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Case Report

Intermittent superior mesenteric artery syndrome in a patient with multiple sclerosis

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ABSTRACT

A 42-year-old man with multiple sclerosis presented with recurrent vomiting, in the context of recent weight loss. Computed tomography scan of the abdomen and pelvis revealed duodenal compression by the superior mesenteric artery (SMA), consistent with intermittent SMA syndrome. Subsequent gastroscopy and barium meal follow-through showed resolution of the obstruction. SMA syndrome is rare and has not previously been reported in a patient with multiple sclerosis. We hypothesize that loss of the aortomesenteric fat pad on the background of contorted body habitus from multiple sclerosis placed the patient at risk for intermittent positional compression of his duodenum.

Introduction

Superior mesenteric artery (SMA) syndrome is a rare but serious cause of proximal small bowel obstruction, whereby the third part of the duodenum (D3) is compressed in the acute angle between the SMA and the aorta. It remains challenging to identify on solely clinical grounds, and radiological investigations are the basis of establishing a definitive diagnosis. This patient's presentation was made more atypical by the presence of multiple sclerosis (MS) and lack of risk factors. This unusual case adds to the sparse literature on SMA syndrome and its heterogeneous presentation.

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Case presentation

A 42-year-old male presented with 7 days of vomiting and nil flatus. He had not opened his bowels for 9 days. He denied pain, recent weight loss, urinary symptoms, systemic symptoms, sick contacts nor recent travel. He reported a similar episode 2 months prior with spontaneous resolution.

Past history consisted of non-ST elevation myocardial infarction, depression, and secondary progressive MS, leaving the patient wheelchair bound. One month prior, the patient had suffered a left supracondylar femoral fracture following a fall from sitting height, which had been repaired with open reduction and internal fixation. He had undergone postoperative rehabilitation and regained his premorbid level of function. His regular oral medications were daily amitriptyline 200 mg, nocte mirtazapine 45 mg, as well as twice daily (BD) carbamazepine 400 mg and docusate/senna 50/8 mg. The patient daily consumed 20 cigarettes, 2 standard drinks of alcohol, and 5 grams of marijuana.

On examination, the patient was well, overweight, and displayed chronic abnormal spinal posturing due to MS. Vital signs revealed sinus tachycardia of 105 beats per minute, blood pressure 140/74 mm Hg, temperature 37.8 °C, and oxygen saturation of 99% on room air. Chest auscultation revealed that left basal decreased air entry and crepitations. The abdomen held no scars and was moderately distended with generalized mild tenderness. There were no other findings. The patient had a long-term urethral catheter, draining clear yellow urine. A nasogastric tube (NGT) was inserted, immediately draining 3500 mL of bilious fluid.

Initial serum investigations revealed hemoglobin of 142 g/L, white cell count 20.4 $\times$ 10^9/L, platelets 365 $\times$ 10^9/L, potassium 3.3 mmol/L, estimated glomerular filtration rate >90 mL/min/1.73 m^2, and albumin 31 g/L. C-reactive-protein was 76 mg/L. Other electrolytes, liver function tests, and Troponin T were normal. Urine dipstick was positive for protein, leukocytes, and blood and culture revealed chronic colonization with *Pseudomonas aeruginosa*. Chest x-ray displayed left basal consolidation.

The impression was of proximal small bowel obstruction complicated by aspiration pneumonia. Postulated mechanisms included internal hernia or malignancy. The leading differential diagnosis was gastric outlet obstruction.

A contrast computed tomography (CT) scan of the abdomen and pelvis revealed gastro-duodenal distension with a transition point at D3 due to compression between the SMA and aorta. The aortomesenteric angle and distance were 18° and 8 mm, respectively (Fig. 1). Left lower lobe pneumonia was seen.

