



## Gastric pneumatosis: An unusual presentation of superior mesenteric artery syndrome

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### ABSTRACT

We present a case of extensive gastric pneumatosis with portal venous air due to superior mesenteric artery syndrome in an underweight, developmentally normal seventeen-year-old boy. Despite his alarming imaging findings, his abdominal exam was benign and he was successfully treated nonoperatively.

Pneumatosis intestinalis is an imaging finding often associated with bowel necrosis; gastric pneumatosis is rare and alarming when encountered. Etiologies include any pathology that results in increased stomach distension and pressure. We present a case of gastric pneumatosis and portal venous air in a seventeen-year-old boy secondary to superior mesenteric artery syndrome, a rare sequelae of this condition. Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

### 1. Case report

A seventeen-year-old previously healthy male presented to an outside facility with a 12-h history of abdominal pain, nausea, and emesis. At the time, he was afebrile and tachycardic with a distended, tender abdomen. His white blood cell count was 16,400 cells/mm<sup>3</sup> and he was acidotic with a pH of 7.31, a base deficit of 10, and a venous lactate of 2.5 mg/dL. Computed tomography (CT) scan demonstrated massive gastric and proximal duodenal distension with decompression beyond the retroperitoneal sweep as well as a small pneumoperitoneum and significant gastric pneumatosis and portal venous gas (Fig. 1). Additionally noted on the CT scan was a reduced 13° aortomesenteric angle and a narrow 2 mm aortomesenteric distance, resulting in compression of the transverse duodenum compatible with superior mesenteric artery (SMA) syndrome (Fig. 2). A nasogastric (NG) tube was placed yielding high volume bilious output and immediate resolution of his pain and nausea. He was transferred to our facility for emergent

surgical consult.

Upon arrival, he denied any previous episodes and his past medical history was only significant for a seizure disorder well-controlled on topiramate. He had no history of intestinal pathology and denied alcohol or tobacco use, reflux, previous emesis, significant weight loss and any other associated symptoms. His father had ulcerative colitis. On physical exam he remained slightly tachycardic, but was normotensive and afebrile in no acute distress with a completely benign abdominal exam. Notably, his weight was only 44.2 kg making his body mass index (BMI) 16.4, less than 5th percentile for sex and age.

At this point, after review with our radiologists, the working diagnosis was distal duodenal obstruction, due to SMA syndrome leading to tense gastric distension and microperforation with tracking of air in the wall of the stomach. Despite the worrisome CT findings, his clinical exam was so reassuring that we proceeded with nonoperative management. Intravenous (IV) fluid was administered, repeat vitals and abdominal exam were normal, and follow-up labs normalized. Additionally, the NG output turned nonbilious and the patient remained pain-free.

After initial resuscitation, the patient was given total parenteral nutrition (TPN) and an upper gastrointestinal series (UGI) was performed on hospital day 3. This demonstrated resolution of gastric pneumatosis and supported the diagnosis of SMA syndrome demonstrating abrupt delay in passage of contrast with evidence of external compression of the transverse duodenum (Fig. 3). The psychiatric consultant observed no evidence of body dysmorphism, but did note restrictive eating habits and they recommended ongoing outpatient

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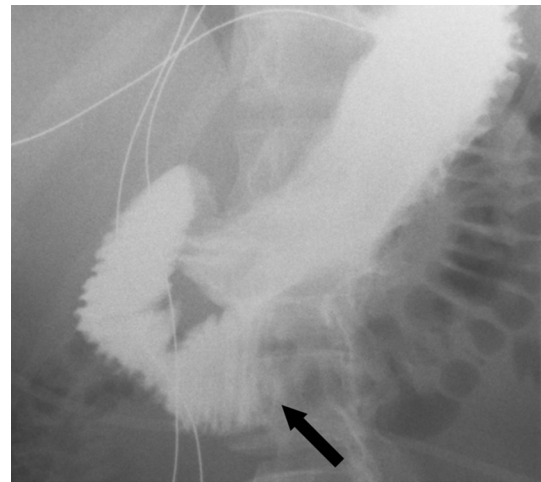
**Fig. 1.** Coronal CT demonstrates significantly dilated stomach and duodenal bulb (asterisk) with portal venous gas (arrowheads), gastric pneumatosis (arrow), and pneumoperitoneum (triangle).

follow-up. A nasojejunal (NJ) tube was placed for feeds. Over the following days, his oral intake gradually increased and his NG tube was removed. He was discharged on hospital day 6 eating a regular diet with cycled NJ tube feeds (45 kcal/kg/day) at night for nutritional supplementation.

At follow-up one week later he had gained 2.5 kg. By one month post-discharge his BMI was low normal at 18.3 and a repeat UGI demonstrated resolution of the duodenal obstruction. His NJ tube was removed and his appetite and weight remained stable at two-week follow-up.

