Intestinal volvulus after conservative management of incidental midgut malrotation discovered at laparoscopic appendectomy in a teenager

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Abstract: Midgut malrotation (MMR) is the abnormal rotation of the foetal midgut around the axis of the superior mesenteric artery which in symptomatic neonates requires a Ladd's procedure. We present a rare case of midgut volvulus occurring in a teenager 3 days after observational management of incidentally discovered MMR during laparoscopic appendectomy. A Ladd's procedure was performed and the patient is well at one-year follow up. We suggest prophylactic Ladd's procedure remains the treatment of choice for MMR even when discovered incidentally.
Keywords: Malrotation; Midgut malrotation; Midgut volvulus; Ladd's procedure; Left side appendicitis; Bowel obstruction

S Afr J Surg 2016;54(3)

Introduction

Midgut malrotation (MMR) is a congenital disorder where partial or complete failure of the foetal midgut to rotate around the axis of the superior mesenteric artery (SMA) occurs. Most cases present in the neonate with abrupt onset of bilious vomiting, often associated with abdominal distension.[1] Occasionally the diagnosis is delayed to childhood, where those older than 2 years are more likely to demonstrate chronic symptoms, including nonbilious vomiting, colicky abdominal pain and failure to thrive. [2] It is rare for MMR to present in adulthood as many of these patients remain asymptomatic and are usually discovered incidentally during investigation for other abdominal complaints. [3, 4]

Case report

A 14-year-old female presented to the emergency department with persistent, severe abdominal pain. The pain started periumbilically and progressively worsened. Two days later it localized in the lower abdomen. There was associated gastrointestinal upset with nausea and vomiting. She was well before without prior abdominal symptoms or medical or surgical history. On physical examination, her vital signs were within normal limits. Guarding and rebound tenderness were elicited in the epigastrium and lower abdomen, most pronounced in the right lower quadrant. The white cell count was 7 170 /mm^3 and basic serum biochemistry was normal. Chest radiography did not show signs of a perforated viscus and the abdominal films were normal. The clinical diagnosis of acute appendicitis was made and a laparoscopic appendectomy offered. Operative findings were in keeping with early left sided appendicitis in a setting of MMR without volvulus. The appendix was removed and the MMR was managed conservatively. Histopathology confirmed acute appendicitis. Her postoperative recovery was uneventful and she was discharged after receiving appropriate counseling and informed to return timeously in the presence of danger signs.

Three days later she presented to the emergency unit complaining of non-bilious vomiting and abdominal pain which began the night before. She was acutely ill and dehydrated, with a non-distended abdomen. There was tenderness to deep palpation in the epigastrium. Blood gas analysis revealed a hypochloremic metabolic alkalosis. The plain radiographs had no typical features of midgut volvulus. Contrast-enhanced abdominal computed tomography (CT) showed typical features of MMR with volvulus (Fig. 1). The patient was taken for emergency laparotomy where the diagnosis was confirmed. The duodenojejunal loop did not cross the midline and the caecocolic loop was found in the left upper quadrant, peritoneal bands were obstructing the duodenum, and adhesions surrounded the superior mesenteric artery (SMA). All intestines were viable. A Ladd's procedure was performed. She made an uneventful recovery and was discharged on the sixth postoperative day. Follow-up to one year was uneventful.

