Intestinal Malrotation in Adults: A case series

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INTRODUCTION

Midgut malrotation is a rare congenital anomaly as a result of non-rotation, incomplete rotation or reversed rotation and fixation of midgut. Mostly it presents with bowel obstruction or volvulus in early life. Presentation in adult is usually variable, ranging from recurrent non-specific colicky abdominal pain to acute abdominal emergencies. Delayed diagnosis due to non-specific symptoms in elective cases, and conversion to laparotomy due lack of intra-operative anatomical understanding in emergency cases imposes increase in patient dis-satisfaction, delayed recovery and healthcare cost burden. We report an interesting case series of various presentations causing significant diagnostic challenge due to atypical clinical and radiological signs.

TAKE HOME MESSAGE

General Practitioners should be aware of such atypical presentations of rare conditions causing recurrent abdominal pain. They should consider appropriate further imaging investigations and referrals for best possible patient care.

IN THE ERA OF LAPAROSCOPIC SURGERY, A GENERAL SURGEON SHOULD ENCOUNTER SUCH AN UNUSUAL CASE ONCE IN A LIFETIME. IT IS OF OUTSTANDING IMPORTANCE TO THE CLINICAN; SOMETIMES PATIENTS BEING LABELED AS HAVING MUNCHHAUSEN SYNDROME.

WHY IS IT MISSED?

Case 1: A 35-year-old male patient, presenting with a 2-day history of right lower quadrant abdominal pain, associated with vomiting, fever and reduced appetite. On examination there was right iliac fossa tenderness. Blood tests showed a white cell count (WCC) of 11 and C reactive protein (CRP) of 37. Therefore, the patient was assumed to have acute appendicitis and was taken to theatre.

Case 2: A 48-year-old female, presented with 1-day history of severe supra-pubic pain with background of on and off episodes of lower abdominal pain for many years, feeling nauseous without vomiting. Her abdominal examination and blood results were normal. A CT scan reported intestinal malrotation with midgut volvulus, without acute obstruction or perforation.

Case 3: A 66-year-old male presenting in outpatients with recurrent attacks of severe abdominal pain and sub-acute obstruction of small bowel, investigated with a computed tomography (CT) scan, which revealed intestinal malrotation.

TREATMENT

Case 1: Laparoscopy converted to laparotomy revealed a severely inflamed perforated appendix (3x10cm) with intestinal malrotation with Ladd’s bands. We proceeded to appendicectomy and division of Ladd’s procedure.

Case 2: Intra-operatively we found distended loops of small and large intestine so was converted to mini laparotomy. Complete non-rotation was confirmed along with midgut volvulus without bowel ischemia. She underwent caecectomy to the left lateral abdominal wall and appendicectomy.

Case 3: He was electively taken to theatre for division of Ladd’s band. He recovered well and was discharged on second post-operative day. He was followed up for 2 months, without any complaints.

WHY IS IT MISSED?

Non-specific recurrent abdominal pain with normal clinical and laboratory findings gives false assurance of patients wellbeing to the clinician; Sometimes patients being labeled as having Munchhausen syndrome.

Appendicities which lead to pain in the left lower quadrant is extremely uncommon and can occur with congenital abnormalities that include true left-sided appendix or as an atypical presentation of right-sided, but long appendix, which projects into the left lower quadrant. This with wrong impression of either diverticulitis or urinary tract infections is treated conservatively with a course of antibiotics.

ANATOMY AND EMBRYOLOGY

Formation of primary intestinal loop by rapid elongation in 8th week of embryonic life, the caecal blind limb develops into ileo-caecal junction and part of jejunum. This process is accompanied by rotation of the intestinal loop, initially in sagittal position, completes a 270° counter clockwise rotation around the superior mesenteric artery (SMA) and first rotation is complete by 5th gestational week. (2)

The second rotation is completed by 10th week. This rotation starts posterior to the superior mesenteric artery (SMA) and the SMA lies to the left of the superior mesenteric vein (SMV). This process is accompanied by rotation of the intestinal loop. Initially in sagittal position, completes a 270° counter clockwise rotation around the superior mesenteric artery (SMA) and first rotation is complete by 5th gestational week. (2)

The reversed rotation occurs when primary intestinal loop rotated in clockwise direction rather than counterclockwise direction. This results in proximal jejunum, the first part to return comes to left side and later returning loops gradually turns right. This is completed rotation of the intestinal loop. Incomplete form is known as malrotation. The caecum can be attached to posterior aspect of the duodenum, transverse colon or midline of abdomen. This rare variant can be difficult to diagnose with preoperative evaluation and intra-operative assessment.

Transverse colon lies near the midline, high up. It grows then into right and descends from right upper quadrant to right iliac fossa. The mesenteric root with superior mesenteric artery and vein lies near the transversalis fascia and peritoneum layer.

In these cases “Ladd procedure” is indicated to divide the bands, revealing the cause of obstruction.(3)

REFERENCE


