Clinical Presentation of Midgut Malrotation in Children Beyond the Neonatal Period

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Midgut malrotation usually presents acutely in the neonatal period. Presentation outside the neonatal period is uncommon and may at times be atypical. Medical records of 17 children over one month of age presenting with midgut malrotation were reviewed. Contrary to the classical acute presentation in the neonates, older children were found to present with widely varying pattern of clinical features that were often non-specific and less dramatic, leading to a significant delay in the diagnosis. These included failure to thrive, chronic non-specific abdominal complaints and malabsorption syndrome. It was observed that while the mortality associated with acute volvulus is well recognised, the morbidity associated with chronic/intermittent volvulus in infants and older children is not adquately appreciated. Because of the unpredictability and high mortality and morbidity associated with acute and chronic volvulus, prompt diagnosis and surgical correction in the form of Ladd's procedure is recommended in every patient with midgut malrotation.

Key words: Midgut malrotation, acute volvulus, chronic volvulus, Ladd's procedure

Midgut malrotation (MM) results from an arrest of rotation in the second stage of normal rotation [1,2]. The process was nicely described by Mall[3] and, Frazer and Robbin[4] based on studies of a large number of embryos. Dott[5] correlated the embryological observations with clinical presentation, and Ladd[6] standardised various operative steps. In MM the duodenum fails to cross the midline, and the caecum fails to descend to the right iliac fossa. Ladd's bands arise from the subhepatic area and course across the accordionated duodenum in an attempt to fix the malpositioned right colon and caecum. The root of the mesentery remains narrow and predisposes to the development of volvulus that in turn results in interruption of the mesenteric venous drainage and arterial inflow leading to intestinal gangrene[2].

MM generally presents in the neonatal period with signs of upper gastrointestinal obstruction, like bilious vomiting, cecause of duodenal obstruction secondary to Ladd's bands. In the neonates, there is a high incidence of volvulus that may present additionally with persistent irritability, abdominal distension and tenderness, bleeding per rectum and shock[2,7]. Differential diagnosis at this age includes duodenal atresia, gastroenteritis and necrotising enterocolitis (NEC).

Beyond the neonatal period, MM may present with widely varying clinical features that often tend to be non-specific and less dramatic. This may at times lead to a delayed and erroneous diagnosis, and inappropriate management. Experience with children presenting beyond the neonatal period is described, along with a literature review.

Patients and Methods

Seventeen children older than one month having midgut malrotation were admitted to the regional paediatric surgical units at the Children's Hospital, Sheffield, UK during 1990 and Chelsea & Westminster Hospital, London, UK during 1992-95. Their medical records were reviewed retrospectively to observe various modes of clinical presentation in relation to age. Their age ranged between 2 months to 15 years, with a mean of 4.63 years. The male to female ratio was 2:1.4. After the diagnosis was made, all patients underwent a Ladd's procedure i.e. division of Ladd's bands and widening of the root of the mesentery.

Results

In the infants, the main clinical features include intermittent vomiting, loose motions, failure to thrive, feeding problems and apnoeic attacks. All of these infants had been initially investigated by physicians to detect medical illnesses and had been treated by several formula changes and therapies including anti-emetic medications. The diagnosis was finally established by upper gastrointestinal contrast studies and ultrasound scan of the superior mesenteric vessels. Two infants developed an acute volvulus. One required resection of a limited segment of the bowel and received total parenteral nutrition for a period of two weeks before tolerating oral feeding.

In older children, intermittent non-specific abdominal pain, non-bilious vomiting and growth failure were the main clinical features. Physical examination of the abdomen was unremarkable in all patients.Ultimately, the diagnosis was made on upper gastrointestinal contrast studies. In a diabetic child, delay in surgical referral occurred because his intermittent symptoms were thought to be due to his diabetes. He developed an acute volvulus, but at laparotomy, the gut was found to be viable. Because of chronic symptoms and the absence of any associated positive physical and laboratory findings, one patient had even been referred to a psychiatrist. In our children MM was an incidental finding during laparotomies for other diseases.

The mean duration of symptoms was 19.96 days (range 2-42 days) in the infants and 8.25 months in children older than one year (range: 2 months-3 years), reflecting a significant delay in surgical referral. There was no mortality in this series. All patients remain symptom free at 6 months to 4.5 years follow up.

