CASE REPORTS

RECURRENT SMA SYNDROME IN A TEENAGE FEMALE

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Abstract

Recurrent superior mesenteric artery (SMA) syndrome has been described in literature, though the true incidence of recurrence is not well known. A high index of suspicion is required for the diagnosis of recurrent SMA syndrome. A 13 year-old female with a previous history of SMA syndrome presented to the emergency department with an acute onset of vomiting and abdominal pain without associated weight loss. She was found to have a recurrence of her SMA syndrome. Imaging revealed a narrow aorta-SMA angle. Even in the absence of weight loss and an atypical presentation of symptoms, a history of SMA syndrome may indicate an anatomic predisposition to the condition and needs to be considered.

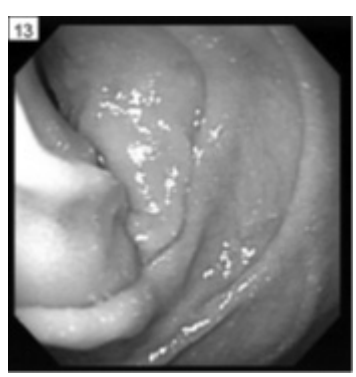
Introduction

Acute superior mesenteric artery (SMA) syndrome is a rare cause of intestinal obstruction occurring secondary to compression of the third portion of the duodenum by the superior mesenteric artery. (1, 2) The incidence of SMA syndrome is estimated between 0.013-0.3% of the population. (3) In the pediatric population, it presents most commonly secondary to acute weight loss such as with anorexia nervosa, or following spinal surgery such as corrective surgery for scoliosis (also known as cast syndrome). (1, 3-5) There is a female predominance to the condition. (3, 6) Diagnosis requires a high degree of clinical suspicion and can be confirmed by CT imaging. Treatment is primarily supportive care and recurrence in the pediatric population is rare, having only been described in single case reports in the literature. (2) A single retrospective study in South Korea reported recurrence of SMA syndrome in the adult population as high as 15.8%, though recurrence rates are otherwise not well reported (7). Here, we describe a 13 year-old female with a previous history of SMA syndrome who presented to the emergency department with acute onset of vomiting and abdominal pain without associated weight loss. She was found to have a recurrence of SMA syndrome. Imaging revealed a narrow aorta-SMA angle.

Case Report

A 13 year-old female presented to the emergency department with 2 days of vomiting and epigastric abdominal pain. She began experiencing abdominal pain in the evening that progressed after a few hours to include nausea and vomiting. The vomiting occurred almost hourly, but was worsened by oral intake and would occur within 15 minutes of oral intake. It was non-bloody and non-bilious, with no associated fevers, diarrhea, constipation, dysuria, or change in the appearance of her urine. Importantly, she had not experienced any preceding weight loss. She denied sexual activity and her last menses were 2 months prior. Her prior medical history was significant for an episode of acute SMA syndrome diagnosed four years prior in the setting of an acute presentation of vomiting and abdominal pain with recent weight loss. She also has a history of grade 1 spondylolisthesis. On initial examination, her vital signs revealed tachycardia with a pulse of at 103/minute and a normal body mass index [BMI] of 19.5 (50th percentile). There was epigastric abdominal tenderness. Investigations showed leucocytosis (white cell count of 11,300 cells/cumm) with normal liver and renal function tests, amylase, lipase, ESR, CRP, and urinalysis. Urine pregnancy test was negative. She was admitted to the hospital and continued to experience emesis especially after eating or drinking. On hospital day 2 an upper gastrointestinal series with barium showed dilatation of the first, second and proximal third portions of the duodenum. Contrast was not able to cross the midline in the supine position with minimal passage with patient in the right lateral position. A tentative diagnosis of recurrence of her SMA syndrome was made and a nasojejunal tube was placed. Endoscopy revealed a dilated 2nd portion of the duodenum and narrowing of the 3rd portion with no intrinsic duodenal abnormalities (Figure 1). CT scan of the abdomen with contrast showed narrowing of the aorto-superior-mesenteric-artery angle at 25 degrees. After supplementation with nasojejunal feeds her vomiting improved and she was discharged home on day 14.

Figure 1. Esophagogastroduodenoscopy showing narrowing of the 3rd portion of the duodenum at the level of the SMA.



Discussion

SMA syndrome is a rare cause of intestinal obstruction with a reported incidence of 0.013-0.3%. (1, 8) It is thought to occur secondary to a decrease in amount of retroperitoneal fat separating the aorta and superior mesenteric artery. The 3rd portion of the duodenum runs between these two vessels. In patients with certain predisposing factors such as a narrowed aorto-SMA angle or a "high-riding" duodenum, loss of this fat "cushion" can allow for extrinsic compression and obstruction of the 3rd portion of the duodenum by the SMA. (4,5) On an average, the SMA branches of the abdominal aorta at a 45 degree angle, with a reported range of 38-68 degrees. (5,6) The patient reported here demonstrated a slightly narrowed aorto-SMA angle at 25 degrees.

In the pediatric population, SMA syndrome

most commonly occurs following rapid weight loss, growth without concurrent weight gain, or in orthopedic patients with casting after spinal surgery. It may be associated with malabsorptive disorders, gastroenteritis, anorexia nervosa, spinal deformities or injury, or hyperthyroidism. (1,4,8) It occurs more frequently in females and adolescents, with some studies reporting a median age of presentation of 13 years. (1, 6) Our patient was also 13 years old but she had no predisposing factors except for a previous episode of acute SMA syndrome. Typically, patients present with a protracted history of symptoms including nausea, post-prandial epigastric abdominal pain, emesis, and anorexia. (9) However, acute presentations of this syndrome have been reported. (1,10). When SMA syndrome is being considered based on a patients history and physical exam additional diagnostic studies include radiographic investigations. These begin with plain abdominal x-rays typically followed by studies involving oral contrast such as an upper gastrointestinal series or a CT of the abdomen with oral contrast. Additional investigations may include CT arteriography or MR arteriography. (3,4,6) Treatment includes gastric decompression, correction of any electrolyte or fluid abnormalities and providing adequate nutrition. (3) In most cases nutritional support can typically be provided enterally via feeds past the level of the obstruction through the use of a nasojejunal tube. (3,6)

In conclusion, a high index of suspicion is required for the diagnosis of recurrent SMA syndrome. Even in the absence of weight loss and an atypical presentation of symptoms, a history of SMA syndrome may indicate an anatomic predisposition to the condition and needs to be considered.

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