Superior Mesenteric Artery Syndrome

INTRODUCTION

Superior mesenteric artery (SMA) syndrome was first described in 1842 by Rotikansky during an autopsy for duodenum compression. Wilkie in 1921 defined the patho-physiological changes of the third (transverse) portion of the duodenum when obstructed by arteriomesenteric compression and used the term “chronic duodenal ileus”. He published the first comprehensive SMA syndrome case series of 75 patients in 1927. Since then, approximately 500 cases of SMA syndrome have been reported.

SMA syndrome, also known as Wilkie disease, duodenal arterial mesenteric compression, duodenal ileus, aortomesenteric artery compression, and cast syndrome, is an uncommon and sometime life threatening gastrointestinal-vascular disorder. Some studies report the incidence of SMA syndrome to be 0.013-0.3% of the general population. Approximately 0.013-0.78% of routine upper GI barium studies identify SMA compression suggesting this diagnosis. 75% of the patients reported with SMA syndrome are aged 10-30 years and predominantly females. A delay in the diagnosis of SMA syndrome can result in malnutrition, electrolyte imbalance, gastric perforation, pneumatosis and hypovolemia, with a reported mortality rate up to 33%. In the usual clinical setting its nonspecific presentations can result in under-diagnosis and severe outcomes.

The recognition of SMA syndrome as a distinct clinical entity remains controversial because it can be confused with other anatomic or motility-related causes of duodenal obstruction. The left renal vein may also be entrapped and compressed between the aorta and the SMA, a clinical setting referred to as “nutcracker syndrome”. The symptoms of SMA syndrome do not always correlate well with abnormal anatomic findings on radiologic studies, and may not resolve completely after treatment. It was often regarded as a diagnostic dilemma and a diagnosis of exclusion. In this article we will review the pathophysiology, symptoms, diagnosis and treatment of SMA syndrome.

Pathophysiology

Anatomically, the third portion of the duodenum typically crosses caudal to the origin of SMA, passing between the aorta and the superior mesenteric artery at the level of 3rd lumbar vertebral body, and being suspended by its attachment to the ligament of Treitz (Figure 1). SMA normally arises from the anterior aspect of the aorta at the level of the L1 vertebral body, and is enveloped in fatty and lymphatic tissue. It forms a takeoff angle of approximately 45° (normal range 38-65°) from the abdominal aorta with the normal mean aorto-mesenteric distance of 10-28 mm.
Any factor that decreases this takeoff angle and narrows the aorto-mesenteric distance can compress the third part of the duodenum as it passes between the SMA anteriorly and the spine posteriorly, resulting in SMA syndrome. In the case of SMA syndrome, this takeoff angle can be sharply narrowed to approximately 6-25° and the aorto-mesenteric distance can be decreased to 2-8 mm, causing entrapment of the third part of the duodenum and mechanical obstruction at the level of the third and fourth parts of the duodenum (Figure 2).\textsuperscript{6,18,20,22}

Predisposing conditions for SMA syndrome that can change the aorto-mesenteric angle have been categorized into three groups. Group A is a function of weight loss. Significant weight loss leading to loss of the mesenteric fat pad and accompanying decreased body mass index is the most common cause of SMA syndrome. Reduction of the retro-peritoneal fat, which lies between the duodenum and the spine allows the SMA to compress the duodenum against the vertebrae. In the absence of an appropriate fatty support, the angle at which the SMA leaves the aorta promotes more compression of the third portion of the duodenum.\textsuperscript{23,24}

This weight loss can be a consequence of medical, psychological or surgery disorders, malignancy, malabsorption syndromes, human immunodeficiency viral infection, trauma, anorexia, burns, bariatric surgery, diabetes mellitus and eating disorders.\textsuperscript{7,25-30}

Group B is attributed to external causes, such as the corrective spinal surgery for scoliosis with instrumentation or body casting resulting in prolonged post operation recovery. This procedure can displace the origin of the superior mesenteric artery by relative lengthening of the spine, reducing the aorto-mesenteric takeoff angle and decreasing the mesenteric artery’s lateral mobility, which is also termed as “cast syndrome”.\textsuperscript{14,31-33} It has been demonstrated that the incidence of SMA syndrome after surgical procedures for correction of spinal deformities varies between 0.5 and 4.7\%.\textsuperscript{14,34-36}

Group C is a function of intraabdominal anatomy, either congenital or acquired, such as compression or mesenteric tension (e.g. aortic aneurysm), low origin of the superior mesenteric artery, following esophagectomy where gastric pull up into the chest changes the upper GI anatomy and peritoneal adhesions.\textsuperscript{37-39} A congenital short ligament of Treitz may pull the duodenum up towards the insertion of the aorto-mesenteric angle predisposing to SMA and malrotation of the small bowel is another predisposing factor.\textsuperscript{40}

**Figure 1.** Anatomy of Superior Mesenteric Artery, Duodenum, and Aorta. The third part of the duodenum crosses caudal to the origin of SMA, passing between the aorta and the superior mesenteric artery and suspended by its attachment to the ligament of Treitz.

