SURGICAL GRAND ROUND

Intestinal Malrotation: Presentation in the Older Child

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Introduction

Intestinal malrotation is not an uncommon condition but diagnosis is often unsuspected or delayed. We present an older child with intestinal malrotation to high light important aspects of diagnosis and management.

Case Presentation (Dr. Sabiu)

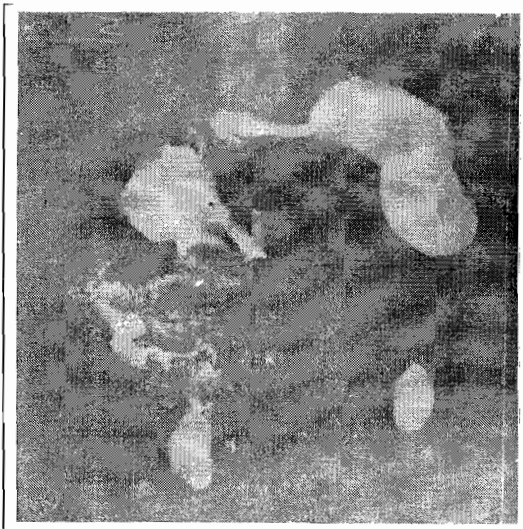
A Five-year-old boy presented with a one-week history of colicky abdominal pain, bilious vomiting and constipation. He was able to pass stool on the day of presentation, which was non-bloody and non-mucoid. There was no history of malaena stool. He had been having episodes of bilious vomiting since the age of three weeks lasting one to two days. No history of chronic diarrhoea and developmental milestone were normal.

Physical examination revealed severe dehydration, afebrile and small for age. The abdominal was distended and there were peristalsis but no tenderness. No mass was palpable. Bowel sounds were exaggerated. Rectal examination was normal. The working diagnosis was intestinal obstruction secondary to malrotation with midgut volvulus. Other differentials included congenital bands, intussusception and parasitic infestation.

Plain Abdominal radiographs showed (erect and supine) paucity of gas shadows in the bowel. Full blood count showed packed cell volume of 31%, white cell count 4.8 x 10^9/L. Serum urea and electrolytes showed urea of 1.4 mmol/l (2.5-.6.5), sodium 120 mmol/L (136.-145), potassium 1.5 mmol/L (3.6-5.2), chloride 70mmol/L (94 - 108) and bicarbonate 32mmol/L (24 - 32).

The patient was resuscitated with intravenous fluid, nasogastric tube decompression and broad-spectrum antibiotics. Abdominal distension subsided, vital signs became stable, and visible peristalsis disappeared. The patient was able to open his bowel by this time. An upper gastrointestinal series was then done and it showed the duodenojejunal junction on the right side of the midline and jejunum and ileum also on the right side (Figure 1).

Figure 1: Upper Gastrointestinal Series Showing the Duodenojejunal Junction and Upper Small Bowel on the Right Side of the Midline



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At laparotomy, the peritoneal cavity was clean. The caecum was in the left hypochondrium and duodenojejunal junction in the right hypochondrium with a Ladd's band extending from the caecum over the second part of duodenum and getting attached to the right posterior peritoneum. The mesentery had a very narrow and was oedematous and thickened with multiple lymph nodes. A Ladd's procedure was performed. Postoperative course was uneventful and the patient has remained well at 9 months of follow up.

Discussion

Malrotation refers to a condition in which the midgut, that part of the intestine supplied by the superior mesenteric vessels extending from the duodenojejunal flexure to mid transverse colon remains unfixed and suspended on a narrow based mesentery.[1] The diagnosis and management of malrotation requires a thorough knowledge and understanding of embryology of intestinal rotation and fixation and also an understanding of the varied presentation of this condition.

Malrotation has a variety of clinical presentations and may be associated with other anomalies, including jejunal and duodenal atresia, oesophageal atresia, prune belly syndrome and Hirschsprung's disease. Malrotation is an intrinsic component of diaphragmatic hernia and omphalocoele and gastroschisis.[2]

Embryology

Rotational anomalies occur when the proximal and distal loops of the midgut fail to complete their required 270° anticlockwise rotation during return of the midgut from its ventral extracoelomic herniation. This normally occurs between ten and twelve weeks of gestation. [3] Rotation and fixation normally vary in degree from " non rotation" in which the proximal loop lacks 90° of normal rotation leaving duodenojejunal junction with ascending colon and caecum remaining on the left side of the peritoneal cavity. It is commonly associated with abdominal wall defects and diaphragmatic hernia.[2] In incomplete rotation or clinically "significant malrotation", the duodenojejunal loop remains to the right of the spine but colonic loops rotate 180° passing in front of the superior mesenteric artery and failing to descend to the right lower quadrant. This leaves a narrow base mesenteric attachment, hence volvulus is a common complication. Peritoneal bands that normally would extend to the right colon from right peritoneal gutter persists as "Ladd's band" , which arise in the right upper quadrant beneath the liver and course across the duodenum in an abortive attempt to fix the malpositioned right colon.[2]

Clinical Presentation

Malrotation may go undetected through out life. Approximately 60% of cases are encountered in the first month of life and over 40% of these within the first week. Sporadic cases occur through out life.[1] The clinical presentation depends on the age at presentation. In the neonatal period, it may present either with acute strangulating obstruction or with recurrent episodes of subacute obstruction. Both are characterised by bilious vomiting from duodenal obstruction or midgut volvulus.[1] In infants and children, a wide spectrum of clinical symptoms occurs, which include bilious or non-bilious vomiting, anorexia or nausea, intermittent apnoea, intermittent abdominal pain, diarrhoea or constipation[1] as was the case with our patient. Other features include malnutrition and failure to thrive due to malabsorption from chronic volvulus. Volvulus is the single most important cause of morbidity and mortality from malrotation.

