INDEX OF SUSPICION

Abdominal Pain, Distention, and Vomiting in an Adolescent

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PRESENTATION

A previously healthy 14-year-old boy presents to an emergency department with left-sided abdominal pain and vomiting. The pain developed acutely 2 days prior and was not relieved by acetaminophen; it was sharp, intermittent, and 7 out of 10 on the pain scale and progressively worsened especially with movement. The abdominal pain was also associated with a 2-day history of abdominal distention and an inability to tolerate oral intake or take deep breaths. On presentation, he had a headache, numerous episodes of dark brown emesis, and an absence of bowel movement for 5 days; he normally has daily bowel movement. He admits to having a good appetite and eating a varied diet but reports progressively worsening early satiety with an unintentional 10-lb weight loss over the year. He denies caloric counting, excessive exercise, binge eating, purging behavior, or other past medical and psychiatric history. He has no history of fever, chills, rash, dizziness, lightheadedness, cough, congestion, throat pain, dyspnea, chest pain, night sweats, recent illnesses, past surgeries, sick contacts, recent travel, sexual activity, or drug use. His immunizations are up to date.

On presentation, his temperature is 98.5 °F (36.9 °C), heart rate is 130 beats/min, respiratory rate is 18 breaths/min, and blood pressure is 107/71 mm Hg, with oxygen saturation of 100% on room air. His height is 68.9 in (175 cm), an increase from 65.9 in (167.4 cm) over the past year, and his weight is 98.5 lb (44.7 kg), a decrease from 109 lb (49.4 kg) over the past year. His height and weight 2 years prior were 62 in (157.5 cm) and 92 lb (41.8 kg), respectively. His body mass index (BMI) is 14.5 kg/m^2 (<1st percentile), having decreased from 17.65 kg/m^2 (33rd percentile) 1 year prior. He is alert and thin and appears uncomfortable. He is tachycardic without murmurs, rubs, or gallops. Lungs are clear bilaterally. He has hypoactive bowel sounds, abdominal distention with rigidity, and generalized tenderness with mild rebound and guarding. There are no masses, hepatosplenomegaly, or costovertebral angle (CVA) tenderness. Psoas and Rovsing signs are negative. He has cool peripheral extremities and dry skin without rashes. Laboratory studies reveal an elevated white blood cell (WBC) count of 17 K/μL, hemoglobin at 19.4 g/dL, hematocrit of 56.7%, and platelets at 433 K/uL. Comprehensive metabolic panel is significant for elevated alanine transaminase at 43 U/L (reference range: 10–40 U/L) and aspartate transaminase at 87 U/L (reference range: 10–50 U/L), sodium at 148 mEq/L,

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albumin at 6.2 g/dL (reference range: 3.3–6.1 g/dL), blood urea nitrogen (BUN) at 46 mg/dL, and creatinine at 2.16 mg/dL. Lactate is 3.52 mmol/L (reference range: 0.5–2.20 mmol/L), and urinalysis reveals small blood with a specific gravity of 1.038. Abdominal radiograph demonstrates diffuse paucity of gas in the central abdomen with air-fluid levels in the gastric fundus and subhepatic region and significant stool burden (Figure 1). An intravenous (IV) line is placed, fluids are started, and an abdominal computed tomography (CT) is obtained. The CT demonstrates dilated stomach and proximal duodenum with compression of the pancreas posteriorly and branches of the celiac artery, specifically the common hepatic artery and splenic artery (Figure 2). Additionally, ischemic changes of the right hepatic lobe (Figure 3) and spleen are visualized. A nasogastric (NG) tube is placed, and 2800 mL of dark brown fluid is aspirated. General surgery and gastroenterology are consulted. He is made nil per os and admitted to the pediatric intensive care unit (PICU).