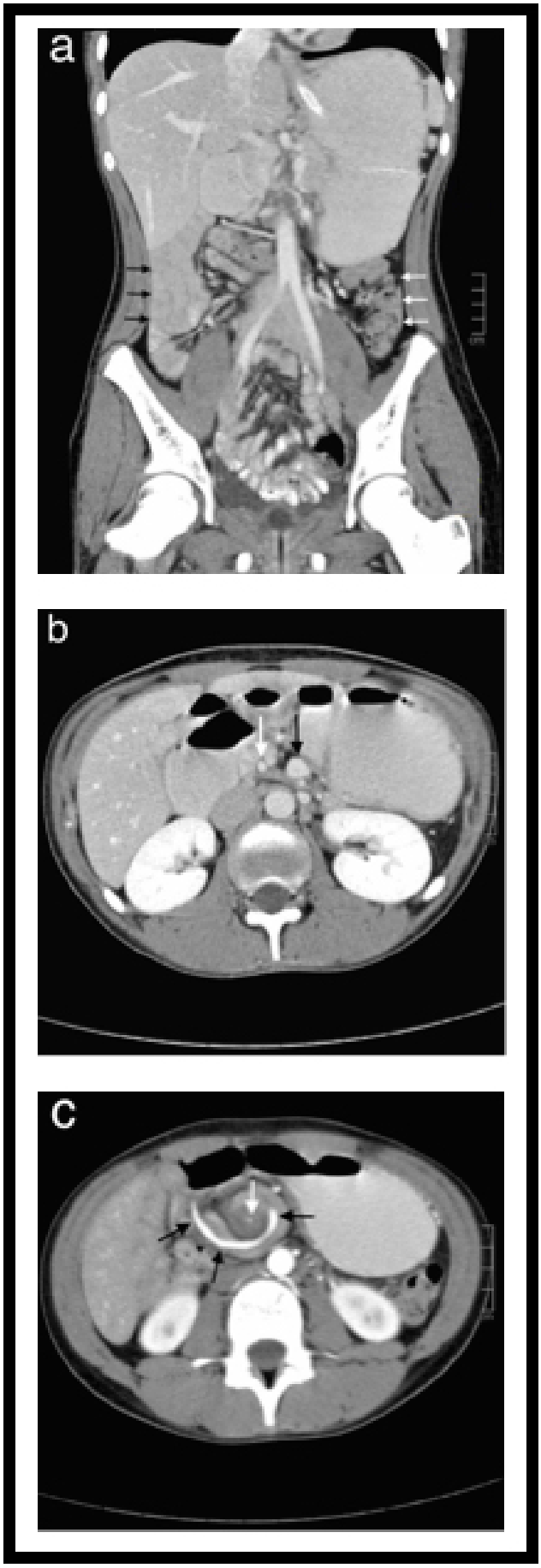

Figure 1. Contrast enhanced computed tomography of the abdomen: ***a*** *Predominantly right-sided small bowel (black arrows) and left-sided colon (white arrows);* ***b*** *Inverted relationship of the superior mesenteric vein (black arrow) situated to the left of the superior mesenteric artery (white arrow) instead of to the right;* ***c*** *The 'whirl sign' demonstrating small bowel loops and mesentery (black arrows) encircling the superior mesenteric artery (white arrow) and vein.*

Discussion

The primitive gut begins to develop at approximately four weeks gestation and can be divided into the foregut, midgut, and hindgut by the end of the fifth week.[1] After a sequence of events that occur between five and twelve weeks, the midgut, which is primarily involved in intestinal rotation around the axis of the SMA, settles in a normally orientated position: the duodenum crossing the midline and lying to the left and posterior of the SMA, with the caecum and ascending colon in the right abdominal gutter.[1,5,6] Depending on its proximal or distal position relative to the SMA, the midgut can be divided into pre-arterial (duodenojejunal) and post-arterial (caecocolic) loops. Classic malrotation is characterised by aberrant positions of both pre-arterial and post-arterial loops.[7] This results in a narrowed mesenteric base with poor posterior fixation, which places the patient at risk for midgut volvulus and the devastating consequences that can follow.[3,8,9]

Plain abdominal films are often undertaken as the initial imaging investigation. Features that are suggestive of malrotation include a gasless abdomen, duodenal 'double bubble' sign, a pattern of small bowel obstruction, and paucity of large bowel gas shadows on the right or small bowel loops on the left.[1,7] Upper gastrointestinal contrast studies are the investigation of choice for suspected MMR.[7,10,11] Findings may include failure of the duodenal C-loop to cross the midline, corkscrew appearance of the duodenum, and small bowel largely confined within the right side of the abdomen.[1,10] Occasionally the results of limited upper gastrointestinal studies are equivocal. In such cases, it is useful to ascertain the position of the caecum, either by delayed upper gastrointestinal images if the patient's condition allows, or immediate contrast enema. The position of the pre- and post-arterial loops can then be used to define the proximal and distal ends of the midgut attachments, and allow an inference about the width of the mesenteric base.[7] When used alone, contrast enema has only a modest ability to exclude MMR.[10,11] CT is also used to diagnose MMR. Typical findings include abnormal position of the small and large bowel, which are found predominantly on the right and left respectively; and inverted position of the superior mesenteric vessels, with the superior mesenteric vein (SMV) situated to the left of the SMA instead of its normal position to the right of the SMA.[1] This reversed mesenteric vessel relationship has led to suggestions that ultrasound (US) may be used to detect MMR. However, the ability of US to diagnose MMR is controversial owing to concerns regarding misleading results: abnormal superior mesenteric vessel arrangement is found in

people without MMR, and not all cases of MMR demonstrate this vessel arrangement abnormality.[7,9]

