Midgut volvulus due to congenital malrotation in adults: A case report

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Abstract

Intestinal malrotation is a rare congenital anomaly. It may occur in adults but it is very rare. Our case is a 20-year-old male from Khartoum (Sudan) presented to the emergency department with acute onset abdominal pain, vomiting, and abdominal distension diagnosed as having intestinal obstruction due to congenital malrotation complicated by midgut volvulus. Midgut volvulus secondary to congenital malrotation in adults is a very rare condition, and abdominal CT scan with IV contrast is helpful for preoperative diagnosis. Treatment is by Ladd procedure.

Introduction

Intestinal malrotation is an uncommon anomaly complicating approximately 0.2% of births. It is usually diagnosed during the first weeks of life. In adults, it is extremely uncommon and a rare cause of intestinal obstruction. The incidence in adults can be estimated between 0.001% to 0.19%. Symptomatic intestinal malrotation in adults is not common as most of them are discovered accidentally, but they can cause several presentations ranging from nonspecific symptoms with mild complaints to acute abdomen requiring surgical intervention [1].

Case presentation

A 20-year-old male from Sudan presented to Omdurman teaching hospital Emergency department complaining of acute onset colicky abdominal pain that started 48 hours prior to presentation. He also had vomiting and abdominal distention. He had a clear medical background with no history of operation before. He had not opened his bowel for 48 hours before admission. After initial assessment, positive findings were abdominal distension, hyper-resonance in percussion, and exacerbated bowel sound, and all blood lab results were normal.

Abdominal X-ray showed multiple air fluid levels and dilated loops of bowel. Emergency computed tomography (CT) scan was obtained which demonstrated features of midgut malrotation where the DJ flexure in the right side and the superior mesenteric artery right to the superior mesenteric vein “positional crossing and whirlpool volvulus of SMA” (picture E,F). The dilated caecum was located in the left upper quadrant (Picture C,D). There is a sign of small-bowel volvulus. There were also a few dilated loops of small bowel in the upper abdomen. Chest X-ray did not show air under the diaphragm.

Management was started at the emergency department with nasogastric tube insertion. Intravenous fluids, broad spectrum antibiotics, and patient was prepared for an emergency exploratory laparotomy. The findings at operation included dilated small bowel in the upper abdomen and the dilated caecum was found on the left side of the abdomen (Picture A). DJ flexure in the right side (Picture B). Partial torsion and necrosis of 155 cm of the small bowel just 56 cm from DJ flexure. Loops of small bowel occupying the right paracolic gutter and the right iliac fossa. There were fibrous bands over the distal part of the duodenum, on the right side of the abdomen, confirming midgut malrotation. The twisted necrotic small bowel was resected and anastomosis was performed, the congenital band was divided, and an appendectomy was carried out with correction of the anatomical malrotation. The patient had an uneventful postoperative recovery.

Picture A: Dilated cecum and small-bowel.
Discussion

The intestine is made of foregut, midgut and hindgut. During the fourth week of embryonic development the midgut increases in length and the abdominal cavity can’t accommodate it and so it herniates through the umbilicus. During the tenth week of development the midgut return back to the abdomen and while doing so it rotate a 270° counter clock wise around the axis of the superior mesenteric artery, which supplies it , so that the right colon is placed on the right side of the body. Failure of this process result in the formation of the midgut malrotation. The exact cause is not established yet but there is often a fibrous band that tethers the right colon preventing it is rotation [1]. According to stinger classification there three types of malrotation. Type1: non-rotation, type 2: duodenal malrotation and type3: duodenal plus caecal malrotation. Also, there is malfixation of the mesentery with narrow pedicle that can result in volvulus. Several conditions is associated with midgut malrotation like Martinez-Friay syndrome where malrotation associated with multiple GI atresia [2]. In paediatrics intestinal malrotation presents early with symptoms of small bowel obstruction ((abdominal distension and bilious vomiting) and can be diagnosed with upper GI contrast series. However, most adult with malrotation are often diagnosed accidently during imaging or exploratory laparotomy for other causes [3]. Many patient might be asymptomatic and most of them have nonspecific GI symptoms like abdominal pain and bilious vomiting. Malrotation in adults is rare cause of intestinal obstruction and its rate in autopsy is 1 in 6000 [4]. In children upper GI series study can detect malrotation, But in adults a contrast enhanced abdominal CT scan can help guide the diagnoses via viewing relation of the superior mesenteric artery and vein (artey is found to the left of the vein in malrotation). Mid gut volvulus is easier to be found in upper GI series (shows corkscrew sign of the proximal small bowel), but because of most adults undergoes a contrast CT scan you might find whirlpool appearance (swirling appearance of bowel and mesentery twisted around the axis of the superior mesenteric artery). Other features are duodenal obstruction, mesenteric vasculature congestion and evidence of malrotation with the caecum on the left [5]. Children and adults who were found to have symptomatic malrotation should undergo surgery [6]. ladd’s operation it the treatment of choice in both children and adults [7]. ladd’s operation involves counter clock wise detorsion of the small bowel, surgical division of ladd’s band(fibrous tissue that tethers the intestine) widening of small bowel mesentery , performing appendectomy and replacement of the small bowel to the right and caecum and colon to the left. It can done via midline incision or laparoscopy. Despite limitation some authors had concluded that use of laparoscopy in treating cases of malrotation in safe and effective [8]. However, in cases presenting with volvulus managing it with laparoscopy can be impossible and it’s safe to operate via a midline incision [7].

Conclusion

Midgut volvulus secondary to congenital malrotation in the
adult it’s a rare condition, abdominal CT scan with IV contrast is helpful for preoperative diagnosis which show positional crossing and whirlpool volvulus of SMA and those are the characteristic manifestation of midgut malrotation. Patients present with symptoms and signs of intestinal obstruction. Treatment is emergency surgical intervention (Ladd’s procedure). The Ladd procedure consists of division of Ladd’s bands, widening of the mesentery, appendectomy and to leave the anatomical malrotation uncorrected.

References