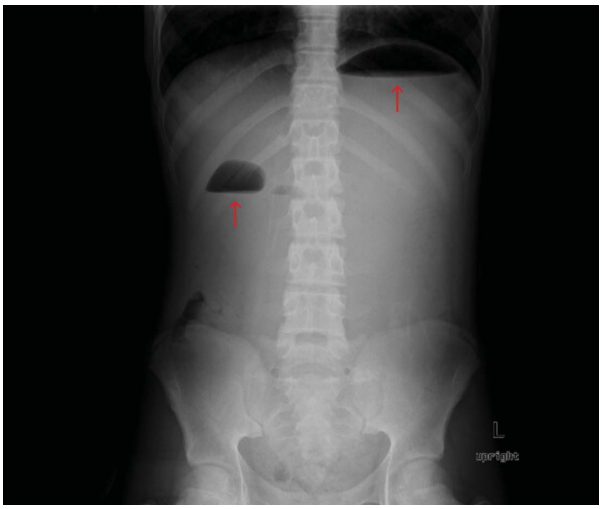

FIGURE 1. Abdominal X-Ray Demonstrating Air-Fluid Levels (arrows) in the Gastric and Subhepatic Regions. Arrows indicate the air-fluid levels in the respective regions.

DISCUSSION

Differential Diagnosis

In this adolescent boy presenting with vomiting, acute abdominal pain, and distention associated with early satiety and chronic weight loss, with exam findings significant for hypoactive bowel sounds; abdominal tenderness; rebound and guarding; and laboratory evidence of dehydration, acute kidney injury, and hepatosplenic ischemia, the differential diagnosis is broad. Initial differential diagnosis included gastrointestinal and urologic causes (appendicitis, constipation, urinary tract infection [UTI], nephrolithiasis) and intestinal obstruction (volvulus, adhesions, bezoar, tumor). Other etiologies on the differential diagnosis list include rheumatologic and systemic conditions (systemic lupus erythematosus [SLE], scleroderma, celiac disease, HIV, neoplasm, hereditary angioedema) and motility-related disorders, such as pseudo-obstruction and psychological conditions (bulimia, anorexia nervosa). Although appendicitis, UTI, and nephrolithiasis can present with acute abdominal pain, nausea, vomiting, and elevated WBC count, the location of the pain and lack of supportive

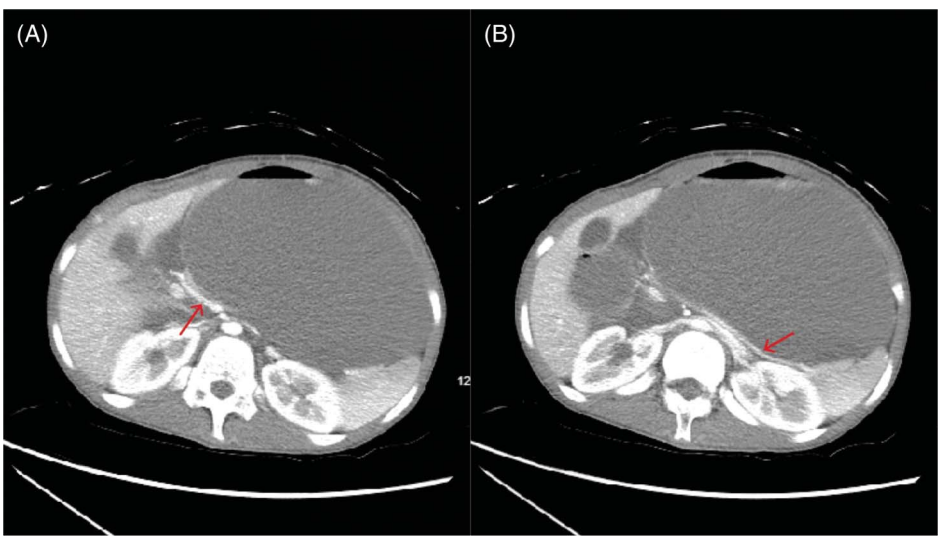

FIGURE 2. Axial Computed Tomography Demonstrating Compressions Caused by Dilated Stomach and Proximal Duodenum. Compressions are shown in the common hepatic artery (A; arrow) and splenic artery (B; arrow).

