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MALROTATION OF THE GUT WITH MIDGUT VOLVULUS IN AN ADULT: A RARE CASE REPORT

Radiology

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ABSTRACT

Intestinal malrotation is a rare congenital condition caused by the absence of or incomplete rotation of the small bowel during the embryonic period and is defined as any deviation from the normal 270 degree counter clockwise rotation of the midgut. If this condition is not timely recognized, it may result in disastrous consequences, such as gangrene of the small gut. 64-80% of cases present primarily during the neonatal period prior to the first month of life, as acute intestinal obstruction due to volvulus of small intestine. The presentation of intestinal malrotation in adults is rare, and occurs in approximately 0.2-0.5%. We present the case of a 17-year-old male who presented to the emergency room with recurrent episodes of colicky abdominal pain and bilious vomiting. Contrast-enhanced computerized tomography (CECT) revealed malrotation of the gut with midgut volvulus. Contrast enhanced CT showed the abnormal anatomic location of a right sided small bowel, a left-sided colon and an abnormal relationship of the superior mesenteric vein (SMV) situated to the left of the superior mesenteric artery (SMA) instead of to the right and the characteristic 'whirpool or whirl sign' describing the swirling appearance of bowel and mesentery twisted around the superior mesenteric arterial axis with the presence of midgut volvulus. In patients with acute obstruction, this differential should be kept in mind, especially if the patient has no previous abdominal surgery or evidence of tuberculosis.

KEYWORDS

Intestinal Malrotation, Midgut Volvulus, Adult, Whirlpool Sign, Corkscrew Sign, Laproscopy, Ladd's Band

Introduction

Intestinal malrotation is a clinical entity that encompasses partial to complete failure of the 270 degrees counterclockwise rotation of the midgut around the superior mesenteric vessels in the fetal life [1]. Its incidence is one in every 200 - 500 newborns [2]. The incidence of symptomatic cases is one in 6,000 newborns [2]. Presentation in adults is very rare. A high index of suspicion is often necessary to diagnose this condition in adults.

Case Presentation

A 17-year-old male presented to the emergency room with complaints of central abdominal pain for the past eight to nine hours and multiple episodes of vomiting. The pain was cramping, located in the central abdominal area. The pain occurred every one to two hours and each episode lasted 15-20 minutes. The pain was associated with episodes of bilious vomiting that provided partial relief. His last bowel movement was two days ago. The patient had been experiencing multiple such episodes for the past ten years. No history suggestive of tuberculosis was present. The patient had no previous abdominal surgery. The patient's blood pressure was 110/74 mm of Hg. The patient's pulse rate was 108 beats per minute. The systemic examination was unremarkable. The upper abdomen was distended. The abdomen was firm and mild tenderness was present. No free fluid was present. Exaggerated bowel sounds were heard. Rectal examination was normal. Routine hematological investigations and renal function tests were within normal limits. Abdominal x-rays revealed no significant abnormality(no dilated bowel loops or air fluid levels were noted).

Ultrasonography was done, which revealed small bowel loops and mesentery swirling around superior mesenteric arterial axis (whirlpool sign) (Figure 1).

Urgent contrast-enhanced computed tomography (CECT) abdomen was done and revealed the small bowel and duodenojejunal (DJ) flexure to be lying on the right side of the abdomen and not crossing over to the left, large colon was seen predominantly on left side of the abdomen (Figure 2), caecum was seen in the left mid abdomen, Twisting of superior mesenteric vein, proximal jejunal loops, ascending colon and terminal ileum was seen around the superior mesentric artery (whirlpool sign) (Figure 3). and also the reverse relationship of superior mesenteric vein to superior mesenteric artery (Figure 4).

Significant narrowing of superior mesenteric artery was seen approx 5.4 cm from its origin for a length of 1.2 cm. Significant narrowing of superior mesenteric vein and proximal jejunal loops was also seen. Resultant mild dilatation of duodenum was seen. Free passage of oral contrast was seen in large and small bowel loops distal to narrowing. Above findings were suggestive of intestinal malrotation with midgut volvulus.

A diagnosis of malrotation of the gut was made, and the patient was planned for urgent surgery. Typical findings of malrotation were seen intraoperatively small bowel loops predominantly on the right side of the abdomen, large bowel loops on left side, and the DJ flexure not crossing the midline and remaining to the right of the midline. Ladd's procedure was performed. The postoperative period was uneventful and the patient was discharged on day 7. The patient has been doing well for the last two months and is totally symptom-free.

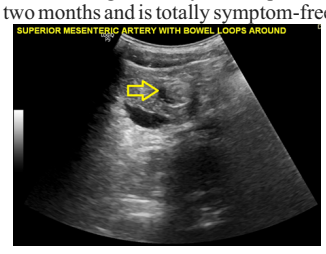

Figure 1: Ultrasound image showing Whirlpool Sign (Yellow arrow)

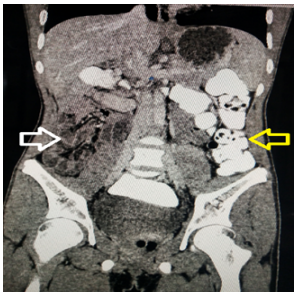

Figure 2: CECT coronal image showing Large bowel on the left side (yellow arrow), and Small bowel on the right side of the abdomen (White arrow)

