CASE REPORT

A puzzling paradox: Congenital bowel malrotation masquerading as duodenal atresia in a case of non-bilious emesis

Nurjasmine Aida Jamani, MMed[1], Amirah Mohd Lotpi, MBBS[1,2], Radhiana Hassan, MMed[3]

[1]Department of Family Medicine, Kuliyyah of Medicine, International Islamic University Malaysia, Jalan Sultan Ahmad Shah, Bandar Indera Mahkota, Kuantan, Pahang, Malaysia, [2]Ministry of Health, Malaysia, [3]Department of Radiology, Kuliyyah of Medicine, International Islamic University Malaysia, Jalan Sultan Ahmad Shah, Bandar Indera Mahkota, Kuantan, Pahang, Malaysia

SUMMARY

Intestinal malrotation is a congenital abnormal bowel position within the peritoneal cavity, usually involving small and large bowels. It is considered primarily a disease of infancy with infrequent occurrence beyond the first year of life. The twisting and malposition of the intestine can cut off the blood supply. If undiagnosed for a prolonged duration, it can be fatal. Diagnosing malrotation of the gastrointestinal (GI) tract in newborn babies in primary care settings can be challenging, where it needs a structured approach with supportive clinical findings and examinations. Although it can be a straightforward diagnosis, approximately 15% of upper GI (UGI) studies result in equivocal findings, leading to false-positive and false-negative interpretations.[1] We report a case of a five-day-old infant presenting with non-bilious vomiting with radiographic imaging of a double bubble sign, which was subsequently found to have bowel malrotation.

INTRODUCTION

Persistent vomiting in an infant is consequential and sometimes poses a challenge in diagnosis in primary care. An isolated episode of non-bilious vomiting in most infants can be due to milk intolerance, gastro-oesophageal reflux, or gastroenteritis. However, continuous emesis can be due to multifactorial life-threatening conditions such as duodenal atresia, annular pancreas, duodenal stenosis, and bowel malrotation. The diagnosis should be made based on the particular onset, age of presentation, nature of clinical conditions, and proper investigations. A simple reassurance or waiting for the vomiting to be self-limiting is not a wise choice to be made, especially in primary care.

Despite various aetiologies that can cause vomiting in an infant, intestinal malrotation should not be missed. Under one year of age, malrotation presents in around 1 in 2500 live-born infants and is more common in males than females. It can cause an infant to be more likely to develop a midgut volvulus in the first few weeks of life, subsequently causing vascular compromise in the intestines.[2] The cardinal features of intestinal malrotation are bilious vomiting and abdominal distension. However, it can be non-bilious, especially for young children and infants.[3] The clinical manifestations may develop quickly and are generally dramatic enough, especially for infants. They may be in shock, and the conditions are critical for survival.[4]

The management of such cases is a mandatory early surgical intervention. The prognosis is good if there is no midgut volvulus, intestinal necrosis, prematurity, or other abnormalities. This case report aims to increase medical practitioners' suspicion towards congenital malrotation upon seeing cases of infants solely presented with vomiting and the importance of early recognition, preventing delay in diagnosis and treatment.

CASE PRESENTATION

A day-five-of-life newborn baby boy was brought by his mother to the primary care clinic for sudden, frequent vomiting, which was non-projectile and associated with abdominal distension for one day. The vomitus contained milk and saliva. There was no bile, mucus or blood. It was not associated with fever, loose stool, upper respiratory tract infection or head trauma. During the first visit to a primary care clinic, the baby was discharged home, and reassurance was given to the parents. However, he was brought to our clinic the next day because of persistent vomiting. He was born full-term with a birth weight of 3.65kg and was exclusively breastfed. The perinatal and antenatal histories were uneventful.

On examination, the baby was afebrile and active on handling. The anterior fontanelle was normotensive. His vital signs were normal, with good pulse volume, and his capillary refill time was less than 2 seconds with warm peripheries. The abdomen was not distended and was soft on palpation. There was no mass palpable, and the bowel sound was normal. Other systems examinations were unremarkable. He was then subsequently referred to the hospital for further management, given the possible diagnosis of intestinal obstruction.

An abdominal radiograph was done, and normal findings were noted. He was then admitted for observation and hydration maintenance. However, while in the ward, the vomiting persisted, and abdominal distension was observed.

