CASE REPORT

Superior mesenteric artery syndrome as a cause for recurrent abdominal pain and vomiting in a 9-year-old girl

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SUMMARY

A 9-year-old girl was admitted under our general surgical team with 2 days of diffuse abdominal pain and vomiting. This was one of multiple admissions for similar symptoms over the past 5 years. She was feverish on admission but haemodynamically stable. On examination, she had a diffusely tender and hypersensitive abdomen, with no guarding or peritonism, and no palpable masses. Of note, the patient was very thin, with almost no body fat. Blood tests were otherwise normal, with a normal abdominal X-ray and abdominal ultrasound. She had undergone three previous abdominal ultrasounds over the past 5 years for similar symptoms, all of which were normal. Following this, CT revealed a diagnosis of superior mesenteric artery syndrome. The patient was transferred to our regional children's hospital for analgesia, nasogastric decompression and nutritional supplementation. She made a swift improvement with plans for ongoing follow-up by the paediatric team.

BACKGROUND

Superior mesenteric artery (SMA) syndrome is extremely rare and the consequences of delayed diagnosis can be severe. Unfortunately it is a condition that many clinicians are unfamiliar with. It is also easy to misdiagnose as a functional disorder, with the result that patients are often misdiagnosed and mismanaged. This case report is also valuable because it highlights learning points pertinent to all clinicians: taking all patient symptoms seriously, not attributing symptoms to inorganic or functional disorders except as a last resort, not casually accepting unsatisfactory diagnoses and not taking radiology reports at face value. It is hoped that this case report will be educational and help clinicians identify rare and unusual pathology at the earliest.

CASE PRESENTATION

A 9-year-old girl was admitted under our general surgical team with diffuse abdominal pain and vomiting of 2-day duration. On admission, the patient was feverish, although haemodynamically stable. General examination revealed a thin girl with very little body fat. She was lying still, inconsolable and hyperventilating. On either side of her sat a very concerned and desperate mother and grandmother. Palpation of her abdomen revealed generalised tenderness and hypersensitivity, but no guarding or peritonism. No abnormal masses were palpable. Her blood tests were otherwise normal.

Over the previous 3 months the patient had been in hospital three times with the same symptoms. It was also noted from her records that since 2009 she had undergone three normal ultrasound scans for similar abdominal pains. An ultrasound on this admission was again completely normal. We asked our paediatric colleagues to review the patient. After a thorough history and examination, the paediatric team concluded that there was no obvious cause for her symptoms, and that there was probably a functional element. They optimised the patient's analgesia and additionally prescribed oral morphine, as required. However, the pain remained uncontrolled, the patient continued to be tearful, and the vomiting worsened. With no obvious cause for her pain, and with increasing desperation and pressure from the child's parents for a satisfactory answer, a CT of the abdomen/pelvis was organised. The CT was initially reported as normal. However, on second review of the images with an experienced senior radiologist, it was noted that there was gastric and duodenal dilation. The angle of the SMA in relation to the abdominal aorta was narrower compared with normal, and distal to the SMA there was collapsed duodenum/jejunum. With a provisional diagnosis of SMA syndrome, the patient was discussed with the paediatric surgery and paediatric gastroenterology teams at our regional children's hospital. Immediate transfer was made under the paediatric gastroenterology team. They instituted nasogastric decompression and nutritional/caloric supplementation, with analgesia optimisation. The obstructive symptoms resolved with the nutritional support and the patient is making a good recovery, with plans for regular follow-up by the paediatric team.

INVESTIGATIONS

Blood tests: white cell count $10.6 \times 10^9/L$, haemoglobin 13.9 g/L, mean corpuscular volume 86.8 fL, platelet count $269 \times 10^9/L$, C reactive protein 13 mg/L, Na 136 umol/L, K 3.8 umol/L, Ur 7.8 mmol/L, creatinine 31 umol/L, estimated-glomerular filtration rate >90 .

Abdominal X-ray (AXR): normal.

Four ultrasounds over 5 years: normal.

CT abdomen/pelvis: compression of third part of duodenum between SMA and abdominal aorta, with proximal duodenal and gastric dilation.

DIFFERENTIAL DIAGNOSIS

► Acute appendicitis

CrossMark

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- ▶ Intussusception
- ▶ Peptic ulcer disease
- ▶ Gastroenteritis
- ▶ Inflammatory bowel disease
- ▶ Mesenteric adenitis
- ▶ Meckel's diverticulitis
- ▶ Urinary tract infection
- ▶ Constipation
- ▶ Irritable bowel syndrome
- ▶ Functional abdominal pain
- ▶ Abdominal migraine
- ▶ Eating disorders

TREATMENT

Medical management: The main aims are rehydration, relief of the obstruction, helping the patient regain weight and restoration of mesenteric fat in order to increase the SMA-aortic angle. This can be achieved through: intravenous fluid and electrolyte replacement, nasogastric/jejunal decompression, enteral feeding past the ligament of Treitz, total parenteral nutrition, and caloric and nutritional supplementation. Small meals with the patient using a knee-chest position can be used to help relieve the obstruction by widening the aortomesenteric angle. Prokinetic drugs such as metoclopramide and cisapride may also be helpful.

Surgical management: Gastrojejunostomy, duodenojejunostomy, division of the ligament of Treitz and duodenal mobilisation. Laparoscopic surgery is increasingly being used to treat SMA syndrome.

