Malrotation of gut with superior mesenteric artery syndrome and multiple jejunal diverticula presenting as acute intestinal obstruction in 6th decade: A rare case report

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ABSTRACT

INTRODUCTION: Intestinal malrotation is a disease of neonates and young children presenting as acute intestinal obstruction. Presentation of malrotation in elderly patients with intestinal obstruction is quite rare with only a few cases reported in literature. We report a case of intestinal malrotation presenting as acute obstruction in sixth decade.

PRESENTATION OF CASE: A 55 year old male presented to the emergency with features of acute intestinal obstruction. Imaging studies revealed intestinal malrotation. Exploratory laparotomy revealed malrotation with compression of 3rd part of duodenum and terminal ileum by superior mesenteric artery with multiple jejunal diverticula. Bypass procedures (duodeno-jejunostomy and ileo-colic anastomosis) with appendicectomy were done.

DISCUSSION: Malrotation of gut is an anomaly usually presenting in neonatal period with complications such as midgut volvulus. Presentation in adult age is rare with most cases being asymptomatic. Ladd’s procedure is the operation of choice with division of the Ladd’s bands and appendicectomy being performed.

CONCLUSION: Surgeons should keep a flexible approach in management of malrotation of gut presenting in late stages of life as more and more clinical variants to the presentations described in literature are being encountered.

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1. Introduction

Intestinal malrotation is a disease of neonates which arises due to anomalous intestinal rotation in fetal life and is usually present in the first month. Presentation in elderly age is quite rare.

Most patients present in the first month of life with bilious vomiting. Adult patients usually present with acute bowel obstruction, associated with intestinal ischemia and midgut volvulus or with chronic obstruction with non-specific abdominal complain like chronic pain. The true incidence of malrotation of gut presenting in adulthood is unknown because majority of patients remain asymptomatic throughout life. Adult midgut malrotation is very rare and its incidence has been reported to be between 0.0001% and 0.19%. A literature review by von Flue et al. cites 40 cases from 1923 to 1992.1,2

We present a case of a 55 year old male presenting with features of acute intestinal obstruction.

2. Case report

2.1. Presenting concerns

A 55 year old male was presented in an emergency department, with complains of severe, generalized, and colicky pain in the abdomen, with distension and multiple episodes of foul smelling vomiting since last 4 days. The patient also complained of non-passage of flatus and motion since the last 2 days (Table 1).

There was no history of previous surgical intervention or any medical comorbidity. Patient was a smoker since last several years.

2.2. Clinical findings

On examination, patient had tachycardia with a pulse rate of 106 bpm, blood pressure 116/84 mm of Hg, and afebrile with a res-
Table 1
Hospital course of the patient.

| Timeline         | Event                                      |
|------------------|--------------------------------------------|
| Pre op day 4     | Onset of abdominal pain and distension with vomiting |
| Pre op day 2     | Onset of absolute constipation, admission, and diagnostic evaluation |
| Day 0            | Exploratory laparotomy                      |
| Post op day 2    | Jejunostomy feeds started                   |
| Post op day 5    | Nasogastric tube removed and oral feeds started |
| Post op day 7    | Abdominal drain removed, patient discharged |
| Follow up        | 2 weeks, 1 month. Patient doing well        |

piratory rate of 20 per minute. The abdomen was distended; mild generalized tenderness to deep palpation was present with voluntary guarding. Bowel sounds were absent. Per rectal examination was unremarkable.

2.3. Diagnostic focus and assessment

The blood investigations revealed mildly raised urea and creatinine levels, most probably due to dehydration. Hemoglobin, white cell count, and rest of the routine investigations were within normal limits. Abdominal x-rays revealed multiple air fluid levels suggestive of intestinal obstruction. Patient was managed with nasogastric decompression and intravenous fluid support but there was no improvement in symptoms. The nasogastric tube output was consistently on the higher side being around 1.5–2 l in 24 h. A contrast enhanced abdominal CT scan revealed clockwise swirling of the SMA and its branches (whirlpool sign) with small bowels lying predominantly on the right side and large bowel predominantly on the left side (Fig. 1). The caecum and ileocaecal junction were said to be present in left iliac fossa. The CT also revealed gross dilatation of the stomach and 1st part of duodenum with tapering of the 3rd and 4th parts at the site of swirling of the vessels. There was a little passage of oral contrast into the small bowel. A possibility of midgut volvulus was suggested to be ruled out by barium study.

2.4. Therapeutic focus and assessment

The patient’s physical condition worsened with rising pulse rate and abdominal distension; hence it was decided to perform an exploratory laparotomy without going for the barium study. On exploration, nonrotation of the gut was found with the small intestines lying to the right of the superior mesenteric artery and large bowel to the left (Figs. 2–5). The caecum was present in midline superior to the umbilicus with the ileocaecal junction slightly to the left of midline. The stomach was dilated. The third part of the duodenum was passing posterior to the superior mesenteric artery and was compressed by the SMA pedicle. The terminal ileum also passed posterior to the superior mesenteric artery pedicle from right to left ending in the ileocaecal junction. The terminal ileum was also being compressed by the SMA pedicle. The appendix was long and thick. There were multiple jejunal diverticula in the proximal jejunum on the mesenteric side. No evidence of any Ladd’s band, midgut volvulus, or bowel gangrene was found. The entire duodenum was mobilized by an extensive Kocher’s maneuver. Duodenjejunostomy was fashioned between the second part of duodenum and proximal jejunum to bypass the obstruction. Another side to side anastomosis between terminal ileum and transverse colon was done to bypass the obstruction at the level of terminal ileum. Appendicectomy was performed and a feeding jejunostomy was fashioned distal to the duodeno-jejunal anastomosis.
3. Discussion

Intestinal malrotation is a rare disease and is more rarely present in late adult life. The usual age of presentation is in neonatal period and early childhood. Seldom have been the cases of malrotation presenting in adulthood as obstruction been reported. What adds to the exceptionality of this case is that, apart from malrotation, other interesting accompanying findings were present. First was the compression of duodenum by the SMA pedicle and second was passage of terminal ileum behind the SMA pedicle. Another incidental finding was the presence of multiple jejunal diverticula. There are no previous reported cases of intestinal malrotation presenting in adulthood with compression of both the duodenum and terminal ileum by the SMA pedicle.