**Figure 2.** Small bowel series with oral contrast could show significant prolonged retention of barium proximal to the third portion of duodenum with distal narrowing at the 4th part, while the jejunum looks normal in caliber.

**Symptoms**

SMA syndrome patients may present acutely or more insidiously with progressive nonspecific symptoms, the severity depending on the degree of duodenal obstruction.\textsuperscript{19} Acute presentations can be related to post-traumatic surgery and often develop from 6-12 days after surgery, and are explained by hyperextension of the SMA compressing the duodenum, sometimes combined
Superior Mesenteric Artery Syndrome

GASTROINTESTINAL MOTILITY AND FUNCTIONAL BOWEL DISORDERS, SERIES #3

with prolonged periods in surgical casts. Another pattern of presentation is in the setting of substantial weight loss that could be related to an eating disorder, limited oral intake or vomiting disorders or cachexia of malignancy or serious depression. A more insidious presentation that may be seen by gastroenterologists involves a long history of abdominal symptoms, where the link to compression of the duodenum is overlooked. Patients with mild obstruction may have only postprandial epigastric pain and early satiety, while those with more advanced obstruction may have severe nausea, postprandial abdominal pain, bilious emesis and weight loss. These symptoms are associated with reduced food intake related to delayed gastric emptying from the retropulsion of food from the duodenal compression with the accompanying compensatory reversed peristalsis. A key aspect of the history that needs to be elicited is that the symptoms are definitely exacerbated by a meal and are better when fasting or overnight. Other aspects of the history include worsening if lying on the right side or supine, and relief by the Hayes maneuver (pressure applied below the umbilicus in cephalad and dorsal direction), lying prone, knee-chest, or left lateral decubitus positioning. These positions of alleviation remove tension from the mesentery and SMA, elevating the root of the SMA and increasing the aorto-mesenteric angle and distance. Findings on physical examination are nonspecific but can include abdominal distension, a succussion splash, and high-pitched bowel sounds.

Because of the nonspecific nature of the symptoms, clinicians need a high degree of suspicion in order to diagnose SMA syndrome. The diagnosis is often delayed and arrived at through the process of excluding other etiologies of intestinal lumen obstruction. The differential diagnosis of SMA syndrome includes other causes of bowel obstruction, duodenal dysmotility and gastroparesis such as diabetic gastroparesis, chronic pancreatitis, systemic autoimmune disease, chronic mesenteric ischemia, idiopathic intestinal pseudo-obstruction, megaduodenum, often caused by connective tissue diseases, especially scleroderma. Post prandial pain of unknown origin, irritable bowel syndrome, dyspepsia, including peptic ulcer and H. pylori gastritis may also be the working diagnosis in patients with nonspecific symptoms (Table 1).

Delay in recognition of SMA syndrome can often result in complications, such as severe electrolyte abnormalities, malnutrition, obstructing duodenal bezoar, gastric dilation with a risk of perforation and pneumatosis, which could portend a fatal outcome. Some patients may be receiving chronic narcotics for pain relief, which will further inhibit duodenal and gastric motility promoting postprandial vomiting.

### Diagnosis

The diagnosis of SMA syndrome is usually established by combinations of abdominal radiographs with barium study, ultrasonography, Computed tomographic (CT) angiography, and magnetic resonance imaging (MRI). Although less high technology and more “old school”, we still recommend an upper GI barium study with a small bowel follow through and Doppler blood flow assessment as the initial diagnostic evaluations of SMA syndrome. Another advantage of abdominal radiographs and ultrasound is the low cost. Their findings may demonstrate a dilated proximal duodenum

---

### Table 1. Differential Diagnosis of Clinical Entities “Mimicking” SMA Syndrome

<table>
<thead>
<tr>
<th>Functional bowel disorders and duodenal, small bowel dysmotility</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diabetic gastroparesis</td>
</tr>
<tr>
<td>Chronic pancreatitis</td>
</tr>
<tr>
<td>Systematic autoimmune disease</td>
</tr>
<tr>
<td>Chronic mesenteric ischemia</td>
</tr>
<tr>
<td>Idiopathic intestinal pseudo-obstruction</td>
</tr>
<tr>
<td>Megaduodenum</td>
</tr>
<tr>
<td>Connective tissue diseases (scleroderma)</td>
</tr>
<tr>
<td>Post prandial pain of unknown origin</td>
</tr>
<tr>
<td>Irritable bowel syndrome</td>
</tr>
<tr>
<td>Dyspepsia</td>
</tr>
<tr>
<td>Peptic ulcer</td>
</tr>
<tr>
<td>H. pylori gastritis</td>
</tr>
</tbody>
</table>