OUTCOME AND FOLLOW-UP

The patient was transferred to the regional children's hospital under the paediatric gastroenterology team for analgesia, nasogastric decompression and nutritional/caloric supplementation. She made a swift improvement and her obstructive symptoms resolved. She is to be closely followed up by the paediatric team.

DISCUSSION

SMA syndrome is extremely rare. It was first described in 1861 by Von Rokitsansky, and there have only been around 400 cases reported in the English literature since then.¹ It is caused by compression of the third part of the duodenum between the SMA and the abdominal aorta. Diagnosis is based on three criteria: duodenal dilation, a reduced aortomesenteric angle (typically less than 25°) and compression of the third part of the duodenum by the SMA.² Missing the diagnosis can result in dehydration, electrolyte abnormalities, malnutrition and even death.³

Patients may present acutely or chronically.⁴ Patients typically present with abdominal pain and distension, tenderness and vomiting. Other symptoms include abdominal hypersensitivity, weight loss, diarrhoea, reflux, feeding difficulties and early satiety. The symptoms are typically aggravated by eating, and classically relieved with postural changes, with the knee-chest, prone or left side down positions increasing the angle between the SMA and aorta. Chronic cases can present recurrently and insidiously over many years with abdominal pain, vomiting and early satiety.

There are many possible causes for a reduction in the SMA-aortic angle and resultant SMA syndrome.^{1 2 4} Anatomical causes include an exaggerated lumbar lordosis, low origin of the SMA and high fixation of the duodenojejunal flexure. Diseases that can cause rapid loss of retroperitoneal and mesenteric fat,

Figure 1 CT axial section demonstrating compression of the third part of the duodenum between the SMA and abdominal aorta, with proximal duodenal and gastric dilation (SMA, superior mesenteric artery; SMV, superior mesenteric vein).

and a decrease in the aortomesenteric angle, include: severe burns, malignancy, anorexia nervosa, malabsorption and hyper-metabolic states. The syndrome can also develop after trauma that forcibly hyperextends the SMA across the duodenum. Other causes include prolonged supine bed rest, spinal cord injury and spinal surgery.

The diagnosis is difficult, particularly because of its often insidious and non-specific presentation. Blood tests may show electrolyte disturbance. Plain AXR may show marked gastric dilation. Barium upper gastrointestinal (GI) series may show compression of the third part of the duodenum with proximal dilation, distal collapse and delay in overall transit time. Upper GI endoscopy may reveal narrowing of the third part of the duodenum. CT, the gold standard, demonstrates the close proximity of the SMA and the aorta, and the calibre change of the duodenum (figures 1 and 2).

Medical therapy and conservative measures should be attempted prior to surgery.¹ The main aims are rehydration, relieving the obstruction, helping the patient gain weight and

Figure 2 CT sagittal section showing compression of the third part of the duodenum between the superior mesenteric artery (SMA) and abdominal aorta, with proximal duodenal and gastric dilation.

restoring mesenteric fat in order to increase the SMA-aortic angle. The conservative measures include: nasogastric decompression, enteral feeding past the ligament of Treitz, total parenteral nutrition, intravenous fluid therapy and electrolyte replacement, and caloric and nutritional supplementation. Small meals with the patient using a knee-chest position can be used to help relieve the obstruction by widening the aortomesenteric angle.⁴ Prokinetic drugs such as metoclopramide and cisapride may also be helpful.¹ Surgery is indicated when conservative measures fail and if there is a prolonged history of weight loss. Surgical treatments include gastrojejunostomy, duodenojejunostomy and division of the ligament of Treitz with duodenal mobilisation.²⁻⁴ There is increasing interest in laparoscopic approaches to treating SMA syndrome, for example, Kim *et al*⁵ reported two cases of SMA syndrome treated with laparoscopic duodenojejunostomy. With such an approach the operative time was reduced, and the recovery was uneventful and rapid.

Our case is important because it highlights some very important learning points for all clinicians. The first point to note is that clinicians should recognise the red flags that suggest rare/unusual pathology is being missed: recurrent admissions for the same non-specific and insidious symptoms, symptoms that are difficult to account for, and increasing patient and family frustration/desperation. The second point is that one should not be ascribing diagnoses such as 'in-organic' or 'functional' abdominal pain without first thoroughly investigating the patient. Indeed, one can argue that such diagnoses should be given only as a last resort. Leading on from this, the third point is that clinicians should not easily accept unsatisfactory diagnoses. One should endeavour to find the root cause of patients' symptoms, and not give up until this is achieved. In our case we did not blindly accept the diagnosis of functional abdominal pain, and we asked for a second opinion on the CT images that were initially reported as normal. It was persistence and continued enthusiasm that ultimately revealed the hidden diagnosis. Of course, many patients with recurrent and non-specific symptoms may not have genuine organic pathology, but it is still good

practice to take all patient's symptoms seriously and investigate them appropriately.

Learning points

- ▶ Recurrent presentations for the same symptoms (particularly non-specific symptoms) should alert clinicians to the possibility that genuine underlying pathology has been missed.
- ▶ One should not attribute symptoms to inorganic or functional disorders without being confident that the patient has been thoroughly investigated beforehand.
- ▶ Do not accept radiology reports at face value, as a second opinion from another more experienced radiologist may reveal a hidden diagnosis.

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Competing interests None.

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