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# Superior Mesenteric Artery Syndrome in a 15-year-old Patient with a Possible Early Sign of Acute Appendicitis - A Case Report

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## ABSTRACT

Superior mesenteric artery (SMA) syndrome is an uncommon condition that involves compression of the third portion of the duodenum, which is the upper part of the small intestines just past the stomach. This condition occurs when the third part of the duodenum is compressed between two arteries—the main artery of the body called the abdominal aorta (AA) and one of its branches called the SMA. The SMA provides blood supply to the small intestines and the first part of the colon. Compression of the SMA against the AA can prevent duodenal contents from draining into the jejunum (upper small intestine), hence the inability to get proper nutrition, leading to weight loss and malnutrition.

Appendicitis is the most common indication for emergency abdominal surgery in childhood and is diagnosed in 1 to 8 percent of children evaluated urgently for abdominal pain. In the United States, the incidence increases from an annual rate of one to six per 10,000 children between birth and four years of age to 19 to 28 per 10,000 children younger than 14 years. It is frequent mostly in the second decade of life. Common signs and symptoms include anorexia, periumbilical pain (early), vomiting after onset of pain and migration to lower quadrant.

This report would primarily address a case of superior mesenteric artery syndrome and its complexities. Due to the condition's nebulous and non-specific presenting symptoms, it was initially believed to be early appendicitis. Also discussed here is how imaging modalities, in particular contrast-enhanced CT scans, can be used to diagnose and assist in patient management.

**Key words:** Superior mesenteric artery syndrome, acute appendicitis

Superior mesenteric artery (SMA) syndrome is an uncommon, acquired cause of vascular compression disorder with resultant obstruction of the third part of the duodenum, with an incidence of 0.1–0.3%.<sup>1</sup> Patients are usually presented with indeterminate and nonspecific symptoms such as nausea, vomiting, epigastric pain, early fullness and discomfort following a meal, abdominal distension, and weight loss, which can mimic acid peptic disease, pancreatitis, and cholelithiasis. Delayed diagnosis may result in significant morbidity and mortality, making it a crucial differential to consider when

a patient does not present a common disease that presents the mentioned symptoms.

Normally, the superior mesenteric artery arises anteriorly from the abdominal aorta at the level of L1, behind the neck of the pancreas and splenic vein, crossing anteriorly to the left renal vein. It emerges anterior to the uncinate process of the pancreas, crosses anterior to the third part of the duodenum, and enters the upper portion of the small bowel mesentery. It then runs along the root of the mesentery downwards to the right. Its branches (jejunum and ileum) are given off to the left, and the proximal and midcolon are given off to the right.

The duodenum is a 20-30 cm C-shaped segment of the small intestine and is a continuation of the pylorus. It is composed of four distinct parts. The first portion

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of the duodenum (D1), located between the pylorus and the gastroduodenal artery, is divided into two parts, the proximal part being free, and the distal part fixed, while D2 and D3 are fixed by close vascular attachments with the pancreas. D2 is where the main bile duct and the main pancreatic duct typically open into the duodenal lumen through the papilla of Vater. D4 is located to the left of the superior mesenteric vessels; it is mobile, but fixed posteriorly by the ligament of Treitz.

Radiologic investigation, such as upper gastrointestinal (GI) series and contrast-enhanced abdominal computed tomography (CT), may be pursued by radiologists and clinicians who have a high index of suspicion based on symptomatology. Other imaging modalities such as Magnetic resonance imaging (MRI) and ultrasound (US) are less commonly utilized in the work-up of SMA syndrome but provide imaging alternatives without the use of ionizing radiation. Reduction of the aortomesenteric angle, aortomesenteric distance, presence of gastroduodenal distention, bowel narrowing as well as delayed gastric emptying or positional obstruction can be observed with fluoroscopy.

The objective was to present a rare case of superior mesenteric syndrome in an adolescent patient.