(continued on page 16)
with an abrupt termination of the barium column in the third and fourth part of the duodenum proximal to the ligament of Treitz with a normal jejunal caliber beyond the ligament of Treitz. In addition, this oral contrast study could show significantly prolonged retention of barium proximal to the third portion of duodenum and hold up before entering the jejunum as well as dilation of the proximal duodenum and stomach associated with retrograde flow of contrast from reverse peristalsis (Figure 3). It is key to have an informed radiologist performing the small bowel series and the clinician needs to be involved and interacting to help interpret the findings with the radiologist. Follow up EGD to investigate these radiographic findings can reveal old food in the stomach, duodenal dilation with or without bezoar formation, and obstruction at the end of the third part of duodenum. Conventional arteriography was traditionally performed simultaneously with the barium study to demonstrate the superior mesenteric artery superimposed upon the barium-filled duodenum with the decreased aorto-mesenteric angle. Ultrasound is a noninvasive method to evaluate the mesenteric artery anatomy and measure the aorto-mesenteric angle. Positional maneuvers, such as having patients in the lateral decubitus or even standing, may identify alterations in the aorto-mesenteric angle. Endoscopic ultrasound has recently been effective in demonstrating in more detail and better definition the anatomic abnormalities associated with SMA syndrome. More advanced imaging studies, such as CT and MRI angiography, are often ordered in the setting of patients with variable abdominal pain with nausea and vomiting and unclear diagnosis. These noninvasive images can provide additional anatomic details such as the compressed bowel in relation to vessels, and calculate the aorto-mesenteric angle and the amount of intra-abdominal and retroperitoneal fat tissue. Recently three-dimensional reconstruction has revealed increased blood flow velocity through the SMA and may unexpectedly reveal other possible causes of the abdominal pain, such as abdominal aneurysm (Figure 4). The diagnosis of SMA syndrome should come to mind in patients with clinical features of duodenal obstruction and imaging results revealing duodenal obstruction in the third portion with active retrograde peristalsis. More studies should then be pursued to define if the aorto-mesenteric angle is ≤25° and the aorto-mesenteric distance is ≤8 mm. Other features to note are high fixation of the duodenum by the ligament of Treitz, or abnormally low origin of the superior mesenteric artery or anomalies of the superior mesenteric artery. Patients with SMA syndrome usually have undergone extensive gastrointestinal evaluation and procedures over a period of time, including upper gastrointestinal endoscopy and colonoscopy, to exclude malabsorptive, ulcerative and inflammatory intestinal conditions. These procedures are expensive, have potential risk for patients, and add to the overall economic burden associated with diagnosing this elusive entity. When faced with a diagnostic dilemma of unknown etiology of abdominal pain or nausea and vomiting, the gastroenterologist should take or re-take a thorough history and review the imaging studies keeping in mind some less common entities in the differential diagnosis, such as the possibility of undiagnosed SMA. Treatment Initial conservative treatment with reversal of any precipitating factor is recommended in all patients with superior mesenteric artery syndrome. Initial gastric decompression by nasogastric tube aspiration can reduce the dilated stomach and proximal duodenum and help to monitor fluid balance. Patients with acute SMA syndrome usually have electrolyte imbalances,
which should be monitored and corrected aggressively. Nutritional support and attempts to increase weight are important initial considerations. These approaches may include an enteral nasojejunal feeding tube if it can be successfully passed through the narrowed duodenum, or a laparoscopic placement of a jejunal feeding tube beyond the ligament of Treitz may be necessary for a short period of 3 to 6 months. Total parenteral nutrition is not recommended because of complications. All these measures are aimed at increasing the fat pad between the spine and duodenum.\textsuperscript{49,50}

Patients with suspected eating disorders need professional nutritional and psychiatric evaluation to help achieve optimal calorie replacement.\textsuperscript{5} Promotility medications, such as metoclopramide, are not appropriate since increasing upper GI motility when duodenal obstruction is present increases the pain component. Antiemetics can be supportive. Younger patients in particular with acute SMA syndrome would benefit from conservative treatment, which should be instituted for at least 3 to 6 months.\textsuperscript{5,8}

Specific indications for surgery in patients with chronic SMA syndrome include failed conservative treatment, long-standing symptoms, continuing weight loss due to abdominal pain, nausea and vomiting and reduced food intake, and marked duodenal dilatation with stasis.\textsuperscript{5,40} (Figure 5) Co-management with dieticians and psychiatry consultation when appropriate are recommended to ensure the best outcomes including concerns for adequacy of wound healing after surgery.