All symptomatic patients and all infants with MMR should have surgical correction.[8,11] The Ladd's procedure was named after Dr. William Ladd who was the first Surgeon-in-Chief at Boston Children's Hospital and it has remained essentially unchanged since his address to the New Hampshire Medical Society in 1936.[6-8] The procedure entails a laparotomy and detorsion of the midgut volvulus if present, division of the peritoneal bands extending from the caecum to the right abdominal gutter (which traverse the duodenum and often obstruct it), widening of the mesenteric base by division of the adhesions surrounding the SMA, placement of the bowel in a non-rotated position, and appendectomy.[1,7,9] Over the past 20 years there have been reports of laparoscopic Ladd's procedures being performed with good long-term results.[3,9] A retrospective study by Stanfill and colleagues confirmed that laparoscopic repair carries certain well-known advantages over laparotomy, such as lower incidence of wound infection and hernia formation, earlier return to feeds, shorter hospital stay, and lower incidence of post-operative adhesive bowel obstruction.[6] Furthermore, laparoscopy can be particularly useful in cases of asymptomatic MMR where other imaging is equivocal.[9]

Intestinal volvulus is a rare but well documented complication following laparoscopic procedures like appendectomy, cholecystectomy, and colonic resection.[6,12,13,14] Predisposing factors for volvulus following laparoscopy include abnormally mobile bowel (e.g. MMR, mobile caecal syndrome and post bowel mobilisation), previous abdominal surgery with resultant adhesions, induction of pneumoperitoneum, intraoperative patient positioning, and postoperative ileus.[15]

The management of essentially asymptomatic MMR after infancy is controversial.[8,9] Many authors believe that all patients with documented MMR should receive a prophylactic Ladd's procedure.[2,5,8,16] They cite the high morbidity and mortality associated with midgut volvulus, persistence of this risk beyond infancy, difficulty in predicting which patients are most at risk for volvulus, and the efficacy of the Ladd's procedure to significantly diminish the likelihood of this event. Furthermore, many patients who were initially thought to be asymptomatic are found to have underlying abdominal complaints upon detailed enquiry.[17]

Other evidence suggests that asymptomatic MMR after infancy can be treated conservatively with close observation and follow up.[3,11,18] Proponents of this watchful waiting state that the risk of midgut volvulus declines after infancy, with most older children and adults undergoing the Ladd's procedure for chronic abdominal complaints. Malek and Burd performed a statistical decision analysis among asymptomatic patients with MMR and concluded that the maximum quality adjusted life expectancy with surgery occurred at age 20 years and that in patients older than 20 years conservative management had more beneficial effects on life expectancy compared with surgery.[8]

Conclusion

The management of asymptomatic MMR after infancy is controversial. Most authors support operative correction of confirmed cases. However, there is also evidence to suggest that many older patients with asymptomatic MMR derive maximal benefit from non-operative management. Patient education and emphasis on seeking prompt treatment can diminish the risk associated with midgut volvulus. However, it remains difficult to predict which patients will develop intestinal volvulus, and the potential for devastating consequences is a compelling factor that supports early intervention in most cases.

Consent

Written informed consent was obtained from the patient and her guardian for publication of this report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of interest

The authors declare that they have no conflict of interest.