This article was accepted: 15 October 2024
Corresponding Author: Amirah Mohd Lotpi
Email: amirahlotpi@gmail.com

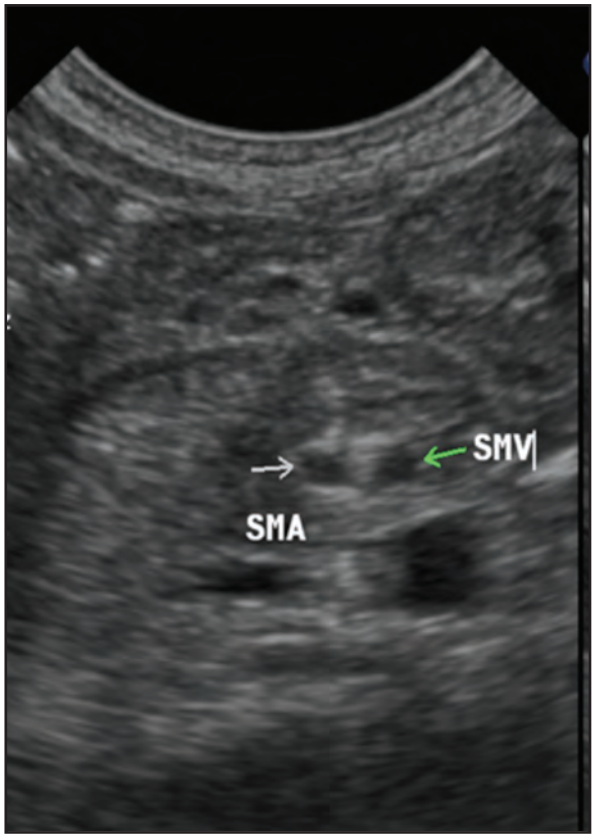

Fig. 1: Ultrasound in axial plane showing the reverse superior mesenteric artery (SMA) and superior mesenteric vein (SMV) orientation

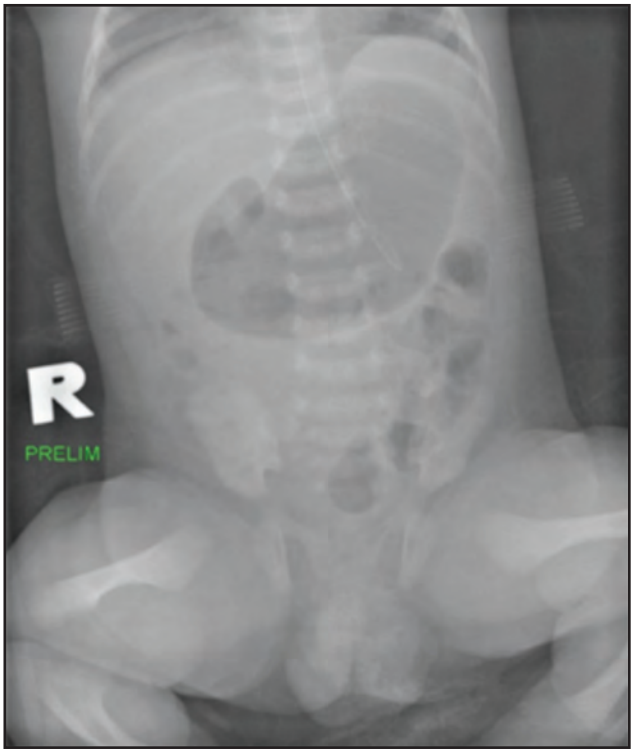

Fig. 2a: A preliminary abdominal radiograph shows air-filled distended stomach with minimal gas distally

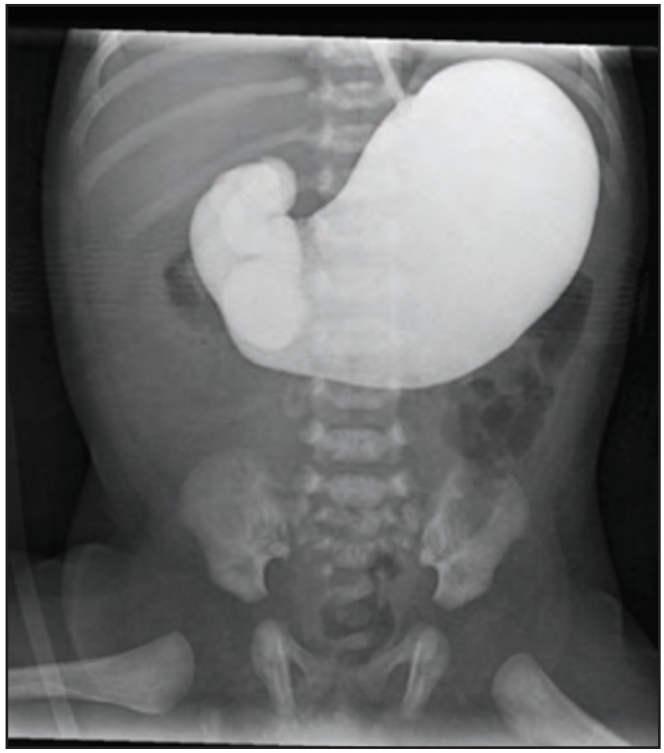

Fig. 2b: Contrast is seen within the distended stomach. Features are suggestive of subtotal duodenal stenosis with presence of distal gas observed

A subsequent ultrasound of the abdomen was performed, showing a suspicious superior mesenteric artery (SMA) and superior mesenteric vein (SMV) relationship inversion (Figure 1). However, no apparent sonographic features suggest midgut volvulus with malrotation. Another abdominal radiograph was done that showed distended stomach with air in distal bowel loops which otherwise, were not dilated. After that, a UGI contrast study was performed, which confirmed malrotation. Other biochemical markers, blood and urine cultures were normal.