## **THE CASE**

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A 15-year-old female student came into the emergency room presenting with 1-day persistence of epigastric pain that radiates to the periumbilical region and is described as sharp and continuous, with a pain scale of 7/10 associated with episodes of bilious vomiting of about ½ cup per bout, nausea, loss of appetite, and generalized body weakness, unrelieved by taking an anti-cholinergic spasmolytic drug every 8 hours (Hyoscine N-butylbromide 10mg/tablet). No fever, dysuria, constipation, or diarrhea were noted. In the interim, previous episodes of epigastric pain and bloatedness aggravated by supine position but are usually relieved through time and a certain position “pronation” were reported prior to consultation. The parent noted an increase in the height of her daughter over the past few years without visual signs of weight gain. The patient reported attending several Christmas parties and drinking huge amounts of milk tea and other food with friends.

In the emergency setting, the patients initial vital signs were normal, with a height of 153 cm, a weight of 43 kg, and a normal BMI of 18.4 (21st percentile

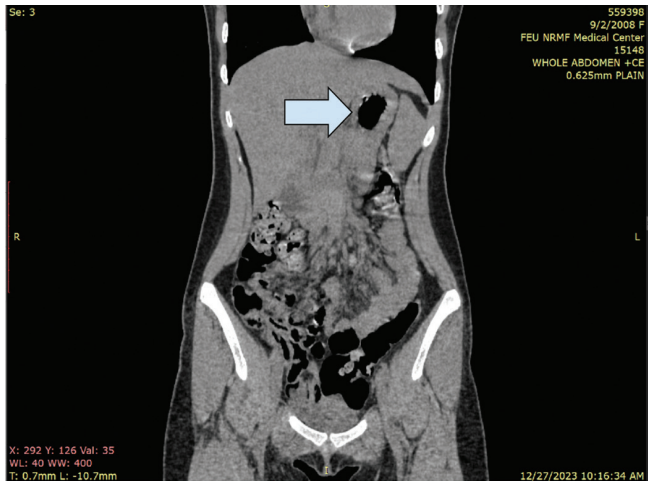
indicating healthy weight). A physical examination of the abdomen revealed hyperactive bowel sounds—tympanitic in all, with minimal tenderness in the periumbilical region. Blood test results showed a WBC count of  $11.9 \times 10^9/L$  (elevated) and neutrophils of 92.9 (elevated), chemistries (sodium 139.9 (normal), potassium 4.0 (normal), creatinine 52.0  $\mu\text{mol/L}$ ), pregnancy test, and ECG showed no significant result, still with nausea, vomiting and generalized body weakness, acute appendicitis was considered.

Elevated wbc count and inconclusive abdominal physical exam led to a plain CT scan being performed. Findings revealed a markedly distended stomach with abrupt narrowing seen in the distal duodenum, likely transverse D3 segment before crossing the aorta. (Figure 1) Minimal mottled densities are seen within the ascending, transverse and descending colons. The appendix is normal in size, measuring 0.5 cm in its widest AP diameter with no surrounding inflammatory fat stranding. No enlarged intraabdominal lymph nodes were seen. The liver is normal in size with normal morphology and tissue attenuation. No mass lesions were noted. The bile ducts are normal in caliber. The gallbladder is unenlarged with no wall thickening or opaque intraluminal lithiasis. The pancreas, spleen, and adrenals are grossly unremarkable. Both kidneys are normal in size with a normal reniform configuration. There is no opaque lithiasis or hydronephrosis. The urinary bladder is empty. The uterus is normal in size and anteverted, measuring 4.4cm x 5.3cm x 3.4cm. No discrete nodule was seen. Both ovaries are normal in size. No adnexal mass was seen. Minimal fluid collection is seen in the posterior cul-de-sac. Final impression includes consideration of superior mesenteric artery syndrome wherein contrast enhanced CT scan was recommended.