**2. Discussion**

This case represents not only a rare pathology but an unusual presentation of SMA syndrome. SMA syndrome, also known as Wilkie syndrome or cast syndrome, is uncommon, but is important to recognize as a potential cause of proximal gastrointestinal obstruction. Pneumatosis intestinalis is a radiologic sign that is defined as gas within the intestinal wall, typically in the submucosal or subserosal layer, caused by mucosal disruption from ischemia [1]. Etiologies range from life-threatening to benign, but the most worrisome is mesenteric ischemia with intestinal necrosis [2]. Because the stomach has such robust blood supply, gastric pneumatosis is an especially negative



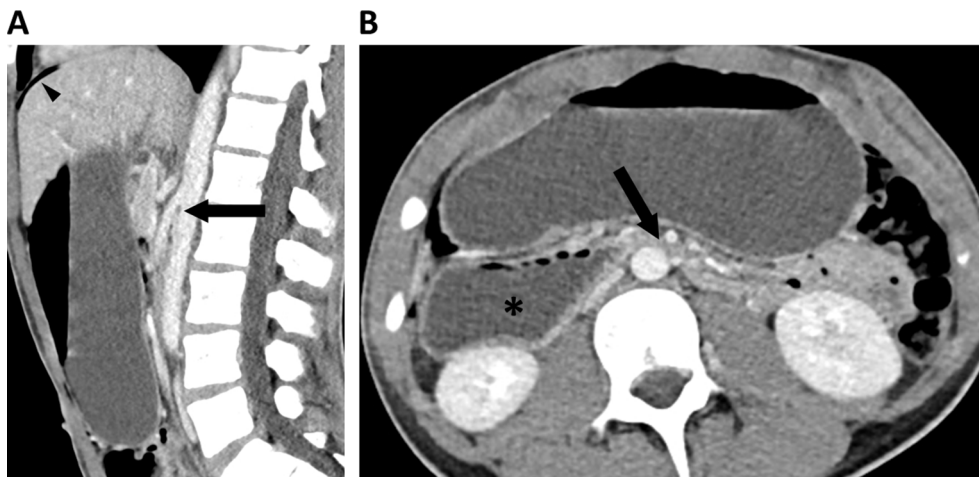
**Fig. 3.** AP fluoroscopic upper GI image demonstrates partial obstruction with abrupt longitudinal extrinsic compression of the transverse duodenum (arrow) followed by a delay in transit of contrast.

finding in the setting of intestinal ischemia.

Rare cases of non-ischemic causes of gastric pneumatosis have been reported. These include duodenal atresia [3], pyloric stenosis [4,5], annular pancreas [6], duodenal stenosis with malrotation [7], and bezoar [8], among others. The mechanism is the same in all cases – intestinal obstruction distal to the stomach that results in distension, increased pressure, and localized ischemia and microperforations of the mucosal lining, allowing air tracking. There are very few case reports of gastric pneumatosis as a presenting sign for SMA syndrome, and most are in adults [9,10].

SMA syndrome is also rare in children, and typically occurs in intellectually or physically disabled children [11], especially following surgery for scoliosis [12]. The most common risk factor present in these patients is critically low BMI, but some children are predisposed to the condition due to a short ligament of Treitz. Weight loss is thought to reduce the visceral fat pad volume between the SMA and the aorta, allowing the SMA to collapse against the aorta causing compression of the duodenum and less commonly the left renal vein. Symptoms are that of a proximal bowel obstruction, including nausea, bilious emesis, early satiety, and weight loss, which initiates a positive feedback loop. Complications of SMA syndrome include electrolyte abnormalities secondary to emesis, gastric perforation, and potentially duodenal bezoar formation [13].

Diagnosis is best achieved by upper GI demonstrating an abrupt halt of contrast at the third portion of the duodenum. CT scan with contrast



**Fig. 2.** A- Sagittal CT demonstrates acute angulation of the superior mesenteric artery with reduced aortomesenteric angle of 13° (arrow), resulting in compression of the transverse duodenum. Distended stomach and pneumoperitoneum (arrowhead) are again noted. B- Axial CT demonstrates dilated stomach and proximal duodenum (asterisk), which tapers posteriorly to the superior mesenteric artery (arrow), where the narrow aortomesenteric distance measures 2 mm (arrow).

may show more detail, as in our case. On cross-sectional imaging, the aortomesenteric angle can be measured, and while the diagnostic cutoff is 25° in adults, one study estimated that up to 20% of asymptomatic children would be incorrectly diagnosed using this value [14]. It is recommended to use the CT findings as an adjunct to the clinical picture for diagnosis rather than to rely on the absolute aortomesenteric angle.

Finally, treatment of SMA syndrome is typically non-operative. After initial resuscitation and correction of any electrolyte aberrations, the principles of therapy are improving nutrition and allowing weight gain to increase the volume of the fat pad between the SMA and the aorta. This is achieved by passing a feeding NJ tube and providing tube feeds until the patient is able to tolerate a diet by mouth. Frequent follow-ups and weight checks are important, as well as psychiatric evaluation to rule out eating disorder. Repeat imaging after the patient has gained weight is useful in determining timing for NJ removal.

Operative approaches are rarely required, and only if non-operative management is unsuccessful [15]. Historically, Strong's procedure was performed, which brings the duodenum into the peritoneal cavity by dividing the ligament of Treitz and mobilizing the duodenum to the right of the SMA [16]. Today, preferred management is by laparoscopic or open duodenojejunostomy with division of the fourth portion of the duodenum to prevent blind-loop syndrome [17].

### 3. Conclusion

Gastric pneumatosis and portal venous gas in a clinically stable patient may be due to duodenal obstruction secondary to SMA syndrome. Non-operative management of SMA syndrome with nutritional supplementation is typically successful and any extraluminal air should reabsorb with resolution of upper gastrointestinal distension.

### Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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### Author contributions

ND/MM drafted manuscript and performed critical revisions of manuscript; FR performed study conception and design and provided critical revisions to the manuscript. All authors have read and approved the final submission.

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