He was then referred to the pediatric surgeon for further management. A Ladd's procedure was performed. Intraoperative findings showed that the duodenojejunal (DJ) flexure is on the right side of the vertebra, with no volvulus but narrowed mesentery. Otherwise, other intraoperative findings, such as the location of the small bowel, large bowel and caecum in this patient, cannot be described further. He was discharged well post operatively. During the follow-up at the age of one month, the baby tolerated feeding well and had satisfactory weight gain. There have been no more vomiting episodes since then.

DISCUSSION

A newborn baby with vomiting is worrisome, and parents will seek immediate medical advice. Vomiting in newborn babies should not be taken lightly. It can happen due to multiple pathologies such as intestinal obstructions, viral gastroenteritis, gastroesophageal reflux disease, pyloric stenosis, Hirschsprung disease and sepsis.[5] In addition, vomiting may result from congenital atresia or an error in embryonic rotation, resulting in malrotation with or without volvulus.[6]

Depending on the onset, quality, frequency, and associated symptoms, medical practitioners must determine whether the infant needs further investigation or sufficient reassurance only. In this case, the healthcare provider attending to the patient during the first visit to the primary care clinic must take a thorough history to rule out all the differential diagnoses mentioned above. This is crucial so that delayed diagnosis can be avoided and the patient can be managed accordingly promptly. In this case, after the second visit to the primary care clinic, the infant was immediately referred to the nearest tertiary centre for further management.

The most significant complication of intestinal malrotation is midgut volvulus, which is life-threatening and may lead to short-bowel syndrome.[6] In addition, a higher rate of complications such as bowel ischemia occur in children, including infants, given a larger fraction of patients in this age group are brought to the primary care clinics with acute presentations. Thus, a physician must have a high index of suspicion based on age, symptoms, and physical examinations before proceeding to perform the necessary investigations.

As mentioned earlier, malrotation diagnosis can pose a challenge in the primary care setting. Clinical presentations for malrotation can occasionally be confused with duodenal atresia (DA).[7] Nevertheless, in DA, infants will present with bilious vomiting in the first 48 hours of life. Clues to differentiating malrotation and DA include antenatal history and radiographic features such as a smooth outline to the most distal point of a double bubble (DA) rather than the 'bird's beak', which suggests malrotation.

In addition, the inversion of the superior mesenteric artery and vein relationship may indicate intestinal malrotation. From the axial plane, at the level of the junction of the superior mesenteric vein (SMV) with the portal vein, the SMV is usually located to the right of the superior mesenteric artery (SMA). Thus, an SMV not lying to the right of the SMA is highly sensitive to intestinal malrotation (Daneman, 2009). As in our case, the reverse orientation of SMV and SMA was suggestive towards intestinal malrotation. Also, an upper gastrointestinal contrast study is 93-100% sensitive and will show a corkscrew appearance when a volvulus is present. The contrast study also helps to define the position of the duodenal-jejunal flexure, the cecum and the proximal colon.[8] However, any suspicion of malrotation warrants emergency investigation and/or operative intervention.[9] Direct visualization of a whirlpool sign helps to establish the diagnosis.[8] In this case, even though it was non-bilious vomiting, the symptom was persistent, followed by abdominal distension after day five of life, which led towards the diagnosis of intestinal malrotation. The management would remain the same.

Ultrasound (US) is crucial in the diagnostic imaging workup for infantile vomiting.[7] Workup for intestinal malrotation should be considered in all patients with bilious emesis, abdominal pain or distention. As in our case, the US abdomen had been done during admission, after the initial abdominal radiograph at the emergency department which showed normal findings. However, malrotation could not be ruled out; thus, a UGI contrast study was performed before surgery was decided upon. An abdominal radiograph was also repeated prior to the UGI contrast study. This also shows that other alarming pathologies still need to be ruled out, even with normal radiographic findings during the first visit. For this condition, the management focuses on avoiding intestinal necrosis and reversing ischemia if it already happens. This is achieved with Ladd's procedure, named after Dr William Edward Ladd, the pioneer pediatric surgeon of North America who first performed the procedure in 1936. Instead of correcting the malrotation, it helps to open the narrow mesenteric pedicle to prevent volvulus from recurring.[5] There is also a debate on whether to choose laparoscopic or open surgery. However, laparoscopic exploration is the procedure of choice compared to open surgery in that it shortens hospital stays and increases the recovery rate.[9] Nevertheless, it will be converted to an open procedure if there is evidence of intestinal necrosis present or difficult local anatomy. The mean hospital stay was 4 days (range 3-12days).[10] In this case, the baby was discharged well.

