Recurrent intestinal volvulus in midgut malrotation causing acute bowel obstruction: A case report

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Abstract

Intestinal malrotation occurs when there is a disruption in the normal embryological development of the bowel. The majority of patients present with clinical features in childhood, though rarely a first presentation can take place in adulthood. Recurrent bowel obstruction in patients with previous abdominal operation for midgut malrotation is mostly due to adhesions but very few reported cases have been due to recurrent volvulus. We present the case of a 22-year-old gentleman who had laparotomy in childhood for small bowel volvulus and then presented with acute bowel obstruction. Preoperative computerised tomography scan showed small bowel obstruction and features in keeping with midgut malrotation. Emergency laparotomy findings confirmed midgut malrotation with absent appendix, abnormal location of caecum, ascending colon and small bowel. In addition, there were small bowel volvulus and a segment of terminal ileal stricture. Limited right hemicolectomy was performed with excellent postoperative recovery. This case is presented to illustrate a rare occurrence and raise an awareness of the possibility of dreadful recurrent volvulus even several years following an initial Ladd’s procedure for midgut malrotation. Therefore, one will need to exercise a high index of suspicion and this becomes very crucial in order to ensure prompt surgical intervention and thereby preventing an attendant bowel ischaemia with its associated high fatality.

Key words: Gut volvulus; Intestinal malrotation; Acute bowel obstruction; Computerised tomography scan; Laparotomy

INTRODUCTION

Intestinal malrotation occurs when there is a disruption in the normal embryological development of the bowel rotation, elongation and fixation. Normal developmental gut rotation takes place around the superior mesenteric artery (SMA) which supplies the midgut. Disturbance of this process will lead to incomplete or non-rotation of the foetal midgut. This condition affects approximately 1 in 500 live births with the vast majority of the affected individuals manifest by the time of their first birthday [1-5]. It has been reported that well over 90% of the affected individuals manifest by the time of their first birthday [1-5].

The diagnosis of midgut malrotation is rarely reported in adults [2,4-9]. A small proportion of the cases go undetected until adulthood when they are incidentally diagnosed in the course of radiological investigations or operative interventions for acute bowel obstruction or other unrelated conditions [3-6]. There is even a lesser

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group of patients presenting later in life with intermittent non-specific acute or chronic symptoms where the diagnosis is particularly difficult to make and the condition can go on for life undetected\(^2,5-9\).

This is a report of a young adult who previously underwent a laparotomy three weeks of age for bowel volvulus and represented later with acute small bowel obstruction due to recurrent volvulus in the setting of midgut malrotation. This unique case is reported to illustrate a rare occurrence of recurrent volvulus following Ladd’s procedure for midgut malrotation. Therefore, a high index of suspicion is required for early diagnosis and prompt surgical intervention in order to prevent the risk of bowel gangrene and its associated high fatality.

**CASE REPORT**

A 22-year-old gentleman presented with three days history of an acute onset central abdominal pain, progressive distension and vomiting. Patient has been experiencing intermittent abdominal pain for weeks and erratic bowel habit with scanty pellet-like stool prior to presentation. He had presented 2 wk earlier and underwent an emergency left inguinal hernia repair which was misdiagnosed as the cause of the intermittent abdominal pain. There was a background history of laparotomy for “twisted” bowel when he was 3-wk-old.

Physical examination at this presentation showed dehydration, distended abdomen with tenderness around the umbilicus. There was no peritonitis and bowel sounds were high pitched and hyperactive. Rectum was empty.

Blood tests were unremarkable with normal parameters for full blood count, urea and electrolytes, liver function tests, arterial blood gases, C-reactive protein and lactate. The abdominal radiograph showed features of a small bowel obstruction. This was subsequently confirmed on the abdominal computerised tomography (CT) scan. The caecum was located to the upper left quadrant with the large bowel on the left of the abdomen and most of the small bowel loops were on the right side (Figure 1A). There was failure of progress of the duodenum to the left side of the spines and aorta (Figure 1B). There was also a reversal of the relationship between the mesenteric artery and vein (Figure 1C). A diagnosis of adhesions causing bowel obstruction in the setting of midgut malrotation was made.