REFERENCES

1. Gamblin TC, Stephens RE, Johnson RK, et al. Adult malrotation: a case report and review of the literature. Curr Surg. 2003;60(5):517-520. [http://dx.doi.org/10.1016/S0149-7944(03)00030-8] [PMID: 14972216]
2. Nagdeve NG, Qureshi AM, Bhingare PD, et al. Malrotation beyond infancy. J Pediatr Surg. 2012;47(11):2026-2032. [http://dx.doi.org/10.1016/j.jpedsurg.2012.06.013] [PMID: 23163993]
3. Nakajima Y, Sakata H, Yamaguchi T, et al. Successful treatment of a 14-year-old patient with intestinal malrotation with laparoscopic Ladd procedure: case report and literature review. World J Emerg Surg. 2013;8(1):19. [http://dx.doi.org/10.1186/1749-7922-8-19] [PMID: 23684081]
4. Birnbaum DJ, Geffroy Y, Goin G, et al. Left side appendicitis with midgut malrotation in an adult. Surg Tech Case Report. 2013;5:38-40. [http://dx.doi.org/10.4103/2006-8808.118627] [PMID: 24470850]
5. Penco JMM, Murillo JC, Hernandez A, et al. Anomalies of intestinal rotation and fixation: consequences of late diagnosis beyond two years of age. Pediatr Surg Int. 2007;23:723–730. [http://dx.doi.org/10.1007/s00383-007-1972-0] [PMID: 17594104]
6. Stanfill AB, Pearl RH, Kalvakuri K, et al. Laparoscopic Ladd's procedure: treatment of choice for midgut malrotation in infants and children. J Laparoendosc Adv Surg. Tech 2010;20(4):369-372. [http://dx.doi.org/10.1089/lap.2009.0118] [PMID: 20218938]
7. Lampl B, Levin TL, Berdon WE, et al. Malrotation and midgut volvulus: a historical review and current controversies in diagnosis and management. Pediatr Radiol. 2009;39:359-366 [http://dx.doi.org/10.1007/s00247-009-1168-y] [PMID: 19241073]
8. Mazziotti MV, Strasberg SM, Langer JC. Intestinal rotational abnormalities without volvulus: the role of laparoscopy. J Am Coll Surg. 1997;185:172-176. [http://dx.doi.org/10.1016/S1072-7515(97)00058-6] [PMID: 9249085]
9. Millar AJ, Rode H, Cywes S. Malrotation and volvulus in

infancy and childhood. Semin Pediatr Surg. 2003;12(4):229-36. [PMID: 14655161]
10. Murphy FL, Sparnon AL. Long-term complications following intestinal malrotation and the Ladd's procedure: a 15 year review. Pediatr Surg Int. 2006;22:326–329. [http://dx.doi.org/10.1007/s00383-006-1653-4] [PMID: 16518597]
11. McVay MR, Kokoska ER, Jackson RJ, et al. The changing spectrum of intestinal malrotation: diagnosis and management. Am J Surg. 2007;194(6):712-717. [http://dx.doi.org/10.1016/j.amjsurg.2007.08.035] [PMID: 18005759]
12. Ly JQ. Malrotation rapidly progressing to midgut volvulus following recent laparoscopic surgery. J Emer Med. 2002;23:295-296. [PMID: 12449969]
13. Lay OS, Tsang TK, Caprini J, et al. Volvulus of the small bowel: an uncommon complication after laparoscopic cholecystectomy. JLAST. 1997;7(1):59-62. [PMID: 9453866]
14. Al Beteddini OS, Sherkawi E. Small bowel volvulus with no malrotation after laparoscopic appendicectomy: Case report and literature review. Int J Surg Case Rep. 2014;5(12):1044-1046. [http://doi: 10.1016/j.ijscr.2014.10.091] [PMID: 25460470]
15. Ferguson L, Higgs Z, Brown S, et al. Intestinal volvulus following laparoscopic surgery: a literature review and case report. JLAST. 2008;18(3):405-410. [http:// doi: 10.1089/lap.2007.0027] [PMID: 18503375]
16. Spigland N, Brandt ML, Yazbeck S: Malrotation presenting beyond the neonatal period. J Pediatr Surg. 1990;25:1139–1142. [http://dx.doi.org/10.1016/0022-3468(90)90749-Y] [PMID: 2273427]
17. Moldrem AW, Papaconstantinou H, Broker H, et al. Late presentation of intestinal malrotation: An argument for elective repair. World J Surg. 2008;32:1426-1431.[http://dx.doi.org/10.1007/s00268-008-9490-3] [PMID: 18347850]
18. Choi M, Borenstein SH, Hornberger L, et al. Heterotaxia syndrome: The role of screening for intestinal rotation abnormalities. Arch Dis Child. 2005;90:813-815.[http://dx.doi.org/10.1136/adc.2004.067504] [PMID: 15890694]