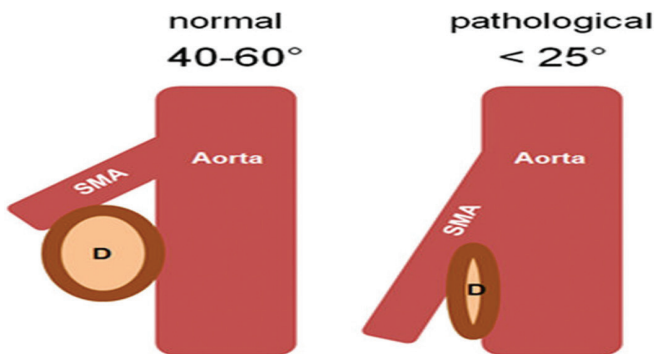
On the interim, patient was managed conservatively with insertion of nasogastric tube, fluid resuscitation and placement on a non per orem diet. A computed tomography scan with contrast was done for further evaluation of the previously noted marked distention of the stomach, showing a significant reduction in size. (Figure 2) However, there still appears to be abrupt narrowing seen within the distal duodenum, likely within the transverse segment D3, which shows a reduction in caliber and appears to be compressed by the superior mesenteric artery and abdominal aorta. The following measurement shows the aortomesenteric distance of about 0.3 cm and the aortomesenteric angle of 17°.



**Figure 1.** Markedly distended stomach with abrupt narrowing seen in the distal duodenum, likely transverse D3 segment before crossing the aorta, and normal-size appendix

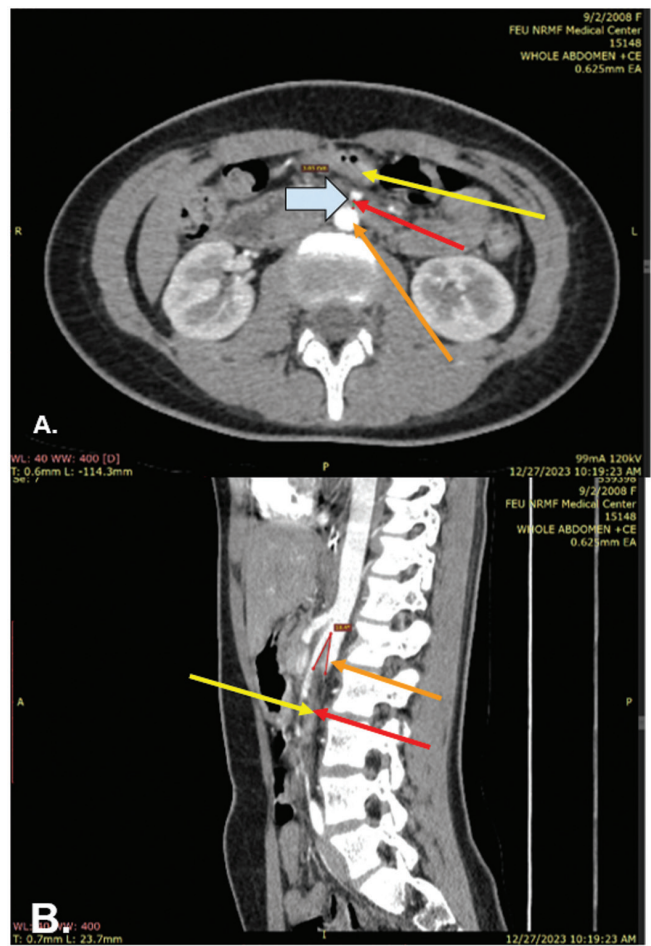


**Figure 2.** Reduced stomach distention. (Arrow)



**Figure 3.** Illustration shows a normal aortomesenteric distance and aortomesenteric angle. In contrast to a pathologically low angle of  $< 25^\circ$ .

Continued conservative management was done with hospitalized observation, nasogastric tube insertion, symptomatic medication, and interval diet



**Figure 4.** Aortomesenteric distance and [B.] aortomesenteric angle of the patient with SMA syndrome. (Arrows: Superior mesenteric artery; Orange: Abdominal aorta; Red: Duodenum).

changes. No progressing symptom was noted during the course of the hospital stay and was deemed improved. The patient was later discharged and advised follow-up with an upper gastrointestinal series with contrast.

On follow-up, the patient underwent an upper GI series, and the findings revealed unremarkable results. As of today, the patient is apparently well and trying to gain weight with small, frequent feedings. There was no recurrence of vomiting or abdominal bloating. Follow up with her consultant every other month for a check-up was done.

