32-Year-Old Man with Abdominal Pain, General Malaise and an Inability to Eat: A Case of Intestinal Malrotation

Katie O' Sullivan
Sixth Year Medicine, TCD

Clinical Points:

- Intestinal malrotation can lead to volvulus (a twist of the gut around its mesenteric axis which compromises blood supply) in adults.
- Symptoms are bilious vomiting and acute bowel obstruction on a background of recurrent, crampy abdominal pain and episodes of nausea and vomiting.
- A high index of suspicion is crucial when a patient repeatedly presents with abdominal pain and an inability to eat as a malrotation of the intestine is a rare but serious condition.
- Management of the condition is surgical and traditionally comprises a Ladd's procedure with variations of the procedure depending on the intestinal anatomy involved.

PRESENTATION OF CASE

Mr. X, a 32-year-old Egyptian man, presented to the emergency department of a country hospital complaining of an eight day history of persistent abdominal pain, general malaise and inability to eat. There was epigastric pain that had an abrupt onset, was sharp in nature and radiated to the lower abdomen. It was associated with nausea, vomiting, constipation and was exacerbated by movement. He had no symptoms of urinary tract infection. Upon admission he experienced one episode of bile stained vomiting. His history revealed two previous presentations to the emergency department complaining of similar abdominal pain. On the first occasion eight days previously no diagnosis had been made. On the second occasion, three days previously, Mr. X was diagnosed with both grade one gastritis and duodenitis and was discharged on a course of triple therapy of full dose proton pump inhibitor omeprazole (20mg OD), amoxicillin (1g BD) and clarithromycin (250mg BD) for seven days. This failed to alleviate symptoms.

He had a positive family history of hypertension and diabetes. He was a non-smoker and non-drinker. On examination the patient was in acute pain, afebrile with a temperature of 37.6ºC. He was mildly tachycardic and hypertensive with a blood pressure of 152/112 mmHg. Other vital signs were normal. His abdomen was non-distended however there was guarding and tenderness in the epigastric and left upper quadrant region. Bowel sounds were present and hernial orifices were free.

INVESTIGATIONS AND DIAGNOSIS

On presentation three days previous to the current presentation, blood results had revealed haemoglobin, haematocrit, biochemistry, clotting and coeliac screens and thyroid function to be normal. C-reactive protein was at the upper limit of normal and alanine aminotransferase was slightly raised. Urine microscopy was unremarkable. Radiological investigations of the abdomen were carried out. A plain film of the abdomen was normal, and abdominal ultrasound confirmed the absence of gallstones and normal liver, biliary tree, spleen and kidneys. Gastric and duodenal biopsies taken during an oesophagogastroduodenoscopy (OGD) revealed grade one gastritis over the fundus, body and pylorus of stomach and grade one duodenitis. Campylobacter-like organism (Clo) testing was carried out and confirmed *Helicobacter pylori*. The patient at this time was commenced and discharged on triple therapy.

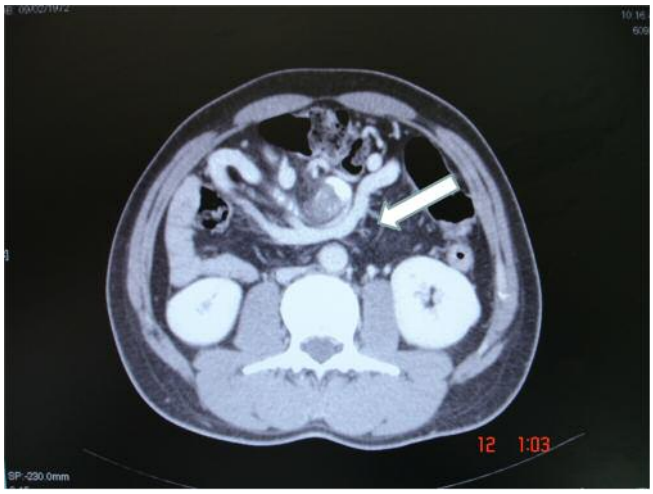

Fig. 1. A CT image of the abdomen of Mr X.
In this CT an arrow points to a whorled appearance of the small mesenteric artery and small mesenteric vein in the central upper abdomen consistent with volvulus. Also evidence of venous congestion of the mesentery exists.

On current presentation, follow through contrast studies of the small bowel revealed delayed passage of barium from the stomach, a midline duodenojejunal flexure, predominate localisation of the small bowel to the right upper abdomen and a high caecal pole. A diagnosis of malrotation was made. In contrast studies malrotation can appear as corkscrew tapering of the duodenum or jejunum. The 'corkscrew sign' is due to intestinal obstruction, but this is not always a reliable sign. Malrotation may also be diagnosed by computed tomography (CT). Abdominal CT was carried out and revealed a whorled appearance in the central abdomen extending below the origin of the superior mesenteric artery.

