



Diurnal Fasting During Ramadan Leading to Superior Mesenteric Artery Syndrome

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ABSTRACT

Superior mesenteric artery (SMA) syndrome is a favorite of anatomists and clinicians because it results from extrinsic compression of the duodenum by the 2 vascular structures forming the aortomesenteric angle (the descending abdominal aorta and the SMA). Although it is an uncommon cause of upper gastrointestinal obstruction, SMA syndrome can cause significant morbidity. It is more common in younger people. Historically, it has been associated with weight loss and eating disorders, but there are several other risk factors that should be considered in the workup. Cases of SMA syndrome are typically managed conservatively, but surgical referral and intervention may be considered in situations that fail conservative management. We present a case of this rare syndrome in a young man with no medical or psychiatric history during diurnal voluntary fasting in the month of Ramadan.

INTRODUCTION

Superior mesenteric artery (SMA) syndrome is an uncommon cause of duodenal obstruction that involves the entrapment and obstruction of the third part of the duodenum between the SMA and the aorta. The disease was first reported in 1861 by Carl von Rokitansky, but the pathology remained undefined until Sir David Percival Wilkie provided a more detailed description of the syndrome in a case series in 1927.^{1,2} Since then, the disease has gained more popularity, but much of the existing literature on the condition is limited to case reports and case series.

SMA syndrome results from narrowing of the aortomesenteric angle (the angle between the aorta and the SMA). This angle normally ranges from 38° to 65°. In most patients with SMA syndrome, the angle is reduced to $\leq 25^\circ$, and as a result, the aortomesenteric distance decreases to ≤ 8 mm, from 10 to 28 mm. This narrowing results in the compression of the third part of the duodenum because it crosses between the aorta and the SMA and may even result in compression of the left renal vein (the so called nutcracker syndrome).^{3,4} The third part of the duodenum is typically cushioned by a mesenteric fat pad as it courses through this angle. In classical cases of SMA syndrome, weight loss leads to loss of this fat pad with decreased acuity of the angle, leading to duodenal compression between the immobile vascular structures. Various conditions may result in rapid weight loss, which causes narrowing of the aortomesenteric angle and predisposes patients to developing SMA syndrome. Once the obstructive syndrome has become established, it becomes self-perpetuating with a vicious cycle of vomiting leading to further weight loss and, thus, further vomiting due to worsening duodenal obstruction.³⁻⁵

Although an accurate prevalence of SMA syndrome is uncertain, the incidence is estimated at 0.01%–0.3%. The epidemiology of SMA syndrome parallels that of eating disorders. It most often occurs in adolescents and young adults (general age range of 10–39 years) but can occur at any age. Females are more commonly affected than males with a ratio of 3:2.^{3,6}

Patients with SMA syndrome may present acutely with recurrent insidious symptomatology or with an acute exacerbation of chronic symptoms. Untreated SMA syndrome is associated with severe morbidity. Most patients are managed conservatively, with surgery reserved for refractory cases not responding to conservative treatment.⁷

Table 1. Pertinent initial laboratory results

Investigation	Result	Reference range
White blood cell count	10,200 cells/ μ L	4,400–10,600 cells/ μ L
Neutrophil %	75.9%	45.3%–74.5%
Hemoglobin	14.5 g/dL	12.9–16.8 g/dL
Hematocrit	45.0%	38.1%–49.0%
Platelet count	201,000 cells/ μ L	161,000–369,000 cells/ μ L
Lipase	200	5–55 U/L

CASE REPORT

A 26-year-old man with no medical or surgical history presented with 2 days of nausea, recurrent bilious emesis, nonradiating epigastric pain, and upper abdominal fullness. He reported that his symptoms were exacerbated after meals. He had nonspecific upper abdominal discomfort after meals for a week before the onset of these symptoms, but they were not significant enough to concern him. He had been fasting in the daytime during the month of Ramadan and endorsed a 12-pound weight loss over the 3 weeks before presentation since Ramadan started. He denied any prior episodes of emesis or postprandial pain and denied any history of peptic ulcer disease or the use of nonsteroidal anti-inflammatory drugs. He reported normal bowel movements and denied gastrointestinal bleeding.

