A Case Report of Superior Mesenteric Artery Syndrome

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INTRODUCTION
Superior mesenteric artery syndrome is a rare gastrointestinal disorder caused by compression of the third part of the duodenum between the superior mesenteric artery (SMA) and the vertebral body of the spine as first described by Rotikansky et al.1 The syndrome typically results from a combination of the decreased angle of takeoff of the SMA from the aorta with modified blood flow and the degree of the retroperitoneal fat pad, which normally cushions the duodenum from the spine. Acute presentations can be post-traumatic due to hyperextension of the SMA across the duodenum, prolonged periods in body casts, or from dramatic weight loss. The more insidious presentation usually involves a long history of abdominal complaints, where the link to compression of the duodenum is unrecognized. Delay in diagnosis can often result in complications in the form of suspected small intestinal obstructions, severe electrolyte abnormalities, nutritional challenges, and chronic postprandial vomiting, suggesting the diagnosis of gastroparesis. If conservative nutritional support measures fail, surgery to perform a duodenoejunostomy has been shown to have good outcomes.2 Here we describe a case of SMA syndrome in a 19 year old female presenting with chronic, recurrent abdominal pain and cachexia.

CASE HISTORY
A 19-year-old Caucasian female with past medical history of seizures presented to the emergency department complaining of epigastric abdominal, midsternal, and lower back pain for the duration of one week. She described the abdominal pain as constant, sharp, and without radiation. Patient stated abdominal pain interfered with her daily activity and ability to sleep. This pain was worse with eating and accompanied by nausea, vomiting with occasional hematemesis. She also described having similar episodes for the past 5 years. When she had the first episode, she sought medical help without resolution of her symptoms. Patient states that she has to sleep sitting down with her legs crossed, and bending over so she can rest her head on the bed. This was the only position in which she could find some relief of her pain. She decided to seek medical attention this time after she realized she was deteriorating with inability to tolerate food and liquids.

Review of systems was positive for weight loss of approximately 10 lbs. in the last month, generalized weakness, oliguria, and anxiety. Home medications consisted of Xanax, which she had not taken for 2 weeks prior to presentation. Patient had positive cigarette smoking history of ½ pack per day since the age of 12, and illegal drug abuse of marijuana, cocaine, and “spice” (synthetic cannabis) use for an estimated 5 years.

PHYSICAL EXAMINATION
Upon physical examination, the patient was alert, but in moderate distress; crying, irritated, and aggressive. Patient was afbrile with stable vital signs except for a blood pressure of 161/91 mmHg. Exciorations were present on her chin area. Her spine was tender to palpation of lumbar area. Abdominal palpation revealed a soft, non-distended abdomen, without hepatosplenomegaly. Tenderness was elicited by palpation over the epigastric area, and bowel sounds were present. Neurology examination was negative.

LABORATORY TEST RESULTS
Initial lab results showed a white blood cell count of 16.04 x10^9/L, hemoglobin 10.2 g/dL, potassium 5.4 mmol/L, HCO3 18 mmol/L, and blood glucose 132 mg/dL. Urine drug screen was positive for barbiturates and opiates. Beta-HCG was negative. Abdominal x-ray indicated paucity of intestinal gas with no pneumoperitoneum.

MANAGEMENT AND CLINICAL COURSE
Patient was admitted with impression of chronic nausea and vomiting, in addition to abdominal pain of unknown etiology. She was treated supportively with scheduled Zofran for antiemetic control, but her abdominal pain persisted, as did her difficulty tolerating any diet.

Abdominal CT indicated a fluid-filled dilated stomach and proximal duodenum with narrowing of the duodenum in the third portion and a collapsed 4th portion. The aorta mesenteric takeoff distance was 9 mm from the aorta. Aorta mesenteric angle was measured as approximately 20 degrees (Figure 1). Barium contrast exam found that the 2nd part of duodenum was dilated with narrowing of the 3rd and 4th part with a normal appearing esophagus, pyloric channel, and a normal small bowel beyond the ligament of Treitz. Stomach filled readily with slight delay in gastric emptying due to pylorospasm (Figure 2). Upper GI endoscopy was conducted with the impression of dilatation of the 2nd and 3rd part of duodenum with noted extrinsic moderate compression at the 4th part of the duodenum, consistent with

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The patient repeatedly threatened to sign herself out of the hospital against medical advice, despite having a life-threatening condition. Given the patient’s prior history of violence and substance abuse, it was determined to be in her best interest to be placed on psychiatric hold.

Following surgical consultation and discussion with the patient, she underwent laparotomy with a duodenoejunostomy for alleviation of her symptoms. Upper GI endoscopy was performed immediately following the procedure to visualize the anastomosis. There was evidence of a widely patent duodenoejuntostomy at the 2nd part of the duodenum, which was characterized by a healthy appearing mucosa. A full thickness antral excisional biopsy was done during the laparotomy and was sent for pathology analysis. No significant atrophy, inflammation, or necrosis was found. C-KIT immunostaining identified an average of eight interstitial cells of Cajal per high-power field (HPF), which is below the normal range of 10-25 per HPF. Patient returned to the medicine service for ongoing care after the laparotomy. She was noted to be constipated with a substantial amount of stool in the colon. Efforts to relieve patient’s constipation were unsuccessful, as patient refused stool softeners and an enema. On postoperative day 4, patient developed coffee-ground emesis and tachycardia. She was found to have a 3 gm drop in her hemoglobin. She received 2 units of PRBC and was transferred to the ICU. Endoscopy in the ICU indicated evidence of recent bleeding with brown blood in the stomach and no active upper GI bleeding. The source of bleeding was at the duodeno-jejunal anastomosis with a localized red clot. Patient was stable and transferred back to the floor for ongoing care. She recovered without further bleeding and began tolerating a liquid then mechanical soft diet. Her abdominal pain did slowly decrease compared to the time of hospitalization. Vicodin (codeine/acetaminophen) was prescribed at discharge for breakthrough pain.