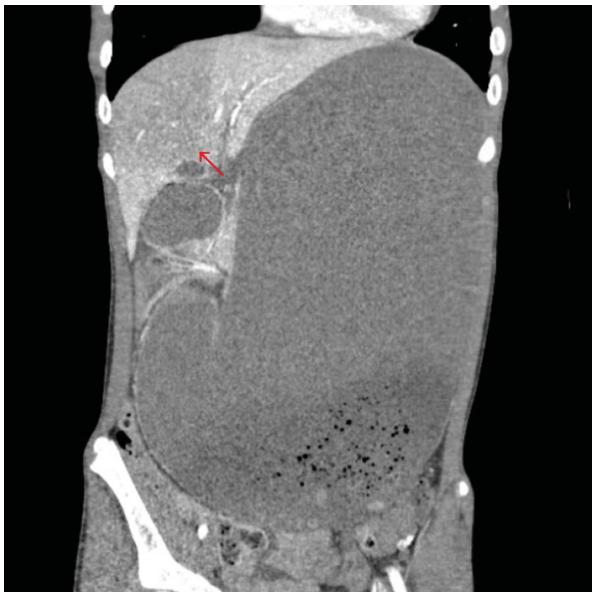

FIGURE 3. Sagittal Computed Tomography of the Abdomen Demonstrating a Grossly Distended Stomach and Ischemic Changes. Ischemic changes (arrow) to the right hepatic lobe are shown.

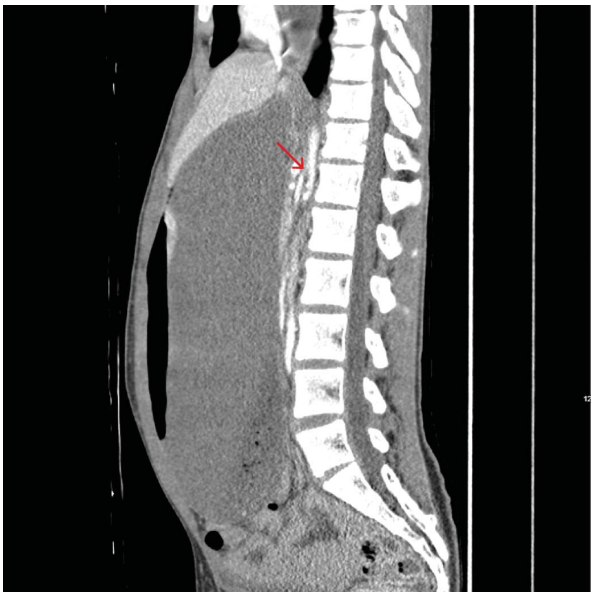

FIGURE 4. Sagittal Computed Tomography of the Abdomen Demonstrating a Narrow Aortomesenteric Angle and a Grossly Distended Stomach. The arrow shows the aortomesenteric angle.

findings, such as fever, psoas or Rovsing signs, and CVA tenderness, made these unlikely. Volvulus and intestinal tumor were considered given the presentation and small bowel obstruction on radiograph but were ruled out by abdominal CT. There was no history of foreign body ingestion or psychiatric illness to suggest bezoar or surgeries to support adhesions. Complete blood count (CBC) results were not consistent with neoplasm, although this does not preclude neoplasm. Hereditary angioedema, which can cause abdominal pain and distention, was ruled out with normal complement levels and function. Scleroderma and SLE were ruled out given negative anti-centromere, anti–double-stranded DNA, and anti-Smith antibodies. Although the chronic weight loss raised concerns for HIV, celiac disease, hyperthyroidism, and inflammatory conditions, the lack of supportive history, such as diarrhea and rash, and negative laboratory findings (HIV antibody/antigen, tissue transglutaminase antibody immunoglobulin A [IgA], total IgA, thyroid-stimulating hormone, free T4, erythrocyte sedimentation rate, C-reactive protein, and fecal calprotectin) made these unlikely.