The patient was fasted and treated conservatively, with intravenous (IV) resuscitation and NGT on free drainage. He was prescribed IV pantoprazole 40 mg, ceftriaxone 1 g, and azithromycin 500 mg daily and metronidazole 500 mg BD, as well as subcutaneous 40 mg enoxaparin daily. All oral medications were ceased, with IV levetiracetam, 500 mg BD substituted for his usual carbamazepine.

On the third day of admission, following removal of the NGT, the patient underwent diagnostic gastroscopy to D3. Mild distal reflux esophagitis was seen, as well as multiple non-bleeding superficial clean-based gastric ulcers in the gastric fundus, body, and antrum. Biopsies of these revealed only inflammatory change, with no evidence of dysplasia nor *Helicobacter pylori*. In the duodenum, the mucosa was normal and the lumen widely patent, with no evidence of malignancy, ulceration, duodenal web nor extrinsic compression (Fig. 2). Water was flushed into D3 with easy passage downstream. The NGT was resited at the procedure's conclusion.

Barium meal with plain film follow was subsequently performed. While D2 remained dilated and D3 narrow, there was prompt passage of contrast to the small bowel narrow with no evidence of obstruction (Fig. 3).

The clinical impression was of intermittent SMA syndrome. The patient's vomiting resolved on day 3. On day 4, the patient tolerated a clear fluid diet and passed flatus, so the NGT was removed, and the patient placed on normal diet thereafter, without complaint. His bowels eventually reopened on day 7.

His pneumonia improved and antibiotics were converted to oral amoxicillin/clavulanic acid 875/125 mg BD, which were continued for 5 days. IV levetiracetam was ceased and the patient restarted on oral carbamazepine. Pantoprazole was converted to oral formulation and continued post discharge. All other usual oral medications were restarted.

The patient was discharged on day 8. Surveillance gastroscopy at 3 months confirmed resolution of his gastric ulcers. At 4-month follow-up, he had suffered no further episodes.

Discussion

SMA syndrome is a rare disorder in which D3 is compressed between the aorta and SMA. This can result in partial or complete duodenal obstruction that may present acutely, chronically, or intermittently. Variously known as Wilkie's syndrome, aortomesenteric syndrome, mesenteric root syndrome, cast syndrome, or chronic duodenal ileus, it was first described by Rokitansky in 1861 based on autopsy findings [1], with the first case series published by Wilkie in 1927 [2]. It may occur simultaneously with or distinct from Nutcracker syndrome, in which the left renal vein is compressed between the aorta and SMA [3]. Due to the syndrome's rarity, there is a paucity of high-quality data. Since 1950, the largest case series comprises 27 patients [4].

The syndrome's pathophysiology involves a decreased aortomesenteric angle and narrowed aortomesenteric distance. Typical patients have pre-existing low values due to inadequacy of the aortomesenteric fat pad that usually supports the area, often with body mass index less than 18 kg/m^2 or weight percentile for height of <5% [3]. They may also have an abnormally high ligament of Treitz with corresponding upward fixation of the D3 into the aortomesenteric angle, or a low SMA origin [2]. A precipitating event further collapses the space occupied by D3. This may be due to weight loss, rapid height increase, aneurysm, or trauma. Significant weight loss shrinks the supportive fat pad and may be intentional or due to eating disorders, malignancy, malabsorption, or following bariatric surgery [2]. Height increase may occur due

