CASE REPORT

Malrotation of the gut harbouring midgut volvulus in an adult with situs inversus totalis: a clinical delay and treatment dilemma

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SUMMARY

A 22-year-old man experiencing infrequent episodes of abdominal pain, distension, non-bilious vomiting and constipation, was diagnosed with malrotation of the gut. He was treated conservatively over the past 10 years. He was referred to our hospital owing to recent aggravation of symptoms. He had no signs of peritonitis. On imaging, malrotation of the gut, with midgut volvulus and situs inversus totalis, was found. Diagnosis was confirmed during laparotomy. A large sac was present to the right of midline. Detorsion of the sac was performed and the sac was opened. The duodenum and caecum were found in the left upper abdomen adherent to the lateral abdominal wall. Adhesiolysis was performed and extrinsic compression at the duodenum relieved. The large bowel was placed on the right side and small bowel was placed on the left. Appendectomy and feeding jejunostomy were performed. Thorough analysis (clinicoradiological) is necessary before considering conservative management in patients known to harbour a congenital anomaly of the gut.

On examination, he was haemodynamically stable and had no signs of peritonitis.

INVESTIGATIONS

Haemogram, serum electrolytes and leucocyte count were in normal range. Chest and abdominal roentgen revealed dextrocardia; there was no free air under the diaphragm (figure 1). CT of the abdomen revealed Situs anomaly (figure 2), distended loops of small intestine, and whorled and twisted mesentery (figure 3). The branches of the superior mesenteric artery (SMA) were hypertrophied and tortuous. Diagnosis of MOG and MV in a patient with SIT was made.

DIFFERENTIAL DIAGNOSIS

Adhesive intestinal obstruction, volvulus of intestine and internal hernia.

BACKGROUND

Malrotation of the gut (MOG) with midgut volvulus (MV) in patients with situs inversus totalis (SIT) is a rare clinical condition seen usually in adults. Surgery performed for asymptomatic MOG in children often entails repeated hospital admissions for symptoms of adhesive intestinal obstruction in the postoperative period. Whether or not to operate on an asymptomatic MOG is a topic of controversial discussion. Conservative management of MOG involves increased responsibility on the part of the treating surgeons, therefore thorough clinicoradiological investigations are necessary before initiation of conservative treatment in patients with MOG. We present a rare case of MOG harbouring MV in an adult with SIT maintained on conservative treatment for the past 10 years, which eventually required surgery for suspected diagnosis on clinicoradiological findings.

CASE PRESENTATION

A 22-year-old man with infrequent episodes of abdominal pain, distension, non-bilious vomiting and constipation, managed conservatively for the past 10 years, was referred to our hospital due to recent aggravated symptoms (abdominal pain, bilious vomiting and distension of the abdomen) that were refractory to conservative treatment.

Figure 1 Chest X-ray posteroanterior view. Arrow A showing dextrocardia with apex of the heart situated on the right side of the body. Arrow B showing fundal gas shadow on the right side below the diaphragm.

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Figure 2 CT scan showing stomach and spleen resting on the right side and liver on the left. Arrow showing dextrocardia with apex of the heart situated to the right of the body.

TREATMENT

During laparotomy, the liver and gall bladder were found in the left hypochondria and the spleen was present in the right hypochondria, confirming SIT (figure 4). There was a large sac containing loops of intestine present to the right of midline. The sigmoid colon was present in the right iliac fossa. The caecum and short ascending colon (short mesentery) were abnormally

Figure 3 CT scan (chest and abdomen): arrow pointing towards whorled superior mesenteric artery.

Figure 4 Situs inversus (liver and gall bladder present in the left upper quadrant) and arrows pointing towards Ladd bands.

positioned in the left upper abdomen. Detorsion of the entire sac was performed four times (720° detorsion) and the sac was opened. Intestines were found twisted, entangled within themselves and viable. The ligament of Treitz was absent, the duodenojejunal junction was found twisted around itself and adhering to coils of jejunum located anterior to the SMA on the left. Tight peritoneal bands (Ladd bands) extended from the caecum to the left lateral abdominal wall across the duodenum and gall bladder, causing extrinsic compression and obstruction of the junction between second and third sections of the duodenum (figure 4). The fourth segment of the duodenum along with duodenojejunal junction and coils of the proximal part of the small intestine were present in a sac, whorled and twisted around itself located on the right of midline. MOG with MV was confirmed. The coils of intestine were untwisted and mesenteric widening was performed by carrying out adhesiolysis (figure 5). The large bowel was placed on the right and small bowel was placed on the left. Appendectomy and feeding jejunostomy were performed. The feeding jejunostomy was performed for interim feeding during the immediate postoperative period; it was carried out in view of the tight extrinsic constriction caused by Ladd bands at the junction of second and third segments of the duodenum, creating a narrow passable small lumen. Appendectomy was performed to avoid subsequent confusion in the diagnosis in case the patient presented with abdominal pain, as the patient had SIT.