## DISCUSSION

Duodenal compression secondary to vascular compression, such as superior mesenteric artery syndrome, also known as Wilkie's syndrome, is a rare but well-recognized clinical entity with an

incidence ranging from 0.1–0.3% as a potential cause of abdominal pain. Traditionally, this is common in young females with anorexia or hyperemetic states and a history of rapid weight loss. Females are more commonly affected than males, with a female-to-male ratio of 3:2. It is more common in patients aged 10 to 39 years old and even rarer in children. Acute or chronic nonspecific signs and symptoms contribute to its difficulty in diagnosis.

The most common symptoms of having superior mesenteric artery syndrome reported are the following: epigastric pain, nausea, and vomiting. Other symptoms include abdominal distention, early satiety, weight loss, and postprandial epigastric pain, which worsens in the supine position and is alleviated with the release of the angle through pronation or the left lateral decubitus position. The intermittent or chronic pain scale depends on the severity of the obstruction. Severe intestinal obstruction in the acute phase leads to a life-threatening dilatation of the stomach. Non-specific and intermittent postprandial pain, nausea, vomiting, and weight loss are common in chronic states. The recurrent cycle of nausea and vomiting causes insufficient food intake, which in turn causes weight loss and exacerbates the syndrome. The aforementioned symptoms may be mistaken for an abdominal emergency. In this case, acute appendicitis was suspected, being the most common cause of abdominal surgery in childhood.

Myung Seok Shin, et al. conducted a retrospective study on eighteen children with SMAS. The reviewed data included demographics, co-morbid conditions, clinical courses, nutritional status, treatments, and outcomes in addition to presenting symptoms. Postprandial discomfort (67.7%), abdominal pain (61.1%), and early satiety (50%) were the three most prevalent symptoms. Before a diagnosis, the symptoms persisted for a median of 68 days. Weight loss (50%), growth spurt (22.2%), and bile reflux gastropathy (16.7%) were the most common co-morbid conditions. In 72.2% of the patients, the body mass index (BMI) was normal. Thirteen (72.2%) of the patients had successful medical management. The treatment lasted a median of forty-five days. It was found that 5 patients (27.8%) had moderate outcomes, 4 patients (22.2%) had poor outcomes, and 9 patients (50%) had good outcomes without recurrence. Poorer results were generally linked to a duration of medical management of more than six weeks ( $p = 0.018$ ). Patients with normal BMIs or no weight loss frequently developed SMAS. Children

with SMAS should be treated medically for at least six weeks before considering surgical treatment because medical treatment has a high success rate.

According to Akin, et al., up to 40% of superior mesenteric artery syndrome cases have no cause. Postprandial epigastric pain, bilious vomiting, nausea and early heartburn are the symptoms of acute angulation between the superior mesenteric artery and the aorta. Three categories remain for the factors influencing this condition: 1) marked weight loss resulting in the loss or depletion of retroperitoneal fatty tissues; 2) external compression using belts or spica casts; and 3) anatomical defects and congenital anomalies, such as scoliosis correction surgery, a high insertion of the duodenum at the ligament of Treitz, along with other conditions. SMA in a young population such as ours is generally unexpected due to the lack of classic risk factors, namely, severe weight loss, psychological disorders, or spinal surgery.

Currently, the cause of SMA syndrome development in children who do not lose weight is unknown, as per Okomoto et al.'s case study. However, there is conjecture that a lack of weight gain in relation to height results in a reduction of visceral fat and puts kids at risk for SMA syndrome. Acute fluid intake and delayed food passage at the duodenum can result in a temporary blockage that goes away with postural adjustments.