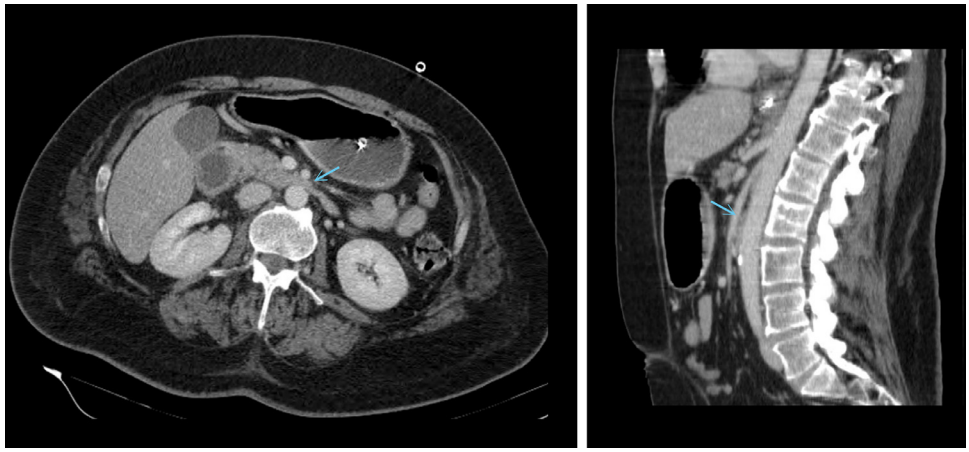

Fig. 1 – Contrast computed tomography scan of the abdomen and pelvis, arterial phase. (*Left, axial view*) The distended stomach and second part of the duodenum (D2) can be seen, followed by abrupt tapering at the third part of the duodenum (D3) (*blue arrow*), as it passes between the superior mesenteric artery (SMA) anteriorly and the aorta posteriorly. (*Right, sagittal view*) D3 (*blue arrow*) can be seen squashed the SMA and aorta. The aortomesenteric angle and distance were 18° and 8 mm, respectively. (Color version of figure is available online.)

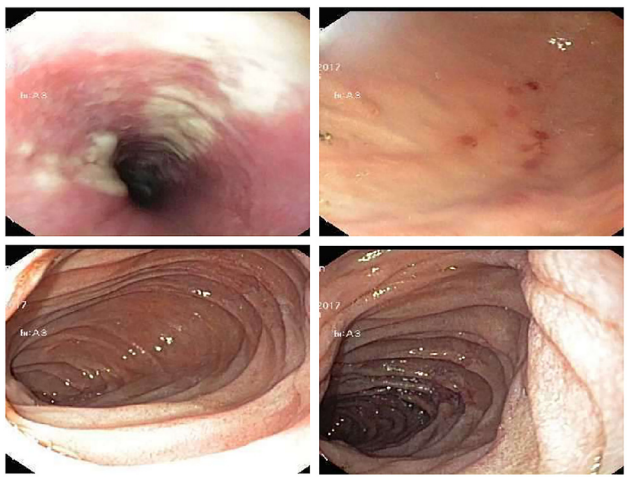

Fig. 2 – Gastroscopic views. (*Top left*) Distal esophagus displaying mild reflux esophagitis. (*Top right*) Gastric antrum, with nonbleeding superficial clean-based ulcers. (*Bottom left*) Duodenal bulb, normal in appearance. (*Bottom right*) D2 looking into D3, with widely patent lumen and no evidence of ulceration, duodenal web nor extrinsic compression.

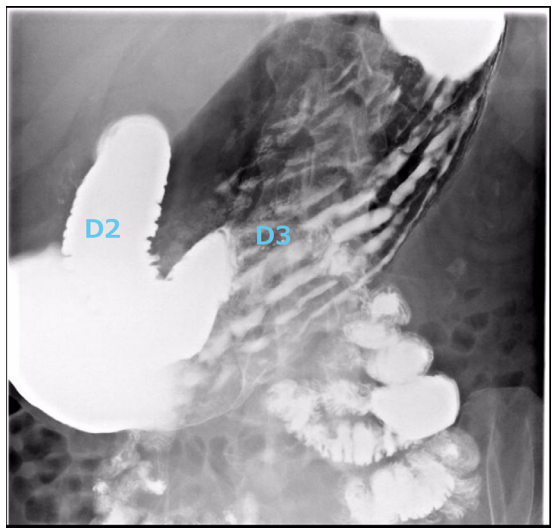

Fig. 3 – Barium meal with plain film follow, 90 minutes after barium swallow. Contrast fills the dilated D2, with D3 almost empty. However, contrast is seen to drain into the jejunum, consistent with a patent D3 lumen.

to rapid linear growth in adolescence without corresponding weight gain, or as a result of spinal surgery such as for scoliosis. Abdominal aortic aneurysm may occlude the aortomesenteric space. Trauma may also cause the syndrome through either traumatic SMA aneurysm or weight loss from prolonged immobilization from other injuries. Finally, for some patients, the syndrome may be idiopathic, as in this case.

