Unusual association of diseases/symptoms

Unusual presentation of midgut malrotation with incidental nutcracker syndrome in adulthood: case report and literature review

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Summary

Malrotation of the midgut is generally regarded as paediatric pathology with the majority of patients presenting in childhood. The diagnosis is rare in adults, which sometimes results in delayed diagnosis and treatment. We present the case of a 28-year-old woman who presented with vomiting and mild acute pain in the abdomen. CT scan showed abnormal location of the midgut and abnormal relation of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) and duodenal jejunal flexure, confirming midgut malrotation. Peroperative findings showed the duodenum, small bowel loops located on the right side of the abdomen. The caecum and appendix were located at the midline in the epigastrium with the presence of Ladd's bands. SMA right and anterior to SMV whirled around SMA. Incidentally, the intraoperatively dilated left renal vein was compressed between the SMA and the aorta, which was confirmed retrospectively on CT scan with no symptoms related to the condition.

BACKGROUND

It has been reported that the incidence of malrotation in adults is ~0.2%.^{1 2} Initial presentation of symptomatic midgut malrotation is rare in adults. However, a significant number of cases remain quiescent during childhood. Incidental diagnosis may then occur in adulthood, when imaging investigations are carried out for other symptoms or during surgery for unrelated pathology. One can now confirm one's suspicions with CT and ultrasound in terms of determining whether the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) positions are normal or reversed.³

We do not find any case of malrotation incidentally presented with nutcracker syndrome in the literature, so it is a very unusual presentation.

CASE PRESENTATION

A 28-year-old woman was admitted to the Department of Gastroenterology with a history of vomiting and mild pain in the abdomen since the last 3 months. She was passing flatus and motions regularly. There were no associated symptoms. The patient had not expressed similar complaints. She had no medical or surgical history.

On examination, the patient was afebrile and thermodynamically stable. The abdomen was soft, scaphoid, with mild tenderness in the central, epigastric region. There was no evidence of peritonitis or obstruction.

Routine admission blood tests including serum electrolytes, arterial blood gas analysis, liver function test and clotting profile were normal. However, a full blood count demonstrated a haemoglobin level of 13.5 mg/dl with a slightly raised total cell count of 14.9×10^9 and a neutrophil count of 9.7×10^9 .

A chest x-ray did not reveal air under the diaphragm; abdominal standing x-ray did not show any dilated bowel loops in the centre of the abdomen and appear gas filled. The diagnosis remained clueless until an emergency CT scan was obtained, which demonstrated the feature of malrotation. The duodenum was malpositioned below and to the right of the ascending colon and hepatic flexure. The caecum was located at the midline in the epigastrium. There were also dilated small intestinal loops on the right side of the abdomen and an inverse relationship of SMA to SMV with a whirlpool sign (figures 1–5).

The patient was resuscitated with fluid and was prepared for the laparotomy. The findings at operation

Figure 1 CT scan image shows abnormal position of duodenal jejunal flexure on right side.

Figure 2 CT scan showing inverse relationship of superior mesenteric artery to superior mesenteric vein and jejuna loops on right side.

included dilated small bowel loops in the right side of the upper abdomen, and the caecum was in the epigastrium and the left side of the upper abdomen. There was a fibrous band over the distal part of the duodenum, on the right side of the duodenum confirming midgut malrotation.

INVESTIGATIONS

The patient also underwent upper gastrointestinal scopy (UGI), showing features of a dilated first and second part of the duodenum; further information could not be obtained as the scope could not pass beyond it.

DIFFERENTIAL DIAGNOSIS

CT scans of patients with adhesive small bowel obstruction caused by either surgically confirmed adhesive bands or matted adhesions were analysed, showing simple obstruction patterns (single abrupt transition zone, beak sign, 'fat notch' sign), patterns of closed-loop obstruction (two adjacent beaks, C-shaped bowel and radial distribution of mesenteric vessels), the location of the obstruction in the abdominal cavity, and the presence of a whirl sign

Figure 3 CT scan image shows compression of left renal vein between aorta and superior mesenteric artery.