OUTCOME AND FOLLOW-UP

The patient was discharged on seventh postoperative day and gained 22 kg over 12 months (68%). He is currently involved in his regular routine and enjoys an improved quality of life.

Figure 5 Arrow A showing release of Ladd bands. Arrow B showing narrow mesentery before adhesiolysis and Arrow C showing widened mesentery after adhesiolysis.

DISCUSSION

SIT is an autosomal recessive or X-linked anomaly occurring in approximately 0.001% live births.¹ People with SIT generally lead a healthy and normal life. SIT may be associated with congenital anomalies.^{1,2} True incidence of MOG in the adult population of SIT is not known. The latter is difficult to estimate owing to its rare occurrence, asymptomatic nature and only a few patients require surgical treatment. MOG in adults with SIT is often detected during autopsy or found incidentally on imaging tests performed for unrelated clinical conditions or during surgical intervention for other pathologies. MOG is an embryological abnormality more frequently diagnosed in infants and children than in adults. In contrast with traditional teaching, which states that nearly half of patients with MOG present during adulthood,³ the current literature suggests that infants most often present with emesis (93%), while abdominal pain was a common presentation in children (67%) and adults (87%). The incidence of acute presentation declines with age, from 37% in infants to 12% in adults.⁴ Most infants present with acute presentation, while children (59%) and adults (32%) present with a chronic pattern over the course of weeks to months to years, with episodic vomiting and abdominal pain suggestive of intermittent MV.⁴ In this case study, the patient had chronic abdominal pain over the past one decade, with intermittent episodes of increased pain severity requiring hospital admission, and was treated conservatively for diagnosed MOG.

Incidentally detected asymptomatic MOG may not universally require surgical intervention in the absence of volvulus or internal hernia. Treatment of incidentally diagnosed asymptomatic MOG is currently a controversial topic of discussion. Murphy and Sparnon⁵ documented the risk of adhesive bowel obstruction after Ladd's procedure as being significantly high in children who had undergone surgery for malrotation. Mehall *et al*⁶ studied the rate of volvulus in patients with MOG and concluded that the rate of volvulus was 2% in patients with atypical symptoms (postprandial vomiting or gastro-oesophageal reflux) and 16% in patients with typical symptoms (acute colicky abdominal pain or bilious vomiting). Surgery performed on patients presenting with atypical symptoms has significantly more postoperative complications than those in patients operated for acute symptoms.⁶

CT of the abdomen with or without oral contrast is useful in guiding treatment of bowel obstruction. It is also useful in detecting the location of gut obstruction, mesenteric twist and free air in the peritoneum, and the associated intra-abdominal abnormality. Mesentery twists, wrapped around the SMA pedicle creating a distinctive whirlpool appearance on CT scan, as in our patient, are the characteristic findings of MV.^{7,8} In the absence of a definitive treatment algorithm, management of MOG is subjective. Definitive indication for surgery is volvulus of the intestine or internal hernia. On exploration, our patient had a stage II anomaly of intestinal rotation (malrotation) and stage III anomaly of intestinal rotation (unattached duodenum, mobile caecum, absent ligament of Treitz and small bowel mesentery). His recovery was uneventful postsurgery and he had gained 22 kg (68%) at 12-month follow-up.

Learning points

- ▶ Malrotation of the gut (MOG) may present with non-specific and atypical symptoms leading to clinical delay and treatment dilemma.
- ▶ Conservative management of MOG is an added responsibility on the part of the surgeon; it requires patient education and early return to hospital in episodes of acute abdomen, for reassessment and evaluation.
- ▶ MOG in patients with SIT is known to be associated with other congenital anomalies (eg, anomaly of intestinal rotation). Conservative management in such patients should be pursued after thorough clinicoradiological evaluation (imaging studies to exclude diagnosis of volvulus of intestine or internal hernia) to avoid strangulation of bowel and subsequent catastrophe and fatality from short bowel syndrome or intestinal failure.

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Contributors SC and MMP conceived the design. SC was the first operating surgeon, MMP and JA were first and second assistant surgeons respectively.

MU took the operating steps photographs, JA and MU together collected images. Demography of the patient and clinical details were collected by JA and MU and analysed by SC and MMP. Manuscript was prepared by SC, MMP, JA and MU. Final approval of the case report is provided by SC, MMP, JA and MU.

Competing interests None declared.

Patient consent Obtained.

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