The syndrome's epidemiology reflects the common causes, with the typical patient being female aged 10-35 years [2]. This is likely due to the timing of the growth spurt, and the greater preponderance of eating disorders and scoliosis both in young adults and in women.

Patient's present with postprandial copious vomiting, epigastric fullness, and reduced bowel motions may report recent weight loss. Symptoms may improve with maneuvers thought to reduce mesenteric tension and so increase the aortomesenteric angle, such as lying prone or in left lateral decubitus position, and worsen when supine. On examination, patients often are thin, hypovolemic, and display upper abdominal distension and tenderness. Serum testing may reveal especially hypokalemia, hypochloremia, metabolic alkalosis, and acute kidney injury.

The preferred imaging is arterial phase CT AP. The normal aortomesenteric angle and distance are 28°-65° and 10-20 mm, respectively, but in SMA syndrome, typical values are <22° and <8 mm, respectively [1,4], as seen in this case. Gastroduodenoscopy may be employed, to assess for other causes of mechanical obstruction. Barium meal with follow-through fluoroscopy is also often utilized. The first and second duodenal segments may be dilated and D3 abruptly narrowed, with associated delayed gastro-duodenal emptying. Additionally, abdominal ultrasound may be chosen in pediatric cases to reduce radiation exposure, and can assess aortomesenteric angle and distance.

Differential diagnoses for SMA syndrome are gastric outlet obstruction, strictures, duodenal web, or neoplasia of D3, or other causes of duodenal dysmotility, such as diabetes mellitus, congenital duodenal atresia, visceral myopathy, and chronic idiopathic intestinal pseudo-obstruction.

Conservative management is preferred, with decompressive NGT, resuscitation, and nutritional support to re-establish the aortomesenteric fat pad. Enteral feeding may suffice, or need parenteral supplementation. Once obstructive symptoms have resolved, oral diet can be cautiously uptitrated.

Surgery is reserved for patients with significant weight loss or failure of trial of conservative treatment. The optimum duration of such a trial is unclear [2,3]. The most common method is duodeno-jejunostomy, via open or laparoscopic approach, with the anastomosis located anterior to the SMA and in the supra-colic compartment [3]. A modification of this technique, with infracolic anastomosis, has been reported [5]. Gastro-jejunostomy is less common, reserved for patients in whom duodeno-jejunostomy has contra-indications, such as duodenal peptic ulcer disease or dense peritoneal adhesions. Another option is Strong's procedure, also known as duodenal rotation. In this intervention, bowel is positioned counter to the normal embryological midgut rotation. Distinct from these intestinal reconfiguration procedures, Pourhassan et al. demonstrated that a vascular approach is also possible [6]. In a patient with refractory SMA syndrome, they transposed the SMA to the infrarenal aorta, with complete symptom resolution on 9-month follow-up. While most patients will enjoy immediate postoperative symptom improvement, on long-term follow-up, on average only modest symptom reduction is achieved [7].

Historically, deaths from SMA syndrome have occurred due to delayed or missed diagnosis. However, if treated promptly and appropriately, the prognosis is excellent.

In this case, the patient was not an at-risk individual and no inciting cause could be found. The authors hypothesize that the patient's abnormal spinal posturing due to MS caused the duodenum to be held upward snugly into the acute aortomesenteric angle. With a chronically narrow aortomesenteric distance, changes in position and body weight would cause intermittent obstruction. Such intermittent obstruction, often positional, due to an acute aortomesenteric angle is a well-documented entity [2,3]. A literature review was performed and to the best of the authors' knowledge, an association between SMA syndrome and MS has not previously been reported.

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