Figure 4 Pre op image shows high caecum and appendix located on the left side of the abdomen.

and a 'small-bowel feces' sign looks similar to malrotation of the gut. So, the patient's relevant history of past operations has value in some cases.^{4 5}

TREATMENT

The surgical management of intestinal malrotation was first described by William Ladd in 1936 and this remains the mainstay of treatment. The classic treatment for incomplete intestinal rotation is the Ladd procedure, which entails counter-clockwise detorsion of the midgut volvulus (if present), division of the abnormal coloduodenal Ladd bands tethering the midgut and causing extrinsic compression, widening of the mesenteric base to prevent further volvulus and removal of the malpositioned appendix.⁶ These principles have remained the same since Ladd's address to the New Hampshire Medical Society in 1936. Generally, the original Ladd procedure was described for the paediatric population group and the full components of this procedure may not be offered in the adult group.⁷ Most authors are of the opinion that Ladd's procedure is an adequate treatment for intestinal malrotation. Fu *et al*⁸ reported a complete resolution of symptoms in nine and near-complete resolution in two of 11 patients. Dietz *et al*⁹ also reported a complete symptomatic resolution in 8 of their 10 patients treated surgically.

Figure 5 Pre op image shows Ladd's band encircling duodenum.

Variations of the technique used to manage intestinal malrotation have been introduced to prevent recurrent volvulus. We offered a modified procedure to our patient by performing a division of Ladd's bands and an appendicectomy. There was no volvulus and we did not feel that the duodenum needed to be mobilised and straightened in this case.

Generally, symptomatic patients with malrotation should be treated surgically. Ambulation of the duodenum, location of the small intestine and colon as well as anatomical position of mesenteric vessels should be intensively observed in order to exclude midgut malrotation.¹⁰ Moreover, Spigland *et al*¹¹ recommended that all patients with malrotation are candidates for laparotomy, even if they are asymptomatic, because the complications associated with intestinal malrotation are based on anatomical reasons that do not alter with age; thus the potential to develop sudden onset of acute midgut volvulus in an asymptomatic patient, at any age, exists.

Recently, laparoscopic techniques for treating malrotation in both infants and adults have been described. The laparoscopic experience with adults has consisted primarily of isolated case reports, till Mazziotti *et al*¹² recently reported a series of malrotation patients managed successfully with laparoscopic intervention. Moreover, Matzke *et al*¹³ published their comparative results for open and laparoscopic Ladd procedures, concluding that the laparoscopic Ladd procedure is feasible, safe and as effective as the standard open Ladd procedure for the treatment of adults who have intestinal malrotation without midgut volvulus. Similar conclusions are presented by Seymour and Andersen,¹⁴ who also noted that long-term follow-up will be required to determine whether patients are at risk for either new symptoms or recurrent malrotation-related problems such as midgut volvulus.

OUTCOME AND FOLLOW-UP

Our patient has been completely symptom free during 12 months of follow-up.

DISCUSSION

Intestinal malrotation is a rare condition and the diagnosis of malrotation after childhood is difficult and usually not readily considered as the cause of intra-abdominal symptoms. It usually presents with non-specific complaints and this often leads to diagnostic and treatment delay with possible bowel ischaemia and necrosis. Evidence of this portends poor prognosis and death. Therefore, a high index of suspicion needs to be maintained and prompt surgical intervention must be considered in order to prevent an abdominal catastrophe and death.

The diagnosis of malrotation is easily made in the neonatal period, but is often delayed in older patients. Unusual forms of malrotation were more frequent in patients undergoing surgery beyond the neonatal period. In this group there was evidence of chronic venous and lymphatic obstruction with one case of superior mesenteric vein thrombosis and two cases of intestinal gangrene. A Ladd's procedure was performed in all cases and the most frequent postoperative complication was adhesive intestinal obstruction. There were no deaths. Awareness of the unusual presentation in patients who present beyond the neonatal period may help reduce delays in diagnosis and surgical

treatment. We believe that laparotomy is indicated in all patients with malrotation, even if they are asymptomatic. Midgut malrotation is broadly considered a deviation from the normal 270° counter-clockwise rotation of the gut during embryonic development during week 4 of fetal development.⁹ The failure of the normal physiological rotation of the midgut leads to various degrees of anomaly including the entire small bowel remaining on the right side of the abdomen, the caecum, appendix and colon on the left and an absent ligament of Treitz. In addition, the small bowel mesentery may develop a narrow vertical attachment and the peritoneal fibrous bands fixing the duodenum and caecum to the abdominal wall may persist. These congenital bands extend from the right lateral abdominal wall, across the duodenum and attach to the undescended caecum and are known as Ladd's bands.^{2 7 8} Ladd's bands compress the duodenum and can potentially cause duodenal obstruction. The malrotation of the gut and abnormal location of the caecum produce a narrow superior mesenteric vascular pedicle, as opposed to the normally broad-based small bowel mesentery.