The patient was adequately resuscitated and underwent an emergency laparotomy and limited right hemicolectomy with ileocolic anastomosis. The findings at operation were consistent with midgut malrotation, with small bowel on the right side and pelvis, caecum and ascending colon on the left upper abdomen and the duodenal-jejunal flexure on the right side of the ascending colon. The appendix was absent presumably removed at the previous laparotomy. There were minimal intra-abdominal adhesions. The cause of obstruction was small bowel volvulus with dilated, congested but viable bowel and a segment of chronically thickened and strictured terminal ileum presumably the site of previous ileoileal anastomosis in childhood (Figure 2). The patient made a good recovery postoperatively and was discharged home a week after the operation. Follow up in the clinic showed no recurrence of symptoms up until 6 mo after surgery.

**DISCUSSION**

Midgut malrotation is a rare cause of intestinal obstruction in adult life and only few of such cases have been reported in the literature\(^2,4-11\). Recurrent intestinal obstruction is even rarer in adults who have been previously operated for gut malrotation and few of such cases have been reported. Features of intestinal obstruction in patients who have had previous laparotomy always raise the suspicion of adhesions as the aetiological factor. The other possible causes to consider are either postoperative
midgut volvulus or internal herniation and few of the latter have been reported following laparoscopic appendicectomy, cholecystectomy and gastric banding operations\(^{[12,14]}\). The reason(s) for this rare phenomenon following laparoscopic operations is not well understood. Bikoo et al\(^{[16]}\) in their retrospective review of obstructive symptoms in patients post Ladd’s procedure showed that adhesive small bowel obstruction was more common than the most dreaded recurrent gut volvulus.

Midgut malrotation is rarely considered as an underlying diagnosis in adults and may present in various ways. Our patient had a history of abdominal surgery as a child for volvulus but there was no knowledge of the aetiological factor at the time of this presentation. He presented initially two weeks earlier with features of subacute obstruction and finding of a left inguinal hernia. He had a presumed diagnosis of an obstructed left inguinal hernia and underwent an emergency hernia operation. However, this treatment did not resolve his symptoms hence necessitating a representation with an acute bowel obstruction. The initial diagnosis of acute adhesive bowel obstruction was made on the background history of previous laparotomy he had as a 3-wk-old child. The clinical diagnosis of midgut malrotation in adolescents and adults is difficult because it is rarely considered on clinical grounds. Beside, many of these patients remain asymptomatic and majority of them are only discovered incidentally during investigations or laparotomy. Dietz et al\(^{[9]}\) in a series of 10 adults with intestinal malrotation showed that 4 and 5 of them presented with acute and chronic bowel obstructive symptoms respectively and one patient had an acute abdomen due to appendicitis.

Recurrent volvulus as a cause of bowel obstruction following Ladd’s operation for midgut malrotation is very rare both in children and adult life and very few of such cases have been reported in literature\(^{[15–22]}\) (Table 1). Recurrent symptoms in such cases are usually considered to be due to adhesions and one may be inclined to adopt a non-operative approach. Fu et al\(^{[16]}\) reported only two recurrences in a series of 12 adults treated for symptomatic malrotation with one of them requiring a reoperation and the other managed conservatively. It is believed that the increasing use of CT scan will enable one to make such diagnosis with certainty preoperatively as this has the overall advantage of detecting the abnormal location of the midgut as well as any other intra-abdominal anomalies. The finding of midgut malrotation should make one to suspect a possible diagnosis of intestinal volvulus which may require an early surgical intervention so as to prevent the most dreadful and life threatening bowel ischaemia and infarction.

