Superior Mesenteric Artery Syndrome: A Rare and Unusual Cause of Gastrointestinal Obstruction

Emily Mae Yap, M.D.*; Ana Beatriz Medrano, M.D.**; Ira Inductivo-Yu, M.D.***

Abstract

Introduction: Superior mesenteric artery (SMA) syndrome is a rare and unusual acquired cause of functional duodenal obstruction whose diagnosis can be easily missed without knowledge of this condition.

Case Presentation: We report a case of a 27-year-old female, presenting with post-prandial vomiting, early satiety, bloatedness and weight loss for about 10 months. Vital signs were stable. She was grossly underweight with a BMI of 11.72 kg/m^2 (height=1.6m, weight=30kg). Physical examination was unremarkable. Gastrointestinal series revealed a narrowing in the third portion of the duodenum likely secondary to extrinsic compression. Contrast-enhanced CT scan of the whole abdomen was performed with 3D reconstruction. There were no definite signs of gastrointestinal obstruction. However, a narrow/acute aorto-mesenteric angle of 13 degrees compressing the third part of the duodenum was noted. Superior mesenteric syndrome was considered, prompting further work-up. Primary hyperthyroidism was the root cause of the patient's weight loss that lead to this condition. Patient was given nutritional support, parenterally and enterally. She was discharged improved after oral feeding was tolerated and patient started to gain weight.

Discussion: Superior mesenteric artery (SMA) syndrome is an uncommon medical condition brought about by a decrease in the aortomesenteric angle from the usual 45° to less than 15° resulting in vascular compression of the third part of the duodenum leading to gastrointestinal obstruction. A high index of suspicion is needed to prevent the diagnosis from being missed which may in turn lead to unnecessary testing and treatment. If recognized early, the condition may be managed conservatively. Surgical management is only required when conservative methods fail.

Conclusion: Early recognition and a thorough evaluation is therefore imperative so conservative measures can be maximized at the outset.

Keywords: superior mesenteric artery syndrome, gastrointestinal obstruction

Introduction

Superior mesenteric artery (SMA) syndrome is an extremely rare cause of functional intestinal obstruction caused by compression of the third part of the duodenum in the angle between the aorta and the SMA.[1] It has an incidence ranging between 0.013 to 0.3% taken from studies on upper gastrointestinal tract series.[2] Although regarded as controversial in the past[3], it is now a clearly defined syndrome associated with a wide range of predisposing conditions.[1] Marked or rapid weight loss with subsequent loss of mesenteric and retroperitoneal fat as may occur in severe wasting, cancer, burns, severe injuries such as head trauma, dietary disorders such as anorexia nervosa or malabsorption, post-operative states, and deformity or trauma to the spine predispose one to have this condition[3,5]. Normally, the SMA leaves the aorta at an acute downward angle. The third part of the duodenum passes through this vascular angle. The mass of fat and lymphatic tissue around the SMA's origin provides adequate protection to the duodenum against compression. A decrease in the angle between the SMA and the aorta from the usual 45° to 15° results in occlusion of the third part of the duodenum causing symptoms of obstruction such as post-prandial pain, early satiety, and vomiting.[6] Without a heightened index of suspicion, this clinical entity can potentially be missed leading to unnecessary diagnostic modalities and treatment. If recognized early, the condition may be managed conservatively. Surgical management is only required when conservative methods fail.[3] Identification of this syndrome can be a diagnostic dilemma and is frequently made by exclusion.[6]

The aim of this case report is to document the first admitted case of SMA syndrome in our institution highlighting

*Diplomate in Internal Medicine, Fellow-in-training Adult Cardiology, Philippine Heart Center, Manila
**Adult Cardiology - Non-Invasive, Philippine Heart Center, Manila
*** Gastroenterology - Transplant Hepatology, Philippine Heart Center, Manila

Corresponding author: Emily Mae Yap, M.D., Philippine Heart Center, Quezon City, Manila, Philippines
Email:emilyyap@gmail.com

the paucity of this entity. This paper is geared towards increasing awareness of this rare and acquired cause of functional gastrointestinal obstruction.

Case Presentation

A 27-year-old female sought admission in our institution due to post-prandial vomiting, bloatedness, early satiety and weight loss for 10 months and back pain for six months. She denied abdominal pain but complained of constipation, with bowel movement usually occurring only once a week. She lost 10kg during a six-month period. She has no known comorbid or psychosocial illnesses and has an unremarkable family history. She was previously admitted in two other hospitals due to persistence of symptoms and underwent extensive evaluation. Electrolytes were deranged secondary to frequent vomiting and were corrected intravenously and with oral supplementation. Upper abdominal ultrasound was unremarkable. Esophagogastroduodenoscopy revealed reflux esophagitis and minimal erosive gastritis in the antrum. The thyroid function test was consistent with primary hyperthyroidism. TSH was low (0.277, normal range: 0.3-5 mIU/L) while FT4 was high (29.69, normal range: 11-22.5 ng/dL). Ultrasound of the thyroid gland was unremarkable. Methimazole 15mg tab every eight hours was started to inhibit synthesis of thyroid hormones. Since there was no improvement in symptoms, patient opted to be discharged against medical advice. She then transferred to our institution for further work-up and management.