Actual Diagnosis

The detailed CT report revealed compression of the third part of the duodenum (Figure 4) by the superior mesenteric artery (SMA), confirming the diagnosis of SMA syndrome. The angle between the SMA and the aorta was measured to be 22° (normal, 38°–65°). An angle of 25° or less is the most sensitive diagnostic measure for SMA syndrome.[1–3]

The Condition

SMA syndrome is a rare condition defined by compression of the third portion of the duodenum between the SMA and the abdominal aorta (Figure 5).[1] The SMA arises from the aorta at the L1 vertebral level and forms an acute angle (normally 38°–65°) maintained by the intervening mesenteric fat (Figure 5).[1,4] The incidence of SMA syndrome is estimated to be between 0.01% and 0.3%.[3,5] Additionally, females

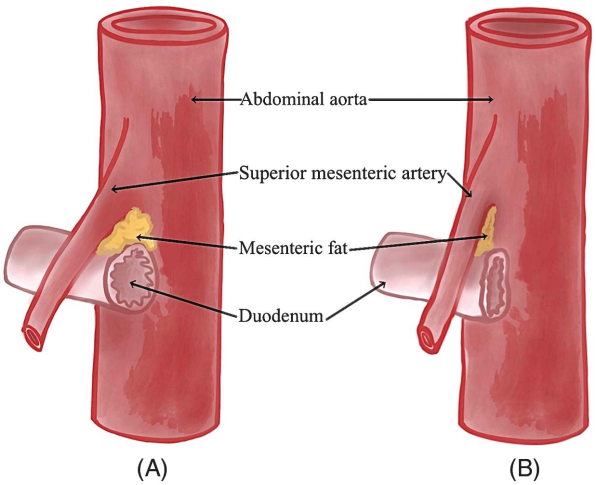

FIGURE 5. Schematic View of Normal Mesenteric Fat and Angle and Compression of the Duodenum due to Loss of Mesenteric Fat. Panel (A) shows normal mesenteric fat distribution and angle. Panel (B) shows compression of the duodenum as a result of loss of mesenteric fat. Artist: Maame Hayfron.

and young adults are more commonly affected by SMA syndrome.[6]

Conditions that can predispose one to the narrowing of this angle include illnesses associated with significant weight loss such as malignancy, malabsorption, hyperthyroidism, and anorexia nervosa.[7,8] In this patient, eating disorders were ruled out after evaluation by a psychiatry service. His progressively worsening early satiety and weight loss over the past year were preceded by a significant increase in height, as evidenced by his growth parameters. Rapid linear growth during puberty has been shown to result in loss of mesenteric fat and SMA syndrome even in the absence of weight loss.[6,9] This patient's pubertal growth likely resulted in the initial loss of mesenteric fat and gradual narrowing of the aortomesenteric space with subsequent early satiety and weight loss. Other conditions include prolonged supine bed rest, trauma (burns or neurological injury), postoperative states (Nissen fundoplication, scoliosis surgery, spinal instrumentation), and anatomical variations of the ligament of Treitz.[6,8]

The clinical presentation of SMA syndrome involves both acute and chronic nonspecific symptoms, including postprandial epigastric abdominal pain, vomiting, nausea, early satiety, anorexia, and bloating, in order of decreasing frequency.[6,10] The pain is usually relieved by lying prone, knee to chest, or in the left lateral decubitus position.[3,8]