Discussion

Typically, MM presents in the neonatal period with sudden onset of bilious vomiting and abdominal distention, secondary to duodenal obstruction from Ladds bands, or development of a volvulus[8,9,10]. Outside the neonatal period, clinical symptoms are often vague and non-specific, rendering the diagnosis difficult. Infants may present with widely varied symptoms like intermittent bilious or non-bilious vomiting, feeding problems, malabsorption patterns associated with diarrhoea, protein-calorie malnutrition and failure to thrive[11,12,13]. As a result, these infants are often referred to paediatricians. Often, the symptoms are attributed to non-surgical conditions like milk intolerance, gastrointestinal allergies and celiac disease[13]. This leads to inappropriate extensive investigations and empirical trials of various therapies like anti-reflux medications, jejunal biopsies, glutenfree diect, lactose-free diet and change of formula. This indicates that the morbidity associated with chronic volvulus is generally not well appreciated. In actual fact, these symptom result from chronic/intermittent volvulus and obstruction resulting in blind-loop syndrome, lymphatic obstruction and protein loss in the bowel[14]. The bowel demonstrates a chronic venous and lymphatic congestion of the mesentery that impairs nutrient absorption and transportation[11,12,14]. Various degrees of chronic intestinal ischaemia may further damage the absorptive mucosal surface. Infants may also present with intermittent apnoea secondary to gastro-oesophageal reflux precipitated by abnormal gastric motility and delayed gastric emptying[15]. Intestinal ischaemia from chronic/recurrent volvulus can also result in gastrointestinal haemorrhage and anaemia[16].

Older children usually present with a longer history of itnermittent or cyclical vomiting, intermittent abdominal pain, bloating, nausea, anorexia, diarrhoea, constipation and chronic malnutrition[14,17,18]. These children with chronic symptoms may become withdrawn and introverted, and face chronic learning and psychomotor disabilities[17]. Many children carry the diagnosis of psychogenic vomiting for months or years before a correct diagnosis of MM is made[10]. The abdominal pain is typically postprandial colicky, transient, vague and not associated with any physical finding[19]. It must be appreciated that vomiting, bile stained or not, and colicky abdominal pain in the absence of abdominal distension and other physical signs do not exclude MM. These patients must be investigated properly to rule out MM before giving them a diagnostic label of abdominal migraine, cyclical vomiting, worm colic and psychiatric illness. Because of an ill-defined and "benign" past history, these children may present acutely because of sudden development of volvulus that may prove disastrous[7]. Rare presentations of MM include obstructive jaundice, peptic ulcer disease due to gastric and duodenal stasis[20], delayed diagnosis of appendicitis due to abnormal caecal position[21], intussusception due to lack of ileo-caecal fixation[22]. A few patients with MM may remain undiagnosed throughout life or present as an incidental finding[18,21].

Associated anomalies and problems are common, with an incidence of 30% - 62%[1,10]. These include gastrointestinal disorders like duodenal atresia, gastro-oesophageal reflux, jejuno-ileal atresias, Hirschsprung's disease and anorectal anomalies. In-utero bowel ischaemia caused by Ladd's bands and volvulus has been implicated in the pathogenesis of the common associated anomalies: duodenal atresia and ileo-jejunal atresias respectively. The other associated anomalies include congential diaphragmatic hernia, gastroschisis, exomphalos and congenital heart defects. MM should be considered in the differential diagnosis when evaluating a child with multiple anomalies who presents with recurrent abdominal pain, feeding problems and failure to thrive[17].

Plain abdominal radiographs often fail to show any abnormality, leading to a delay or failure to pursue the diagnosis. Although favoured in the past, barium enema is less commonly used now-a-days because of high false-positive and false-negative rates[8]. Upper gastrointestinal contrast study is the most helpful investigation, the classic finding being the failure of the duodenum to cross the midline[23]. Recently, the observation of abnormally oriented superior mesenteric vessels on ultrasound scan has been successfully utilised for the diagnosis of MM in centres where services of experience dultrasonologists are available[24].

Patients with MM are always prone to develop an acute volvulus that may lead to extensive bowel gangrene. Sudden onset of volvulus has been documented in previously asymptomatic children of different ages[1,13,20]. The resultant morbidity and mortality can be reduced by virtue of early recognition of the disease process and prompt intervention. The diagnosis is fairly easily made if the classical features are present. However, in older children the diagnosis may often remains dubious because of vague presentation. Awareness of the possibility of MM in children presenting beyond the neonatal period may avoid an unnecessary delay in the diagnosis. Surgical correction must be offered to every patient of MM regardless of age, whether symptomatic or asymptomatic, documented or incidental.

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