Diagnosis

Due to the ever present risk of midgut volvulus and gangrene with possible massive bowel loss, the diagnosis of malrotation should be prompt, hence the need for a high index of

suspicion based on above features and utilisation of investigations. Radiological studies are more effective in diagnosis of malrotation. Plain abdominal radiography may show features suggestive of volvulus, which include double bubble appearance and paucity of gas shadow in the rest of the intestine.[1] Contrast radiology plays on important role, especially upper gastrointestinal series and diagnosis is made based on these features;

1. Incomplete duodenal obstruction usually the third portion.
2. Abnormal position of the duodenojejunal junction to the right of the midline.
3. Abnormal position of the proximal jejunal loops to the right of the midline.
4. Presence of cork - screw appearance of duodenum and proximal jejunum. [4]

Barium enema gives information only about position of the caecum, which may occasionally be normally placed even in the presence of volvulus. [4] The usefulness of sonography and computed tomography in diagnosis of malrotation is as yet inconclusive.[5] However it should be noted that patient presenting with acute peritoneal signs may not be suitable for diagnostic contrast studies.

Treatment

The Ladd's procedure has become the gold standard in the treatment of malrotation over the years and only an occasional recurrence of volvulus after this procedure has been reported. The Ladd's procedure involves excision of any bands, dissection and broadening of the mesenteric base, and placement of the entire small bowel on the right side of the abdomen and large bowel on the left side, with caecum in the left upper guardant. Appendicectomy is frequently performed either by inversion or excision technique.[1] Any gangrenous bowel from midgut volvulus should be resected.

Conclusion

Intestinal malrotation is not an uncommon congenital anomaly. Because of the risk of midgut volvulus, which can lead to midgut gangrene and subsequent short bowel syndrome, which is associated with high morbidity and mortality, there is a need for early diagnosis. In our environment, however, diagnosis is frequently made at surgery or after complication has supervened. This may be due to lack of suspicion. Therefore, intestinal malrotation should be considered in children with recurrent vomiting or recurrent abdominal pain. Diagnosis should be confirmed by appropriate investigations and the condition promptly treated to avoid midgut volvulus and attendant morbidity and mortality.

Comments

Dr. Odigie: What is the advantage of inversion appendectomy and is there any contraindication?

Dr. Sabiu (Response): Inversion appendectomy is a sterile form of appendectomy that avoids peritoneal contamination. Contraindications include operating for acute appendicitis or operating on an appendix previously scarred by inflammation. It is not be to be used in the presence of a faecolith unless the faecolith can easily be milked back into the caecum and also in patients who are explored for abdominal pain of unknown origin or if colonic surgery is being performed. [6]

Dr Lawal: How will you manage a patient with midgut volvulus complicated by massive gangrene and what is the role of caecopexy ?

Dr. Sabiu (Response): In such a situation, owing to the risk of short gut syndrome, the volvulus should be untwisted and the abdomen closed. The patient is given 100% oxygen and resuscitation continued. The patient is returned for second look laparotomy in 12 to 24 hours. Perhaps after this time, some of the bowel may recover so that resection and anastomosis or ostomies can be performed which will not result in short gut syndrome. [7]

On the role of caecopexy and other fixation of the mesentery, it is generally accepted that additional fixation does not significantly

increase the number of early survivors, the number of re-operations or recurrence of volvulus or the proportion of children with abdominal complaints later in life. [7]

Dr. Sabo: Is the appendectomy done on this patient incidental appendectomy?

Dr Ameh: It is part of Ladd's procedure. It is necessary because the caecum is now positioned in the left upper quadrant and there my be diagnostic confusion if appendicitis occurs in future.

Dr. Odigie: Are there surgical procedures for the management of short bowel syndrome?

Dr. Sabiu (Response): There are many surgical procedures for management of short bowel syndrome if after the period of intestinal adaptation patient is still dependent on total parental nutrition. These include, slowing intestinal transit by procedures such as reversed small bowel segment, colonic inter position, construction of valves, recirculating loops and electrical pacing. Other modalities include improving the function of existing intestine by plication, tapering, tapering and lengthening and Kimura technique. Lastly, increasing the area of absorption by growth of neomucosa and intestinal transplantation.

Dr Ameh: Intestinal malrotation is not uncommon here. We reported 14 cases seen in this hospital from 1980 to 1989[9] and in all the patients, diagnosis was made only at laparotomy. Because of the devastating consequences of midgut volvulus, there is need for a high index of suspicion among clinicians to make early preoperative diagnosis. The condition should be suspected, particularly in children with recurrent vomiting, recurrent abdominal pains or failure to thrive. Appropriate investigations should then be done to ascertain the diagnosis. It is important to note that following report of the experience here, awareness has increased and was responsible for the preoperative suspicion and diagnosis in this patient.

Dr. Kalayi (Closing): Malrotation is not uncommon in this environment. Emphasis should be on the need for high index of suspicion to make a preoperative diagnosis.

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