On admission, vital signs were within the normal limits. She was grossly underweight with a BMI of 11.72 kg/m^2. Physical examination was unremarkable. Complete blood count was normal (WBC 8.90, Neutrophils 58%, Lymphocytes 29%, Hemoglobin 133 g/L, Hematocrits 41%, Platelet count 338). Creatinine 0.05 mmol/L was normal. Electrolytes were in the lower limits of normal - Potassium 3.6 mmol/L (NR 3.5-4.9), Sodium 137 mmol/L (NR 138-146), Magnesium 0.82 mmol/L (0.74-0.99 mmol/L) and Calcium 1.16 mmol/L (NR 1.16-1.32). Serum albumin was slightly low 43.1 (35-50.00 mmol/L). Alanine Amintotransferase 25 U/L (NR 9-52), Amylase 48 U/L (NR 30-110); Total bilirubin 3.3 umol/L (3.0-22.0) were normal. Chest x-ray showed a narrowed anterior-posterior diameter of the thorax. A review of the upper gastrointestinal series previously done revealed a narrowing in the third portion of the duodenum likely secondary to extrinsic compression. Contrast-enhanced CT scan of the whole abdomen done on her previous admission was also reviewed with 3D reconstruction. There were no definite signs of gastrointestinal obstruction but a narrow/acute aorto-mesenteric angle of 13 degrees compressing the third part of the duodenum was noted as seen in Figure 1 and 2. Chest CT scan was done. Aortic aneurysm and dissection were ruled out. Dual Energy X-ray Absorptiometry (DXA) was done. Whole body composition showed the following: total

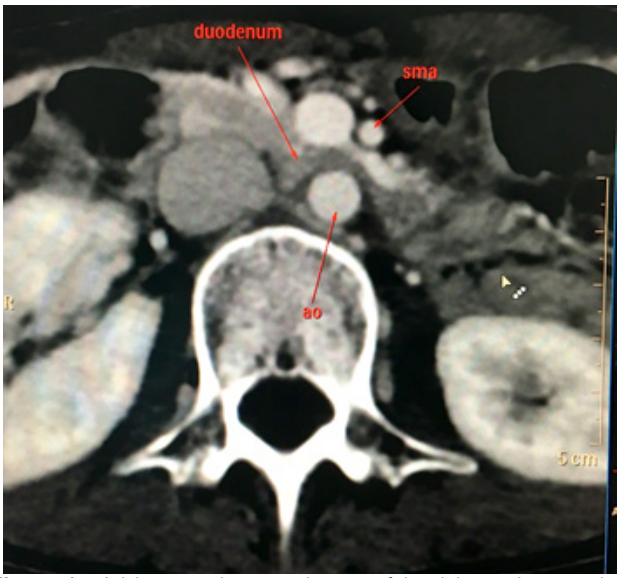

Figure 1. Axial computed tomography scan of the abdomen demonstrating duodenal compression by the aorta (ao) and superior mesenteric artery (sma)

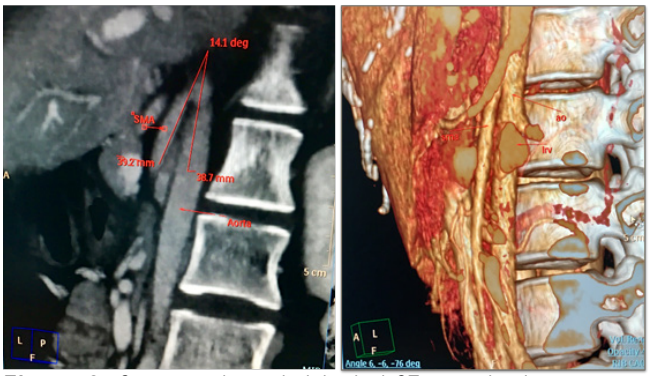

Figure 2. Contrast-enhanced abdominal CT scan showing a narrow aortomesenteric angle of 14.1o on the left and a 3D-reconstructed image on the right