This so called “whirlpool sign’ describes the cross sectional appearances of the twisted mesenteric vessels (2, 3) and is pathognomic for the condition(see Fig. 1.). It is a well recognised clinical sign in neonates and can be seen in adults. This provides an immediate diagnosis of malrotation and allowing prompt action. Absence of a dilated bowel suggested that the volvulus was likely to be intermittent.

Diagnosis of intestinal malrotation on clinical grounds is difficult to make, as illustrated by the case of Mr X. Patients often have numerous medical consultations before a diagnosis is made (1). Therefore, a high index of suspicion is crucial when a patient repeatedly presents with the symptoms described in the case presentation. Another challenge that often presents is the presence of co-existing gastrointestinal complaints, also illustrated by the case of Mr X. When the presence of gastritis was confirmed on OGD, a seemingly plausible reason for his symptoms of abdominal pain was found. Malrotation can often mimic other gastro-intestinal diseases.

MANAGEMENT

Mr X was managed conservatively with analgesia for five days. A laparotomy with division of fibrous bands of tissue, Ladd’s bands, and a caecopexy were then performed. Operative findings confirmed a malrotation and revealed a narrow small mesentery, a wide band compressing the 2nd and 3rd parts of the duodenum and the presence of Meckel’s diverticulum (a persistence of the vitelline duct that occurs in 2-4% of people). The remaining abdominal contents were normal.

Management of malrotation relies on speed of diagnosis and prompt surgical correction. In an acute situation placement of a nasogastric tube, IV access and administration of parenteral antibiotics should be accomplished without delay (4). The traditional surgical procedure, the Ladd procedure consists of: i) envisceration of the bowel and inspection of the mesenteric root, ii) counter clockwise derotation of the volvulus, iii) lysis of Ladd’s bands with straightening of the duodenum, iv) an appendecectomy and v) placement of the cecum in the left lower quadrant. However, as illustrated by our case, there are acceptable variations of this procedure depending on the anatomical geography involved. There have been a number of small studies comparing the use of open and laparoscopic procedures, however, because of the absence of larger studies, no clear recommendations have been made regarding the best method of surgical treatment (5,6).

DISCUSSION

Intestinal malrotation is a congenital abnormality of the gastrointestinal tract that predisposes to duodenal obstruction and midgut volvulus and can lead to ischaemic necrosis of the small bowel if undiagnosed (7). It is a very rare presentation in adulthood (7,8) as 60% of cases are diagnosed within the first month of life, 20% between one month and a year and a further 20% are usually diagnosed before the end of childhood. There is a wide variation in reported incidence from 1:500 to 1:6,000 (9) with a male predominance of 2:1 (10).

During the foetal growth a variety of important time points exist for the development of the midgut. At the beginning of week six, rapid elongation of the midgut begins and liver growth reduces the abdominal cavity pushing the intestine into a cavity in the umbilical cord, a process called physiological umbilical herniation. As the loop continues to grow in length, it rotates around an axis formed by the superior mesenteric artery root. When viewed front-on, this happens in a counter clockwise direction and turns a total of 270º before then returning to the abdominal cavity (1). If the rotation is less than 270º, the mesentery may have a narrow base and allow the bowel to rotate on the superior mesenteric axis, creating a volvulus and compromising blood supply. At week ten, intestinal loops begin to return to the abdominal cavity. The factors involved are not entirely known, but abdominal expansion and a reduced rate of liver growth are thought to play a role. The proximal jejunum enters first and goes to the left of the abdominal cavity. This is followed by the remaining loops which settle to the right. The caecal bud is the last part to re-enter. At first it lies in the right upper quadrant but then descends into the right iliac fossa and therefore places the ascending colon and hepatic flexure on the right hand side of the abdominal cavity (11,12). A fusion process then adheres the gut to the posterior abdominal wall resulting in fixation of the colon in the retroperitoneal space. The fascial attachments involved in holding the right and left colon in place are called the fascial fusion planes of Toldt, while the ligament of Treitz fuses the duodenum to the retroperitoneum. It is these broad based attachments which usually limit the mobility of the gut and prevent volvulus and kinking of the vascular supply to the gut. Abdominal rotation and attachment are completed by end of the first trimester (13). If a disruption occurs in this rotation six types of malrotation are possible (14), the commonest involving failure of the caecum to move into the right lower quadrant (15). In addition, Ladd’s bands may cross and obstruct the duodenum (7).

All types of malrotation can lead to the development of a volvulus, a twist of the gut around its mesenteric axis. This occurs due to shortness of the mesenteric root (line of mesenteric fixation to the posterior wall of the abdominal cavity) and the narrowness of the resulting suspensory pedicle of the gut (3,10,16). It is the most feared complication of malrotation as it has a mortality rate of 18-25% if it remains undiagnosed (7,17,18,19).

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