Radiology Case Reports 19 (2024) 6108–6111

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Late presentation of midgut malrotation with obstruction in a 5-year-old female: A case report☆,☆☆

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ARTICLE INFO

Article history:
Received 2 September 2024
Revised 11 September 2024
Accepted 13 September 2024

Keywords:
Intestinal obstruction
Malrotation
Whirlpool Sign
Ladd's procedure
Pediatric emergency

ABSTRACT

Midgut malrotations are rarely diagnosed beyond infancy. Delays in recognition and diagnosis can result in death. Here, we report the case of a 5-year-old girl who presented with a 1-year history of intermittent abdominal pain and vomiting. An abdominal computed tomography scan with contrast confirmed the diagnosis of midgut malrotation with obstruction; therefore, the Ladd procedure was performed, and the child was discharged uneventfully. Clinicians must maintain a high level of suspicion because this diagnosis is unusual in this age group.

Introduction

Intestinal malrotation is a rare congenital anomaly that occurs when the midgut fails to completely rotate during embryonic development [1,2]. Malrotation is classified as "typical" when the duodenojejunal junction is positioned to the right of the midline and "atypical" when it is placed on the left side but does not rise above the pylorus [1,2]. It has an incidence of approximately 1 in 500 live births [2–4]. Over 75% of patients are symptomatic within the first month of their lives, and 90% develop symptoms within the first year [2–4].

Here, we present a case of nonclassic malrotation anomalies in a 5-year-old girl who had intermittent abdominal pain for 1 year.

Case report

A 5-year-old girl was brought to our emergency department (ED) with complaints of intermittent abdominal pain and vomiting that had worsened in the last 2 days. The patient was healthy until 1 year prior when she began to have episodes of

Abbreviations: CT, computed tomography; ED, emergency department; OR, operating room.

☆ Competing Interests: The authors declare that they have no competing interests.

☆☆ Acknowledgments: The authors would like to thank the parents who gave us the permission for writing this case. Funding: Not applicable.

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https://doi.org/10.1016/j.radcr.2024.09.064

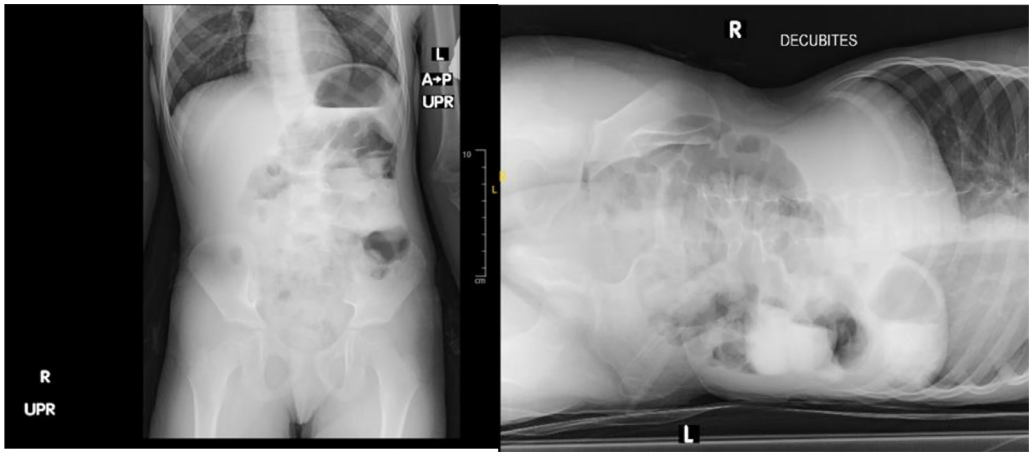

Fig. 1 – Abdominal plain radiographs showed multiple air fluid levels with numerous dilated bowel loops.

abdominal pain. Her mother added that these symptoms were periodic and would last for 3-5 days once monthly; each time she sought medical advice, they treated her for constipation and discharged her safely home.