DISCUSSION
In the Superior Mesenteric Artery (SMA) syndrome, the 3rd portion of the duodenum is compressed in the angle between the SMA anteriorly and the spine posteriorly, causing mechanical obstruction at the level of the 3rd and 4th portions of the duodenum.5,4 This uncommon and sometimes life threatening condition of intestinal obstruction was first described by Carl Von Rokitansky in 1842.5 Wilkie provided a patho-physiological description of the condition he named “chronic duodenal ileus” in 1921, and later reported this pathology in the literature in 1927.6

SMA syndrome has been described under various other names, including Wilkie disease, duodenal arterial mesenteric compression, duodenal ileus, aortomesenteric artery compression, and cast syndrome.4,6,8 Anatomically, SMA normally arises from the abdominal aorta at a takeoff angle of 25-60 degrees, and the mean aortomesenteric distance is 10-28 mm.8,9 In the case of this syndrome, this takeoff angle is reduced to less than 25 degrees of the angle and the aortomesenteric aperture is usually between 2-8mm.8,9

Predisposing conditions for SMA syndrome have been described. These conditions can change the aortomesenteric angle, and can be categorized into groups. Group A is a function of weight loss.

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Reduction of retro-peritoneal fat, which lies between the duodenum and the spine allow the SMA to compress the duodenum against the vertebrae. This weight loss can be the result of cancer, trauma, eating disorders, rheumatoid disease, cardiac cachexia, human immunodeficiency virus infection, bariatric surgery, and diabetes mellitus. In the absence of an appropriate fatty support, the angle at which the SMA leaves the aorta promotes more compression of the 3rd portion of the duodenum. Group B is attributed external causes, e.g. scoliosis surgery with instrumentation or body casting resulting in prolonged post operative recovery, requiring supine position. 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. Group C is a function of intra-abdominal anatomy, such as compression or mesenteric tension (e.g. aortic aneurysm). A congenital short ligament of Treitz may pull the duodenum up into the root of the aortomesenteric angle and predispose to SMA or malfunction of the small bowel. Our patient also had a decrease in the average number of interstitial cells of Cajal, which is seen in a gastroparesis setting and suggestive of gastric emptying that may have been affected by the dilated duodenum related to a retrograde pressure gradient from the SMA compression.

The incidence of SMA syndrome is between 0.013% and 0.3% in the general population. Clinicians need a high degree of suspicion in order to diagnose this entity. The diagnosis is often delayed and made through the process of excluding other etiologies of intestinal lumen obstruction. Symptoms of SMA syndrome are nonspecific and include intermittent abdominal pain, nausea, bilious vomiting, constipation, early satiety, and postprandial bloating. The entity symptoms are exacerbated by a meal and reduced by not eating. The pain is characteristically relieved by prone, knee- chest, or left lateral decubitus positioning. Patients may complain of chronic abdominal pain, the severity depending on the degree of obstruction, and this may continue for months or even years depending on diagnostic outcomes. Because of this delay in recognition, some patients may be receiving chronic narcotics for pain relief, as was the case in our patient.

Diagnosis is corroborated by combinations of CT, CT angiography, and magnetic resonance imaging (MRI) studies. A barium study is commonly performed as the initial investigation. Barium studies are important for the diagnosis. A classic picture obtained is a dilated proximal duodenum with an abrupt termination of the barium column in the 3rd and 4th part of the duodenum proximal to the ligament of Treitz with a normal jejunum beyond the ligament of Treitz. Recently, multiple imaging studies have been used such as CT angiography and three-dimensional reconstruction. CT and contrast-enhanced CT as imaging modalities allow direct visualization of the compressed bowel in relation to vessels, calculation of the angle between the aorta and SMA, appreciation of the amount of retroperitoneal fat tissue, in addition to having the capacity to reveal other possible causes of the abdominal pain that may not have been appreciated, such as an aneurysm.

Initial treatment of SMA syndrome is conservative and involves gastric decompression by nasogastric aspiration, correction of fluid and electrolyte imbalances, and attempts to increase weight if weight loss is suspected as an underlying etiology of the syndrome. Nutritional support may include a short period of total parenteral nutrition. Most of the patients with chronic SMA syndrome require surgical intervention. Specific indications for surgery include: failed conservative treatment, longstanding symptoms, progressive weight loss, and duodenal dilatation with stasis. Duodenojjunostomy, gastrojejunostomy, and Strong’s procedure (lysis of the ligament of Treitz) are the surgical options described in the treatment of SMA syndrome. Duodenojjunostomy is the most effective of the procedures, with a success rate > 90%, as this was utilized in our patient.

REFERENCE

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