One of the most common complications that can happen post-operatively is intestinal obstruction. There are also cases of readmission within six months after Ladd's procedure for different problems such as wound infection, feeding difficulties, ascites, pneumonitis, constipation, and abdominal pain. Cases of failure to thrive or

gastroesophageal reflux disease (GERD) also need to be given attention. As for this case, parental satisfaction was high, and no long-term morbidity was observed during the subsequent follow-up with the paediatric surgery department. These complications also need to be highlighted during routine check-ups at primary care.

CONCLUSION

A definitive diagnosis of malrotation can be challenging in the primary care setting with limited resources. For primary care providers, an accurate clinical judgment for a case of vomiting in an infant is a must. Any infant with symptoms and signs of acute intestinal obstruction must be investigated further. Reassurance does not suffice. Although this case presented with non-bilious vomiting, intestinal malrotation remained one of the differential diagnoses. Intestinal malrotation must be considered, even though it is rarely found in daily practice, as the final diagnosis is commonly established at the tertiary centre. In a primary care setting, one should never rely only on plain radiograph findings to diagnose or exclude intestinal malrotation. Appropriate referrals should be made to prevent diagnosis delay and complications.

ACKNOWLEDGEMENT

The authors would like to thank the patient and his parents for agreeing to publish this case.

DECLARATION

The authors have no conflict of interest to disclose.

REFERENCES

1. Applegate KE. Clinically Suspected Malrotation in Infants and Children: Evidence-Based Emergency Imaging. In 2018. p. 583–95. Available from: http://link.springer.com/10.1007/978-3-319-67066-9_37
2. Coste AH, Anand S, Nada H, Ahmad H. Midgut Volvulus [Internet]. StatPearls. 2024. Available from: http://www.ncbi.nlm.nih.gov/pubmed/14655161
3. Nguyen D, Sessions W, Deitrick J, Olanrewaju A, Meller J. Nonbilious Vomiting in a 4-Week-Old Male: A Case Report and Review of the Literature. Glob Pediatr Heal [Internet]. 2018 Jan 1;5:2333794X1775101. Available from: http://journals.sagepub.com/doi/10.1177/2333794X17751010
4. Alazraki AL, Rigsby CK, Iyer RS, Bardo DME, Brown BP, Chan SS, et al. ACR Appropriateness Criteria® Vomiting in Infants. J Am Coll Radiol [Internet]. 2020 Nov;17(11):S505–15. Available from: https://linkinghub.elsevier.com/retrieve/pii/S154614402030939X
5. Mitsunaga T, Saito T, Terui K, Nakata M, Ohno S, Mise N, et al. Risk Factors for Intestinal Obstruction After Ladd Procedure. Pediatr Rep [Internet]. 2015 May 25;7(2):5795. Available from: https://www.mdpi.com/2036-7503/7/2/5795
6. Heidsma CM, Hulsker CCC, van der Zee D, Kramer WH. [Malrotation with or without volvulus]. Ned Tijdschr Geneeskd [Internet]. 2015;159:A8859. Available from: http://www.ncbi.nlm.nih.gov/pubmed/26173661
7. Adams SD, Stanton MP. Malrotation and intestinal atresias. Early Hum Dev [Internet]. 2014 Dec;90(12):921–5. Available from: https://linkinghub.elsevier.com/retrieve/pii/S0378378214002424
8. Daneman A. Malrotation: the balance of evidence. Pediatr Radiol [Internet]. 2009 Apr 24;39(S2):164–6. Available from: http://link.springer.com/10.1007/s00247-009-1152-6
9. Svetanoff WJ, Srivatsa S, Diefenbach K, Nwomeh BC. Diagnosis and management of intestinal rotational abnormalities with or without volvulus in the pediatric population. Semin Pediatr Surg [Internet]. 2022 Feb;31(1):151141. Available from: https://linkinghub.elsevier.com/retrieve/pii/S1055858622000026
10. Ferrero L, Ahmed Y Ben, Philippe P, Reinberg O, Lacreuse I, Schneider A, et al. Intestinal Malrotation and Volvulus in Neonates: Laparoscopy Versus Open Laparotomy. J Laparoendosc Adv Surg Tech [Internet]. 2017 Mar;27(3):318–21. Available from: http://www.liebertpub.com/doi/10.1089/lap.2015.0544