On extensive search of literature, we could only find one reported case of malrotation with terminal ileum passing behind the SMA pedicle and multiple jejunal diverticula.

Midgut malrotation is mainly a deviation from the normal 270° counter clockwise rotation of the gut which occurs during embryonic life. The embryonic gut is in the form of a straight tube by the end of 4th week of life. During the 5th week, the vascular pedicle to the gut develops and the gut can now be divided into foregut, midgut, and hindgut based on the anatomic blood supply. The midgut is supplied by the superior mesenteric artery. Due to rapid elongation of the midgut, there is physiological herniation of the bowel loops into the umbilical cord with return to abdominal cavity 4–6 weeks later. It is during this return of midgut into the abdominal cavity, the rotation of intestines occurs. The rotation of the intestines can be divided into three stages. Stage I, occurring in weeks 5–10, includes the physiological herniation, a 90° counter clockwise rotation, and return of the midgut into the abdominal cavity. Stage II involves further counter clockwise rotation making a total of 270° and occurs during 11th week. This results in bringing of the duodenal ‘c’ loop behind the SMA and the placement of ascending colon to the right, transverse colon above, and descending colon to the left. Stage III involves the fixation and fusion of the mesentery.

Intestinal anomalies can be classified based on the stage of rotation during which they occur. Stage I anomalies involve omphaloceles; stage II anomalies involve nonrotation, malrotation, and reverse rotation while stage III anomalies include unattached duodenum, mobile caecum, and unattached small bowel mesentry.

The diagnosis is usually made on radio graphical findings. The imaging modality of choice is an UGI contrast study. It demonstrates an abnormal position of the ligament of Treitz along with the appearance of a bird’s beak in the third portion of the duodenum, which indicates an obstruction. The accuracy of the upper gastrointestinal series (UGI) is reported to be over 80% in diagnosing malrotation in the child and adult. Ultrasonography and particularly using color doppler is useful in diagnosing midgut volvulus in which the normal relation of the superior mesenteric vessels is altered or reversed (normally the SMV is to the right of SMA). The abdominal CT findings include a whirlpool sign in the small bowel mesentery representing the twisting of the mesentery around the SMA and also the altered relationship of the SMA and SMV.

The treatment of a symptomatic malrotation is primarily surgical. Midgut volvulus is a surgical emergency. Ladd’s procedure is the operation of choice. In patients not presenting with volvulus, the surgical options include division of the Ladd’s bands with an appendicectomy being invariably performed. The gut is usually repositioned back into the abdominal cavity with the small bowel in the right side and large bowel occupying the left side.

In our case, there might be an arrest of rotation of duodenojejunal loop after partial rotation around the SMA pedicle leading to non-passage of caecum behind the artery and its normal descent.
4. Conclusion

Presentation of malrotation as small bowel obstruction is rare in adults. It is becoming increasingly frequent to encounter variations from the classically described presentations of malrotation, like in our patient, who neither had midgut volvulus nor Ladd’s bands. As no standard operation mentioned in the literature or textbooks covers the treatment of a typical malrotation, surgeons should keep a flexible approach.

Conflict of interest

No conflicts of interest.

Funding

No sources of funding.

Ethical approval

Ethical approval not required.

Author contributions

Dr. Dhananjay Saxena – study design, data acquisition, data analysis, writing, revision. Dr. Abhinav Pandey – revision, editing. Dr. Rana Arun Singh – data collection, analysis. Dr. Prashant Garg – study design, revision. Dr. Rahul Roy – writing, revision. Dr. Rajendra Prasad Bugalia – data collection, design, revision. Dr. Amit Goyal – article revision. Dr. Jeevan Kankaria – design, revision. Dr. R. K. Jenaw – data analysis, article revision, approval.

Consent

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

Guarantor

Dr. Dhananjay Saxena, the contributing author, is the guarantor.

References

1. Von Flue M, Herzog U, Ackermann C, Tondelli P, Harder F. Acute and chronic presentation of intestinal nonrotation in adults. Dis Colon Rectum 1994;37:192–8.
2. Wang C, Welch C. Anomalies of intestinal rotation in adolescents and adults. Surgery 1963;54:839–55.
3. Horwood J, Akbar F, Maw A. A typical midgut malrotation presenting as chronic bowel obstruction in the eighth decade. BMJ Case Rep 2009, 2009: bcr08.2008.0631.
4. Dott NM. Anomalies of intestinal rotation: their embryology and surgical aspects with report of five cases. Br J Surg 1923;42:251.
5. Gohl ML, DeMeester TR. Midgut nonrotation in adults. Am J Surg 1975;129:319–23.
6. Spigland N, Brandt ML, Yazbeck S. Malrotation presenting beyond the neonatal period. J Pediatr Surg 1990;25:1139–42.