Over the past 3 days, her condition worsened, and she experienced an increased frequency of vomiting. Vomiting was described as a small amount, 9-10 times per day, nonprojectile, nonbilious, and nonbloody. Abdominal pain was moderate to severe, colicky, and localized in the umbilical area with no radiation, and no aggravating or relieving factors. The patient had not passed stool for a couple of days. The mother had no history of urinary symptoms, fever, trauma, medication intake, or surgery. She also reported a normal pregnancy and an uneventful delivery of her child. The girl had received all immunizations according to local guidelines.

Upon arrival at the ED, the patient appeared uncomfortable but not ill. She was alert, conscious, and oriented. The patient was well-hydrated and not in acute distress. Her vital signs were initially unremarkable for her age: Temperature 36.9°C; Heart rate 115 beats per minute; Blood pressure 108/69 mmHg; Respiratory rate 22 breaths per minute; and oxygen saturation 100% on room air. An abdominal examination revealed tenderness throughout the abdomen and guarding, with no signs of peritonitis. Additional examinations, including of the neurological, cardiovascular, and respiratory systems, did not reveal any abnormalities. Abdominal plain radiographs were performed and showed multiple air fluid levels with numerous dilated bowel loops (Fig. 1).

The complete blood count and basic metabolic, liver, and renal profiles were within normal limits. During this time, the patient experienced bilious vomiting with the same abdominal examination findings. Therefore, intestinal obstruction was considered the most likely diagnosis. The patient was kept nil per mouth (NPO), a nasogastric tube was inserted to decompress the stomach, she was resuscitated with fluids, and antibiotics were administered.

In the ED, abdominal computed tomography (CT) with contrast showed multiple dilated jejunal loops with a twisted mesentery, giving a whirlpool-like appearance in the right upper abdomen, with the duodenojejunal flexure in the right midline, raising the suspicion of small bowel obstruction due to midgut volvulus (Fig. 2).

Pediatric surgery was consulted. The patient was immediately transferred to the operating room for laparotomy because of the suspected volvulus of the midgut. Intraoperatively, dilated jejunal loops and a twisted mesentery with the duodenojejunal flexure in the right midline and congenital adhesions were observed. Ladd's procedure was performed to maintain gastrointestinal patency and release congenital adhesions. Postoperatively, she did well and tolerated a diet on postoperative day 7. She was discharged on postoperative day 9 with oral analgesia and antibiotics. At surgical outpatient follow-up 1 week later, she had no complaints.

Discussion

Malrotation presents less frequently after the first year of life [2]. Consequently, the diagnosis might be underestimated owing to the varied presentations [2]. The clinical presentation in older children can be either acute or chronic, with nonspecific manifestations that include intermittent episodes of abdominal cramps and frequent obstructive symptoms, along with failure to thrive [2,5]. Moreover, 40% of children with malrotation presenting to the emergency department experience nonbilious vomiting, which may make the diagnosis more difficult for physicians, especially in children over 1 year of age. [6].Patients usually present with nonspecific symptoms that delay the diagnosis [2,6]. Our case presented with intermittent abdominal pain associated with vomiting, which is like that reported in the literature [7–9]. Mir et al. [10] reported that a high index of suspicion is required to diagnose this uncommon pathology when patients present with intermittent abdominal pain and discomfort after meals.

Plain abdominal radiography is considered the initial investigation and might detect some of the features of malrotation, including the double bubble sign, unusual gas patterns, and air-fluid levels, like our case and other reported cases [9,10]. Specific findings suggesting malrotation can be