On presentation, his blood pressure was 103/62 mm Hg; his heart rate was 94 beats/min; his respiratory rate was 17 cycles/

min; and his temperature was 36.8°C. He weighed 63 kg and was 1.86 m in height, with a body mass index of 18.2 kg/m².

Physical examination was significant for mild tenderness and diffuse fullness in the epigastric region. There were no signs of peritoneal irritation. Pertinent initial laboratory results are summarized in Table 1. Liver function tests, lipid panel, electrolytes, blood urea nitrogen, and creatinine were within normal limits.

Noncontrast abdominal computed tomography revealed a massively dilated stomach and proximal duodenum with a transition point around the region of the SMA suggestive of obstruction (Figure 1). The surrounding vascular structures were not clearly delineated given the lack of contrast, but the aortomesenteric angle was estimated to be approximately 20° with an aortomesenteric distance of 6 mm. There was no free intraperitoneal gas and no features suggestive of pancreatic inflammation on the computed tomography scan. Afterward, a barium upper gastrointestinal series with small bowel follow-through showed a markedly distended stomach with proximal duodenal dilation and a sharp transition point between the second and third parts of the duodenum near the midline in the region of the SMA (Figure 2). He was then placed on maintenance intravenous fluids and had an 18F nasogastric tube inserted with 800 mL of bilious gastric contents suctioned out.

The next morning, enteroscopy revealed Los Angeles grade B esophagitis and a large amount of food and bilious secretions in the stomach. The second portion of the duodenum appeared