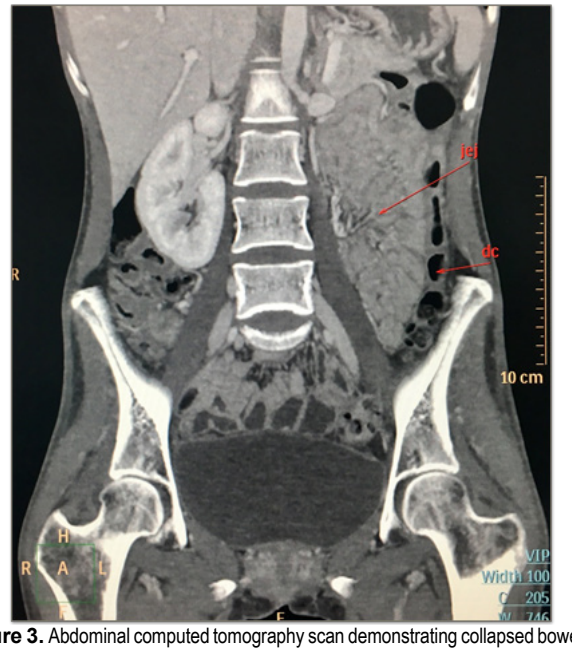

Figure 3. Abdominal computed tomography scan demonstrating collapsed bowels distal to the obstruction in this view (jej - jejunum, dc - distal colon)

body mass of 28.04kg, bone mineral content (percentage) of 1.52kg (5.4%), total lean mass (percentage) of 20.30kg (72.4%) and total fat mass (percentage) of 6.22kg (22.2%). Based on z-scores, the patient was classified as having bone mineral density values below the expected range for age. Superior mesenteric artery syndrome brought about by the patient's progressive weight loss secondary to untreated primary hyperthyroidism was considered. Intravenous fluids (Lactated Ringer's solution) at one ml/kg/hour were started. Electrolytes were corrected accordingly. Nutritional build-up was advised. Dibencozide three milligrams tablet, one tablet, once a day as appetite stimulant and multivitamins capsule one capsule once a day were given. Supplemental parenteral nutrition with 1,000 kcal/day was provided. Small and frequent feedings (six to eight times aday) were advised. A duodenojejunostomy was recommended to bypass the obstruction if oral alimentation would fail. Conservative management was maximized. Patient started to gain half pound a week. She was discharged after three weeks. Out-patient follow up was done two weeks after discharge. She continued to gain weight and reported marked relief of symptoms.

Discussion

Superior mesenteric artery (SMA) syndrome is a rare medical condition brought about by a low aortomesenteric angle resulting in vascular compression of the third part of the duodenum which is relatively immobile.[3] This may present acutely, or with a chronic, insidious history. Acutely, patients may present with signs and symptoms of duodenal obstruction such as nausea, vomiting, abdominal pain or distention, aggravated by eating. Electrolyte imbalance may occur. Chronic cases present many times over many years for investigation of abdominal pain, vomiting, early satiety and anorexia.[2]

Superior mesenteric artery (SMA) syndrome is more common among females and in those with slender built after acute weight loss[3-4] similar to our case. Symptoms are similar to those of small bowel obstruction such as nausea, vomiting, early satiety, anorexia and abdominal pain as presented in our case. Radiologic studies that can be used to establish or confirm the diagnosis include upper gastrointestinal series, ultrasonography, computed tomography scan or CT angiography and magnetic resonance (MR) angiography. In particular, an aortomesenteric angle of <22-25° or an aortomesenteric distance of <8mm support the diagnosis.[3] Unfortunately, this is not routinely measured when one requests for ultrasonography, hence this can be missed on routine exams, highlighting the need to measure the said angle/distance when one is considering the possibility of SMA syndrome. CT or MR scans can demonstrates not only the aortomesenteric angle and distance, but also the presence of surrounding fat, duodenal obstruction and potential culprit (eg. mass). In our case, contrast enhanced CT scan confirmed the suspicion of SMA syndrome as seen in Figures 1 to 3.

Most of the time, the first line of treatment of SMA syndrome includes conservative measures, such as provision of high caloric enteral or parenteral nutrition, and replacement of fluid and electrolytes.[3-5] Those who respond to conservative management have good prognosis. Reversing or removing the precipitating factor[3-4], such as treating the hyperthyroid state in our patient is equally important. Insertion of a nasogastric tube as what was done to our case for gastric and duodenal decompression, and later, for feeding is commonly done. Correction of fluids and electrolytes, as well as an increase in caloric intake which can increase the amount of retroperitoneal fat can subsequently increase the aortomesenteric angle and can alleviate or prevent recurrence of symptoms. In other words, weight gain is usually accompanied by the complete relief of symptoms.[5] Metoclopramide may benefit as a prokinetic.[4] If these measures fail, surgery is then indicated to bypass the compressed duodenum. The procedures that can be done include gastrojejunostomy, duodenojejunostomy (most common) or duodenal mobilization for lowering the duodenojejunal flexure.[3,4]

Conclusion

Superior mesenteric artery (SMA) syndrome is rare. Without a high index of suspicion, the diagnosis can be missed and can lead to unnecessary testing and treatment. Early recognition and a thorough evaluation is therefore imperative so conservative measures geared towards nutritional build up can be maximized. If this fails, surgical management is then considered. The importance of knowing this rare entity should be underscored to heighten awareness.

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