The most common surgical operation for SMA syndrome is duodenojejunostomy, in which the compressed portion of the duodenum is actually bypassed by constructing an anastomosis between the 2\textsuperscript{nd} portion of the duodenum and proximal jejunum anterior to the superior mesenteric artery. Duodenojejunostomy can reestablish the bowel continuity with a success rate > 90%.\textsuperscript{16} (Figure 5C, the preferred surgery) Another version of this duodenojejunostomy is division of the 4\textsuperscript{th} portion of the duodenum. (Figure 5D) This is not the recommended best option since there is division of the 4\textsuperscript{th} portion of the duodenum. (Figure 5D) This is not the recommended best option but rather the approach of leaving the duodenum-jejunal continuity intact is the preferred surgery. Surgical complications include bleeding, leakage or stricture at the anatomies site.\textsuperscript{51} Long term concerns include small bowel bacterial overgrowth in the “blind loop” created in the bypassed 3\textsuperscript{rd} and 4\textsuperscript{th} parts of duodenum.

Another surgical option is gastrojejunostomy (bypass the obstruction by bringing up loop of jejunum to the stomach and anastomosis) (Figure 5B). This may be a potential consideration when adhesions from previous surgeries prevent adequate access to create the Duodeno-jejunal anastomosis. Another surgical approach is the Strong’s procedure (duodenal derotation...
with lysis of the ligament of Treitz). However, this procedure is now largely of historic interest and has a higher failure rate to relieve the duodenal obstruction. Successful laparoscopic techniques for the duodenojejunostomy and Strong procedures have been described. Although current literature is limited to case reports and small studies, laparoscopic approaches offer a less invasive surgical option. An important point to address is that all of these surgeries have one thing in common; the SMA itself is never displaced, re-routed or surgically altered. This is a key anatomic feature of the surgeries.

Long term outcomes in SMA syndrome patients after surgery are limited in the literature. One series of 16 patients found significant weight gain, but most symptoms remained unchanged except for decreased vomiting. The need to address eating disorders, bulimia, and underlying psychiatric issues is a key aspect of post-surgery management. A recent series of 8 patients described improved symptoms but no weight gain. Surgical morbidity and mortality can be affected by other comorbidities, e.g. diabetes, and end-stage renal disease.

CONCLUSION

Superior mesenteric artery (SMA) syndrome is an uncommon but well recognized clinical entity characterized by compression of the third portion of the duodenum between the aorta and the superior mesenteric artery. This can result in an acute presentation or more commonly chronic nonspecific symptoms explained by duodenal obstruction with decreased aorto-mesenteric angle and distance. The SMA syndrome has a spectrum of symptoms, which can be referred to as “great mimickers” of a GI motility disturbance. The main GI motility conditions that would be encompassed are: delayed gastric emptying; dilated duodenum suggesting intestinal pseudo-obstruction; unexplained nausea and vomiting and abdominal pain, suggesting the spectrum of cycle vomiting syndrome on one hand, and irritable bowel syndrome on the other hand. This is a great challenge to the physician in practice.

We recommend that clinicians focus on the following “clinical pearls”: 1) unexplained abdominal pain provoked by eating and accompanied by nausea and vomiting; 2) endoscopic evidence of retained food in the stomach and a dilated proximal duodenum; 3) a slow scintigraphic gastric emptying result (Retention of >60% of isotope at 2hrs and >10% at 4hrs). Armed with these clues, the possibility of SMA syndrome has to come to mind, so the next logical step is to get a “road map” and obtain an upper GI and small bowel series. If the clinical suspicion is borne out by a suggestive contrast study then more sophisticated noninvasive imaging can be pursued to demonstrate the specific features we have summarized in this review. SMA syndrome patients need initial conservative treatment where reversible aspects, particularly nutrition, are the focus. Surgical options are effective for non-responding patients, although complete resolution of all symptoms may not always be achieved. The role of explanation, education and practicing the “art of medicine” are all important features in the total care and outcome of these patients. Making a diagnosis in a patient who has been told there is no explanation for the abdominal pain, nausea and vomiting is very satisfying, particularly when there is a treatable and reversible entity involved. Such is the world of SMA syndrome, an often overlooked entity.

References

Superior Mesenteric Artery Syndrome

For further details visit our website: www.practicalgastro.com

Special rates are available for quantities of 100 or more.