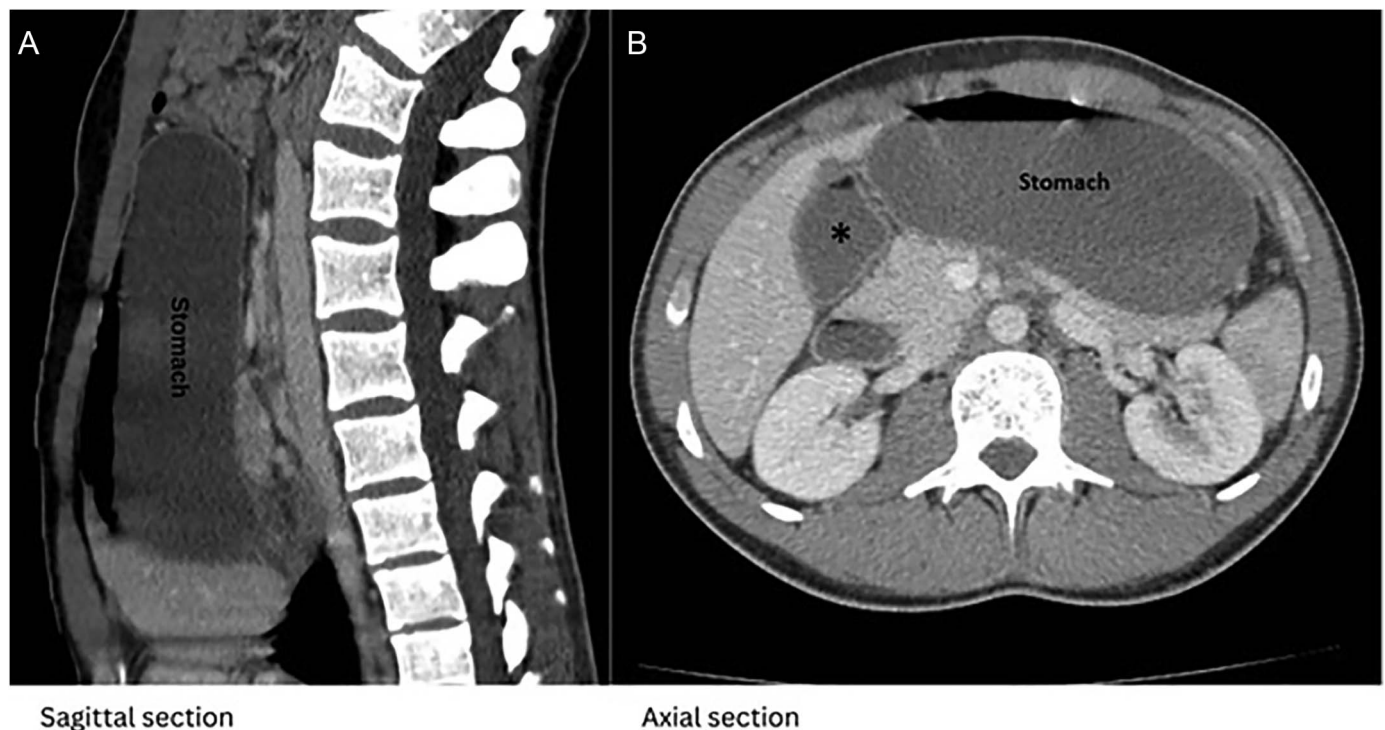


Figure 1. Abdominal computed tomography showing a massively dilated stomach and proximal duodenum (black asterisk). (A) The superior mesenteric artery is seen forming an acute angle with the aorta with an aortomesenteric angle of 20°. (B) Markedly distended stomach and proximal duodenum (black asterisk) seen on axial section.

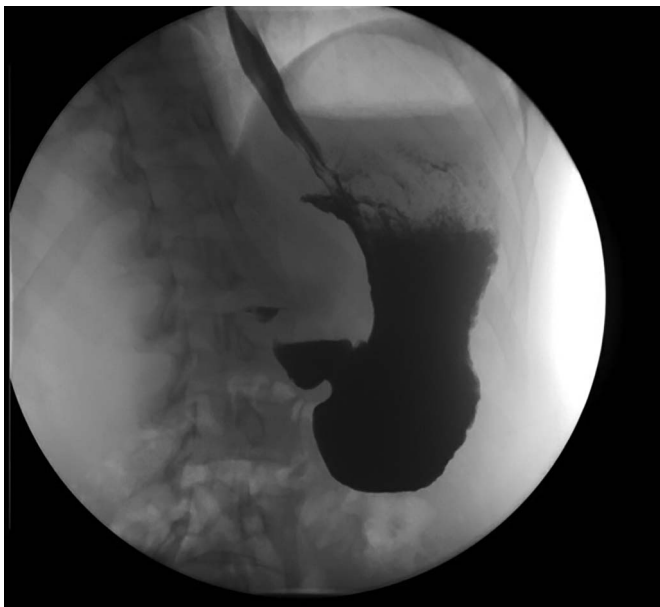


Figure 2. A markedly distended stomach with proximal duodenal dilation.

dilated, and in the distal second portion of the duodenum/proximal third portion, there seemed to be an area of angulation/extrinsic compression that opened with air insufflation (Figures 3 and 4). The scope was advanced into the proximal jejunum without resistance. These findings were suggestive of SMA syndrome. A nasojejunal tube was left beyond the abovementioned area of extrinsic compression.

The patient was managed conservatively with a high-calorie diet in small, frequent meals with liquid consistency through the nasojejunal tube. He improved clinically within a few days and was advanced to a soft diet. He was discharged to continue follow-up in the gastroenterology clinic 9 days after presentation, having gained 5 pounds while being hospitalized. He



Figure 3. Compressed/angulated area in the distal second/proximal third portion of the duodenum.



Figure 4. Angulated area highlighted above the opening with air insufflation.

remained symptom-free after discharge from the hospital, and he has regained weight appropriately.

DISCUSSION

Several factors can decrease the acuity of the angle between the aorta and the SMA. The most common is significant weight loss leading to loss of the mesenteric fat pad. Established risk factors for such weight loss include debilitating illnesses such as malignancies, acquired immune deficiency syndrome, malabsorption, and cerebral palsy; catabolic conditions such as extensive burns and trauma; and eating disorders such as anorexia nervosa. Similarly, SMA syndrome has been described after surgery-associated rapid weight loss as seen after bariatric surgery and esophagectomy.³⁻⁵ In the pediatric population, SMA syndrome can occur in otherwise healthy adolescents after insufficient weight gain relative to an increase in height.^{8,9}