CT and magnetic resonance angiography (CTA/MRA) enable visualization of vascular compression of the duodenum and measurement of aortomesenteric distance. Van Horne (2023) reports that in an investigation that examined 8 cases of superior mesenteric artery syndrome, a distance of 8 mm was found to be both sensitive and specific for the condition, and an aortomesenteric angle cutoff of 22 degrees revealed a 42.8% sensitivity and 100% specificity.<sup>4</sup>

Aortomesenteric (AOM) angles less than 22 to 28 degrees with an AOM distance between 2–8 mm are strongly suggestive of SMA syndrome in the correct patient setting. The normal AOM angle is between 45 and 60 degrees, and the normal AOM distance is between 10–20mm. As these values drop, the likelihood of SMA syndrome increases when paired with any one symptom of SMA syndrome (postprandial epigastric pain, anorexia, vomiting, or weight loss). These values can be obtained from a CT or US. However, computed tomography was done on our patient for her comfort due to excessive abdominal pain and vomiting.

Laboratory tests are usually non-diagnostic, and it has been noted that electrolyte imbalances, as well as protein and albumin levels, can remain normal despite concurrent weight loss. Although uncommon, superior mesenteric artery syndrome warrants consideration due to the substantial morbidity and mortality that can arise from inadequate nutrition, dehydration, and other factors if diagnosis is delayed. Electrolyte imbalances, gastric pneumatosis, portal venous gas, gastrointestinal hemorrhage, and gastric perforation.

Usually, conservative, non-operative medical management is the first course of treatment. This comprises the acute care management protocol's nasogastric tube insertion for gastric decompression, electrolyte replacement, total parenteral nutrition, and fluid resuscitation. The patient is advised to follow posture therapy instructions, which involve lying in the left lateral decubitus position rather than the recombinant position, and to eat small meals to help relieve symptoms. In conjunction with conservative therapy, nutritional support through hyperalimentation is vital in an effort to increase the mesenteric fat pad, raise the AO angle, and alleviate symptoms. If the patient is unable to tolerate conservative measures, then surgery is required with a duodenojejunostomy or gastrojejunostomy. Surgical correction is successful in upwards of 90% of patients. A delay in diagnosis of SMA syndrome can lead to bowel obstruction, perforation, and death.

There have been reports of several SMA syndrome complications. Notably, even in young patients, undiagnosed or severe cases can worsen and lead to potentially fatal consequences like aspiration pneumonia, hypovolemic shock and sudden death. The exact cause of sudden death is unknown, but a number of theories, supported by autopsy reports and published cases, can be put forth. These include severe hypokalemia-induced arrhythmia, dilated duodenum-induced severe compression of the inferior vena cava, and severe pulmonary depression brought on by alkalosis and elevated abdominal pressure. Thus, in severe cases, early reduction of intestinal pressure and prompt adjustments to blood volume and electrolyte imbalances should be necessary. The most common side effect is gastrointestinal damage caused by retained or refluxed peptic acid and bile acid, as well as elevated intraluminal pressure. The incidence of mucosal injury has been reported at 25%–59% in patients with SMA syndrome. Emphysema, necrosis, portal venous gas, and pneumoperitoneum can develop from chronic or inadequately treated

mucosal injuries. Acute pancreatitis and elevated pancreatic enzymes can occasionally arise from disruption of the pancreatic juice flow caused by elevated intraluminal pressure at the second portion of the duodenum. Lipase and pancreatic amylase isozyme should aid in the detection of pancreatic abnormalities because vomiting itself has been shown to elevate serum amylase (primarily from the salivary glands). Aspiration pneumonia, electrolyte imbalances, dehydration, and severe malnourishment are further consequences of frequent vomiting. Occasionally, SMA syndrome coexists with other vascular compression disorders; of these, the nutcracker phenomenon is the most common due to its anatomical proximity to SMA. This condition, which shares symptoms with SMA syndrome, arises when the left renal vein becomes compressed between the aorta and SMA.

## CONCLUSION

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Although uncommon, superior mesenteric artery syndrome should be taken into consideration whenever a patient in pediatric population (adolescent phase) complains of abdominal pain and vomiting, intestinal obstruction is suspected, common differential diagnoses are ruled out, and the cause is unknown. The early detection of such a syndrome prevents a patient from experiencing several life-threatening complications and possible surgical intervention.

In this case, the diagnosis of SMA syndrome was straight-forward, as the patient's CT imaging of the abdomen was highly suggestive of the diagnosis.

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