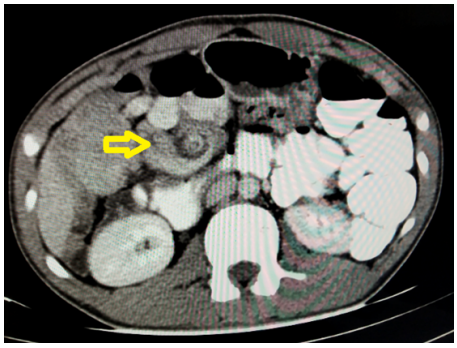

Figure 3: CECT axial image showing Whirlpool sign (Yellow arrow)

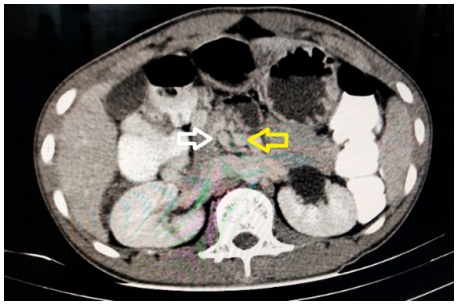

Figure 4: CECT axial image showing SMA on the right side (White arrow) and SMV on the left side (Yellow arrow)

Discussion

Malrotation of the gut is the complete or partial failure of 270° of counterclockwise rotation of the midgut around the superior mesenteric pedicle. The rotation of the intestines in the embryonic period occurs in three stages [3]

Stage I occurs between 5th to 10th gestation week. It includes extrusion of the midgut into the extra embryonic cavity, a 90° counter-clockwise rotation and return of the midgut into the fetal abdomen.

Stage II occurs in 11th week and involves further counter clockwise rotation within the abdominal cavity completing a 270° rotation. The duodenum rotates caudal to the artery, and its C-loop traces this path. The transverse and ascending colon demonstrate the path of rotation of the cecum cephalad to the artery.

Stage III involves fusion and anchoring of the mesentery. The duodenum becomes fixed retroperitoneally in its third portion, emerging at the ligament of Treitz, and the cecum becomes fixed to the lateral abdominal wall by peritoneal bands. The takeoff of the branches of the superior mesenteric artery elongates and becomes fixed along a line extending from its emergence from the aorta to the cecum in the right lower quadrant [4].

Malrotation in adults is rare and occurs with obscured clinical symptoms, such as recurrent abdominal pain and vomiting, often resulting in multiple hospital visits and posing a diagnostic dilemma to the unpolished surgeon [5-6]. This condition may also present in an acute way, due to midgut volvulus, and may result in intestinal ischaemia and gangrene. The consequences of this are disastrous and often result in massive bowel gangrene, death, and short bowel syndrome if the patient survives. Timely recognition of the condition is the key to survival [6].

Clinical presentation of adult intestinal malrotation is usually asymptomatic. Approximately 30% of these patients present with vomiting and 20% have recurrent unspecific abdominal pain. Symptoms can arise from acute or chronic intestinal obstruction that may be caused by the presence of the Ladd bands and/or a volvulus with resultant ischemic necrosis and perforation peritonitis. Some may present as malabsorption associated with diarrhea and nutritional deficiencies caused by bowel lymphedema resulting from lymphatic obstruction by chronic volvulus and resulting in loss of proteins into the bowel lumen. Rare presentations of chronic volvulus include cases of obstructive jaundice by mechanical compression of the biliary tract, chylous ascites and superior mesenteric vein thrombosis, secondary to long-standing lymphatic and venous obstruction [7-10].

Plain abdominal radiographs are not useful and the investigation of choice in adults remains a contrast-enhanced CT scan[6]. The typical findings are reversed relation of superior mesenteric artery (SMA) and superior mesenteric vein (SMV), a whirled appearance of the vasculature entering the volvulus (whirlpool sign), small bowel loops in the right upper abdomen, large bowel loops in left side of abdomen, lack of visualization of the caecum in the right iliac fossa, dilatation of various duodenal loops, and duodeno jejunal flexure to the right (corkscrew sign). The typical reversed relationship of superior mesenteric vessels can be seen on ultrasonography as well. All patients, regardless of age, should undergo surgery as it is impossible to predict the development of catastrophic complications [5].

The classic treatment for incomplete intestinal rotation is the Ladd procedure, which entails counter-clockwise detorsion of the viable midgut volvulus (if present), division of the abnormal colo duodenal Ladd bands tethering the midgut and causing extrinsic compression, mobilization/ kocherisation of the duodenum and the right colon, widening of the mesenteric base by section of possible adhesions near the superior mesenteric vessels to prevent further volvulus and removal of the malpositioned appendix. The aim of this procedure is to reduce the risk of acute volvulus, by locating the small intestine in a non-rotating position and widening the base of the mesentery. Appendectomy is performed due to possible difficulty in the diagnosis of future appendicitis, distant from the classic lower right quadrant position. Laparoscopic Ladd procedure is safe now days due to early discharge and oral intake [11,12].

Conclusion

A high index of suspicion is needed to diagnose malrotation of the gut in adults. This condition should be suspected in patients with recurrent episodes of abdominal pain and bilious vomiting with no history suggestive of tuberculosis or any history to support an adhesive cause. In the emergency setting, an ultrasound and contrast enhanced computed tomography scan looking for the reversed relation of superior mesenteric vessels, whirlpool sign, position of large and small bowel can be very useful. Timely diagnosis will prevent the deadly complications of this disease.

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