The clinical diagnosis in adolescents and adults is difficult because it is rarely considered on clinical grounds. Many patients remain asymptomatic and the diagnosis is discovered incidentally during investigations or laparotomy for other unrelated problems in adult life. Two distinct patterns of adult presentations have been reported in the literature⁹: acute and chronic. Chronic presentation is more common in adults. This is characterised by intermittent crampy abdominal pain, bloating, nausea and vomiting over several months or years. The symptoms may be highly non-specific. However, the range of clinical presentations underlines the need for a high index of suspicion of midgut malrotation, when investigating the cause of intermittent and varying abdominal symptomatology in a healthy young adult.^{8 9} Dietz *et al*⁹ studied a series of 10 adults with bowel obstruction caused by intestinal malrotation. They reported that five adults presented with chronic features and that the duration of symptoms extended to 30 years. Fu *et al*⁸ reported that 6 of 12 patients in their series presented with chronic intermittent abdominal symptoms. Diagnostic delays are common in this group of patients because of the non-specific nature of the presentations. The pathophysiology of these chronic symptoms may relate to the compression effect of Ladd's bands running from the caecum and ascending colon to the right abdominal wall.^{9 15}

The other group of symptomatic adults typically present with symptoms of acute bowel obstruction and these patients may or may not report a history of abdominal symptoms, as with our patient. These patients may, on occasion, have symptoms and signs of an impending abdominal catastrophe. Moldrem *et al*¹⁶ reported that 48.5% of their 33 patients presented with an acute abdomen. Acute presentation may be due to volvulus of the midgut or ileocaecum, reported as the most common cause of bowel obstruction in adults with gut malrotation. Other causes of acute presentation may be related to internal herniation caused by Ladd's bands.

We can expect an increase in the incidental diagnosis of gut malrotation with increasing and widespread use of radiological investigations. Diagnostic features of midgut malrotation can be identified using plain abdominal

radiography, ultrasound scan (USS), CT scan, MRI scan and mesenteric arteriography.^{16 17} Conventional plain radiography is neither sensitive nor specific in the diagnosis of gut malrotation, although right-sided jejunal markings and the absence of a stool-filled colon in the right lower quadrant may be suggestive, leading to further investigation. Abdominal colour Doppler USS may reveal malposition of the SMA, raising the suspicion of gut malrotation with or without the abnormal location of the hollow viscus.^{16–18} Characteristic USS findings of midgut volvulus were first described by Pacros *et al*¹⁹ and include duodenal dilatation with distal tapering and fixed midline bowel and mesentery twisted around the SMA axis. These features classically present as the ‘whirlpool’ sign.

CT scan with or without UGI contrast study is increasingly used preferentially as it is now considered the investigation of choice, providing a diagnostic accuracy of 80%.^{9 16} CT and MRI scans may show the SMV to be in an anomalous position: posterior and to the left of the SMA. In addition, they may show the abnormal anatomical arrangements of the midgut with the duodenum not crossing the spine. Deviation from the normal positional relationship of SMV and SMA was originally described by Nichols and Li²⁰ as a useful indicator of the diagnosis of midgut malrotation. However, abnormal orientation of the SMA–SMV relationship is not entirely diagnostic of malrotation; it can also be seen in some patients without the pathology and a proportion of patients with malrotation may have a normal SMA–SMV relationship.¹⁷ The CT appearance of midgut volvulus is diagnostic of malrotation. The shortened mesentery allows the small bowel and mesentery to twist and wrap around the narrowed SMA pedicle to create a distinctive ‘whirlpool’ appearance on CT scan. This pattern was first described by Fisher in a patient with midgut volvulus. It can be detected with both abdominal USS and CT scan. CT scan findings of gut malrotation and small bowel obstruction without volvulus may show internal herniation secondary to Ladd’s

bands. Demonstrating a retromesenteric duodenum is the reference standard of imaging in the work-up of malrotation, not any other previously published criteria. The US imaging is the most acceptable imaging method for malrotation work-up, in the spirit of the ALARA principle and the ‘Image Gently’ campaign.²¹

Competing interests None.

Patient consent Obtained.

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Learning points

- Intestinal malrotation is a rare condition but should be considered as an important cause of bowel obstruction in adults.
- Radiological modalities are important for the confirmation of the diagnosis in clinically uncertain cases.
- Clinically definitive diagnosis is very difficult as presenting with variable non-specific features and specially in adulthood because it overlooked diagnosis in this age group.