The standard surgical intervention in patients with obstructive symptoms and gut malrotation is Ladd procedure which was originally described in paediatric population by Ladd\(^{[23]}\). This procedure consists of 4 elements including the division of Ladd’s bands overlaying the duodenum; widening of the narrowed root of the small bowel mesentery by mobilising the duodenum and division of the adhesions around the SMA to prevent further volvulus; counterclockwise detorsioning of the midgut volvulus if present and appendicectomy to prevent future diagnostic dilemma of an abnormally located inflamed appendix\(^{[22]}. Most authors are of the opinion that Ladd’s procedure is an adequate treatment for intestinal malrotation but various modifications of this operation have been reported. The full components of this procedure may not be required in the adult group to deal with the bowel obstruction\(^{[19,20]}\). One of the clear objectives of surgical management of midgut malrotation is to prevent recurrent volvulus and there are various techniques used to prevent such complication. This includes re-establishment of the normal gut anatomy by duodenopexy, cecopexy and suture fixation of the ascending colon to the right abdominal wall, in the retroperitoneal position\(^{[8,9]}\). There are reports of increasing use of laparoscopic approach to Ladd’s operation in the literature\(^{[22,23]}\) with excellent outcome.

It was difficult to ascertain the full details of the procedure(s) performed in our patient in childhood as the operation took place in a different hospital with una-

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**Table 1** Reported cases of recurrent intestinal volvulus following previous Ladd’s procedure for midgut malrotation

| Ref.       | Year | No of cases | Diagnosis          | Management of volvulus |
|------------|------|-------------|---------------------|------------------------|
| Fu et al\(^{[16]}\) | 2007 | 3           | 2-recurrent volvulus | 1-surgery              |
|            |      |             | 1-adhesive bowel obstruction | 1-conservative treatment |
| Mazeh et al\(^{[17]}\) | 2007 | 1           | Recurrent volvulus   | Surgery                |
| Alkan et al\(^{[18]}\) | 2007 | 1           | Recurrent volvulus   | Surgery                |
| Tashjian et al\(^{[19]}\) | 2007 | 3           | 1-recurrent volvulus | All had surgery        |
|            |      |             | 1-adhesive bowel obstruction | 1-closed loop obstruction |
| Panghaal et al\(^{[20]}\) | 2008 | 1           | Recurrent volvulus   | Surgery                |
| El-Gohary et al\(^{[21]}\) | 2010 | 10          | 1-recurrent volvulus | Surgery                |
| Bikoo et al\(^{[16]}\) | 2011 | 9           | 9-adhesive bowel obstruction | Surgery |
|            |      |             | 1-recurrent volvulus | Surgery                |
|            |      |             | 8-adhesive bowel obstruction | Surgery |
| This case  | 2012 | 1           | Recurrent volvulus   | Surgery                |
vailable medical records. Our best guess is that he may have had the standard Ladd's procedure at that age as the appendix and the classical Ladd's bands were absent at laparotomy. We presumed he may have also had a bowel resection for ischaemic bowel resulting from volvulus as evident by strictured distal ileum. There was no evidence that a caecopexy and/or fixation of the ascending colon to the right abdominal wall were performed. This patient had a recurrent small bowel volvulus and chronic stricture of the distal ileum causing acute bowel obstruction. Recurrent small bowel volvulus also may have been encouraged by the minimal adhesion formation following the laparotomy he had in childhood. He then underwent a limited right hemicolectomy and ileocolic anastomosis with an uneventful postoperative recovery.

In conclusion, midgut malrotation is rare in adult population but an important factor contributing to bowel obstruction in that group. The most dreadful and life threatening complication of intestinal malrotation both in children and adults is gut volvulus with possible ischaemic changes and associated high mortality. However, recurrent volvulus resulting from intestinal malrotation is uncommon after treatment with Ladd's procedure and only very few of such cases have been reported in the literature. Majority of recurrent bowel obstructive symptoms are due to adhesions from previous laparotomy. Therefore, one will need to exercise a high index of suspicion and an awareness of the possibility of recurrent volvulus even several years following an initial Ladd's procedure. This is crucial to ensure prompt surgical intervention in order to prevent attendant bowel ischaemia and a high fatality rate.

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