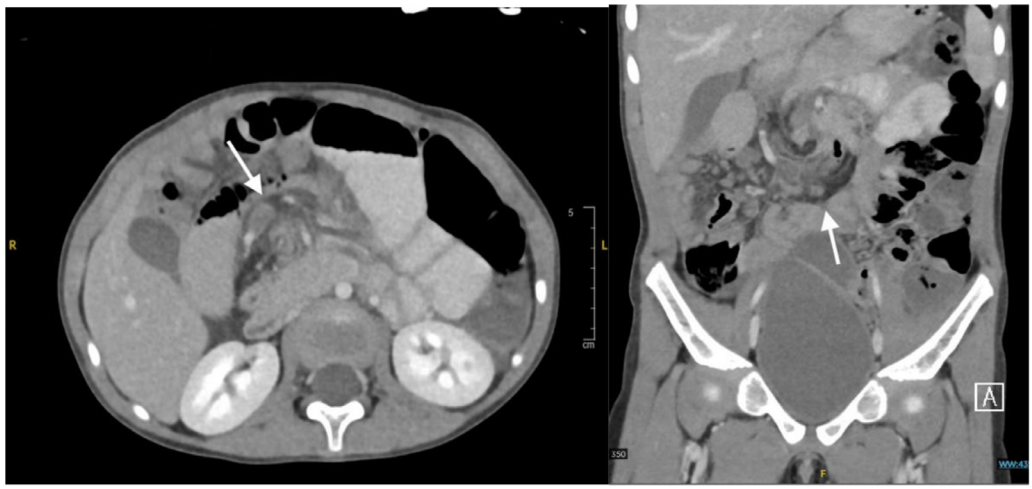

Fig. 2 – Abdominal computed tomography (CT) with contrast: A coronal and sagittal view showed multiple dilated jejunal loops with twisted mesentery, giving a whirlpool-like appearance with the duodenojejunal flexure in the right of the midline.

detected using abdominal CT, mesenteric artery angiography, and gastrointestinal barium studies [10]. In our case, we performed an abdominal radiograph as the first-line investigation, as shown in (Fig. 1), with findings confirmed using abdominal CT with contrast, as shown in (Fig. 2), with similar findings reported by Laham et al. and Gupta et al. [8,9]. Moreover, one other helpful finding in diagnosing a midgut volvulus is the inverted relationship between superior mesenteric artery (SMA) and superior mesenteric vein (SMV) (SMV normally on the left side of the SMA on the abdominal CT). Although this is not always reliable.

A more useful sign to rule out intestinal malrotation on abdominal CT and ultrasonography (US) is the demonstration of the retroperitoneal D3 duodenal segment, which normally should be seen in a transverse plane between the SMV and the aorta [11].

Management principles for pediatric patients who present with acute malrotation and hemodynamic instability should include aggressive hydration and urgent pediatric surgical consultation for exploratory laparotomy as the first line of management [6]. Ladd's procedure remains the mainstay of surgical treatment, with similar surgical procedures in reported cases [6–9]. Intra-operative findings in our case were dilated jejunal loops and a twisted mesentery with duodenojejunal flexure on the right side of the midline and congenital adhesions, with different findings reported in different case reports. Emmerson et al reported a 5-year-old boy with intra-operative firings of malrotation with dusky but viable bowel with midgut volvulus [7]; Laham et al. [8] reported a 15-year-old girl with intra-operative findings of malrotation, stenosed duodenum, and extensive adhesions, similar to our case. Gupta et al. [9] have a case series and reported 2 pediatric cases of a 13-year-old girl and a 17-year-old boy with intra-operative findings of stenosis at the fourth part of the duodenum, cecum, and ascending colon on the left side and cecum and appendix in the subhepatic region, hernia sac, with inter-bowel adhesions. Most reported cases underwent laparotomy and Ladd's procedure and were discharged with an uneventful recovery, like our patient [7–9].

These cases illustrate the variability in presentation, ranging from chronic intermittent symptoms to acute severe obstruction, and underscore the effectiveness of Ladd's procedure in resolving midgut malrotation and volvulus across different age groups and clinical scenarios.

Conclusion

Intestinal malrotation is rare beyond infancy. These symptoms resemble those of other illnesses and pose a challenge for diagnosis. Clinicians must maintain a high level of suspicion because malrotation is a less likely diagnosis in older children.

Ethics approval and consent to participate

This study has been approved by the Institutional Research Ethics Committee in King Fahad Medical City in Riyadh, Kingdom of Saudi Arabia (IRB00010471) . An informed consent for participation and publication of identifying images or other personal or clinical details was obtained from the parents.

Consent for publication

Informed consent for publication of identifying images or other personal or clinical details was obtained from the parents.

Availability of data and materials

Data that support the findings in the current study are available from the corresponding author on reasonable request.

Author contributions

Idea development and case report writing: Tahani AlHarshan, Ibtihal Almeshawi. Drafting the manuscript: Ibrahim AlWakid. Final writing up and critical revision of paper: Yara AlGoraini. Submission of the manuscript: Yara AlGoraini.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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