Imaging plays an important role in diagnosing SMA syndrome. Abdominal radiographs are often nonspecific and may reveal evidence of obstruction (Figure 1). In this patient, the obstruction was presumed to be at the level of the duodenum. Although barium studies may reveal failure of contrast passage beyond the third part of the duodenum, CT scan and magnetic resonance angiography aid in confirming an aortomesenteric angle less than 25° or a parallel distance between the aorta and SMA of less than 8 mm (normal, 10–28 mm).[1,2,11,12] Similarly, the angle can be measured during surgical intervention.[6]

Patients with SMA syndrome are at high risk of developing complications including malnutrition, dehydration, poor weight gain, nutcracker syndrome (left renal vein compression between the aorta and SMA), abdominal compartment syndrome (increased intra-abdominal pressure that leads to ischemia and multiorgan failure), and death.[6,10,13–15] Our patient's findings of hematuria, elevated BUN, creatinine, lactic acid, and abnormal transaminases coupled with ischemic changes to the liver (Figure 3) and spleen were due to the compressive effects of the stomach distention on surrounding structures (Figure 2). This is consistent with previous reports of abdominal compartment syndrome secondary to severe SMA syndrome as a result of delayed diagnosis.[10] Increased intraabdominal pressure can lead to compression of the inferior vena cava and thus a reduction in preload and cardiac output.[16] Consequently, diminished splanchnic flow, compounded by impaired venous drainage, can cause renal failure, prolonged bowel ischemia, and inability of the liver to clear lactate.[10]

Treatment/Management

Management of SMA syndrome is usually conservative and involves insertion of an NG tube for gastroduodenal decompression as well as IV fluids for correction of electrolyte and water imbalance.[1,8] Patients can also be placed in a knee-to-chest or left lateral decubitus position to temporarily increase the aortomesenteric angle.[2,6] Efforts to improve weight and promote an increase in mesenteric fat include enteral nutrition via a postpyloric tube, such as a nasojejunal (NJ) tube, or via total parenteral nutrition (TPN).[6] Surgical interventions, such as laparoscopic or open duodenojejunostomy (an anastomosis between the proximal duodenum, before the point of obstruction, and a loop of jejunum beyond the ligament of Treitz), may be required depending on the severity or if conservative management fails.[2,17,18] Recent studies have found laparoscopic duodenojejunostomy to be more effective, feasible, and safer than open surgery.[17]

Patient Course

The patient's PICU course was complicated by hepatosplenic ischemia, hypernatremic dehydration, and acute kidney injury. After fluid resuscitation, interventional radiology was consulted. However, initial attempts at NJ tube placement beyond the obstruction were unsuccessful. His initial daily NG output was greater than 1000 mL; this eventually decreased over the hospital course. By hospital day 3, his hydration status, CBC, kidney function, and electrolytes had returned to normal. He was transferred to the general pediatric floor and received further workup for underlying causes of weight loss. He was started on TPN and lipids until an attempt at NJ tube was successful. Repeat lactate levels also returned to normal, and serial imaging revealed an improved SMA angle (29°) with resolution of the hepatosplenic ischemia. His abdominal pain, distention, and weight gradually improved, and by hospital day 22, he was started on a slow oral diet. He was discharged home on hospital day 50 with outpatient gastroenterology and neurogastroenterology follow-up. At 6-month outpatient follow-up, he continued to eat normally with no recurrence of symptoms and had increased his BMI to the 26th percentile with a weight of 121 lb (55 kg).

LESSONS FOR THE CLINICIAN

- SMA syndrome is a rare cause of intestinal obstruction with increased morbidity and mortality when there is a delay in diagnosis.
- When evaluating early pubertal adolescents with abdominal pain and postprandial vomiting, pediatricians should have a high index of suspicion for SMA syndrome even in the absence of weight loss.
- Although conservative management and medical treatment are usually recommended as initial therapy, surgery may be required for severe or refractory cases.
- Laparoscopic duodenojejunostomy is proven to be safer, faster, and more effective than the traditional open surgery.

References for this article can be found at
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