Acute presentation of SMA syndrome, as in our patient, is usually characterized by upper abdominal pain, nausea, vomiting, and rapid weight loss. The pain is classically relieved in the prone or left lateral decubitus position because these positions release tension on the small bowel mesentery and increase the aortomesenteric angle. Chronic cases may present with long-standing vague abdominal symptoms or recurrent episodes of upper abdominal pain associated with vomiting, food intolerance, and weight loss. Other less common symptoms are esophageal reflux, early satiety owing to slowed gastroduodenal transit time, and gastric distension.^{3,4}

The diagnosis of SMA syndrome is challenging and often delayed because of the similarity of its presentation with an abundance of more common differential diagnoses. These differentials include acute pancreatitis; peptic ulcer disease; other causes of bowel obstruction; and diseases associated with duodenal dysmotility and megaduodenum, including diabetes

mellitus, scleroderma, and collagen vascular diseases.^{6,10} High clinical suspicion is warranted, and diagnosis is based on clinical evidence supported by radiological findings. Barium radiography demonstrates dilatation of the first and second parts of the duodenum ± gastric dilatation (Figure 2). Contrast-enhanced computed tomography with vascular reconstruction or magnetic resonance angiography enables visualization of the vascular compression of the duodenum and precise measurement of the aortomesenteric angle and distance. Endoscopic examination may visualize a pulsatile extrinsic compression.¹¹

Potential complications of untreated SMA syndrome include hypovolemia, electrolyte abnormalities, gastric perforation, gastric pneumatosis, portal venous gas, and formation of an obstructing duodenal bezoar.^{12,13} Traditionally, initial treatment of SMA syndrome consists of conservative measures with goals including gastric decompression; nutritional rehabilitation; and correction of fluid balance, electrolyte, and acid-base abnormalities. Psychiatric evaluation may be needed to assist the management of a concomitant eating disorder if present, and postural maneuvers during meals and motility agents may be helpful in some patients. Options for nutritional rehabilitation include enteral nutrition bypassing duodenal compression or total parenteral nutrition in extreme cases. However, this should be performed cautiously because accelerated nutritional rehabilitation puts patients at risk of refeeding syndrome, which may worsen electrolyte abnormalities or even precipitate cardiopulmonary complications.^{3–5,14}

Surgery may be considered if conservative treatment fails.⁴ Duodenojejunostomy is the operation of choice to relieve the obstruction, with a success rate of up to 90%. Laparoscopic duodenojejunostomy has recently replaced open bypass as the standard operative treatment. Another less invasive surgical option, known as the Strong procedure, involves lysis of the ligament of Treitz with mobilization of the duodenum. However, this procedure had a failure rate of 25% because of the short branches of the inferior pancreaticoduodenal artery not permitting sufficient duodenal mobilization. Gastrojejunostomy is no longer frequently used because of increased postoperative complications, such as blind loop syndrome, and recurrence of symptoms due to nondecompression of the duodenum.^{3–7} Postoperatively, patients are followed for resolution of their symptoms, and weight gain is monitored. Contrast studies may be performed after 1–2 weeks to demonstrate patency of the repair and normal duodenal emptying.^{15,16}

Diurnal fasting as a potential cause of SMA syndrome has not been previously established. We hope that our demonstration of such rare causation can shine light on variable etiologies of weight loss to include not only pathological causation but also benign etiologies.

SMA syndrome should be considered as part of the differential diagnosis in patients (particularly adolescents and young adults) who present with a history of persistent postprandial vomiting and weight loss. A multidisciplinary approach provides the most beneficial diagnostic and therapeutic outcomes

in this often-underestimated disease. A high index of suspicion is advised to facilitate early recognition and treatment of this syndrome.

DISCLOSURES

Author contributions: PE Ojemolon drafted the manuscript and participated in the review and final approval of the manuscript. I. Maghari drafted the case presentation and participated in the review and final approval of the manuscript. A. Almoghrabi conceptualized the manuscript, collected the cases, and revised the manuscript for intellectual content. PE Ojemolon is guarantor of the article.

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