3].

Although the exact etiology of situs inversus is not established, the ‘flipping’ of organs is theorized to be secondary to absence or defects in special types of embryonal cilia that are responsible for organization of organs and ensuring the correct laterality [4]. Hence there is a strong association with Kartagener’s syndrome in some cases of situs inversus [4,5]. The variability in presentation is also because of the effectiveness or failure of the organs to migrate and get affixed to their correct position. Since the bowel rotates back into the

abdominal cavity during the 5th to 11th week of fetal life, malrotation or abdominal component of situs inversus occurs during this period.

Volvulus is defined as the 'twisting' of gut around its mesentery causing intestinal occlusion. Cecal volvulus is rare and is responsible for only 1% of all cases of intestinal obstruction [6]. Cecal volvuli occurring in situs inversus are sparse [7], although theoretically it is more probable. Abdominal X-ray in our patient demonstrated a large dilated loop in the abdomen with visible haustrations. This led to a high suspicion of cecal volvulus because cecum, unlike the descending colon, may retain its haustrations despite dilatation and can distend massively. Air may be seen in the distended appendix. Another curious finding in this case was presence of spleniculi in cecal mesentery acting as a pivot for twisting of cecum.

Contrast enhanced CT has 90% sensitivity. Some typical signs seen on CT include the 'whorl sign' that consists of a swirl of folded mesentery and surrounding fat [6], the 'bird beak sign' that shows progressively tapering bowel, and 'central appendix sign' due to abnormal position of the appendix near midline [8].

Surgical management of cecal volvulus in a patient with situs inversus may be a simple detorsion with cecopexy and appendectomy. Right hemicolectomy may be performed in cases with compromised bowel or as a measure to prevent recurrence [6]. Simple detorsion without cecopexy has a high rate of recurrence [6]. In our case the cecum and ascending colon were massively dilated and could have resulted in functional paralysis of smooth muscle for a prolonged period. So, we decided to perform a right hemicolectomy. Whether or not to perform primary anastomosis depends on the patient's haemodynamics and state of the bowel intraoperatively [6]. In our patient we performed a right hemicolectomy with end to end ileotransverse anastomosis.

Only a few such cases have been reported in literature [7,9]. Owing to the anomalous position of organs, diagnosis in acute presentation is challenging in both common and uncommon abdominal emergencies. This may lead to a delay in management

with dire consequences. Mortality in delayed management of cecal volvulus has been reported to be as high as 30% [6].

Conclusion

High index of suspicion and timely diagnosis is the key to successful management of patients having situs inversus with cecal volvulus. Management is based on correction of the volvulus, cecopexy or hemicolectomy to prevent recurrence.

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