Case Report

Superior mesenteric artery syndrome in a malnourished female: A rare cause of abdominal pain

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\textbf{Abstract}

Superior mesenteric artery (SMA) syndrome is a rare cause of duodenal obstruction that can go undiagnosed, exacerbating weight loss in an already significantly malnourished patient. Diagnosis is often challenging, however, can be made by keeping a high index of suspicion based on the clinical presentation. The pathology involves a reduction in the amount of fat pad between the abdominal aorta and superior mesenteric artery (SMA) leading to a reduction of aorto-mesenteric angle and consequent compression of mostly third part of the duodenum. Management is usually conservative, however, if conservative treatment fails, surgical intervention is warranted.

Our patient was a 20-year-old female who presented to us with nausea, vomiting, weight loss, and abdominal pain. The presence of obstructive symptoms along with imaging (CT scan) lead to the diagnosis of SMA syndrome and she improved with conservative management. Informed consent was obtained for this study.

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\textbf{Introduction}

Superior mesenteric artery (SMA) syndrome (also known as Wilkie’s Syndrome) was first described in 1861 by Rokitansky \[1,2\]. The incidence of SMA syndrome in the general population is 0.013%-3.0% with a mortality rate of 33% \[3\]. The patient often presents with nausea, painful postprandial epigastric fullness, abdominal pain, and bilious vomiting. Significant weight loss is the most common cause of SMA syndrome. Ef-
forts should be made to rule out other causes of mega duodenum and duodenal obstruction. CT imaging should be used to aid with a diagnosis along with the clinical presentation and Oesophagastroduodenoscopy (OGD) should be considered to investigate for underlying weight loss. Initially, a conservative approach should be adapted to treat patients. If conservative treatment fails, this would warrant surgical intervention.

Case presentation

A 20-year-old female presented to ED with nausea, vomiting, weight loss, and abdominal pain. She also complained of constipation on and off during this period. Previous hospital history of note is admission with weight loss and investigations did not reveal any organic pathology and patient was diagnosed and managed for anorexia nervosa.

On arrival at our department, her vital signs were; pulse 110/min, blood pressure 80/60 mm Hg, and temperature 98 F. On physical examination, she was very cachectic with a BMI of 11.2. Abdominal examination revealed a slightly distended abdomen with mild tenderness in the epigastric region and succussion splash but no palpable masses or clinical signs of peritonitis. Bowel sounds were present. WCC was 12,000/mm$^3$. While the rest of the blood and urine examinations were normal. A computed tomography (CT) scan of the abdomen and pelvis with oral and IV contrast material was advised which showed a distended stomach with dilated duodenum up to the 3rd part with collapsed 4th part of the duodenum but no obvious mass demonstrated. Reduction in retroperitoneal and prevertebral fat and aorto-mesenteric angle were also noted hence raising the suspicion of SMA syndrome (Fig. 1). The patient was admitted for inpatient treatment and was started on IV hydration and gastric decompression via nasogastric tube (Ryle’s tube).

OGD showed dilated stomach and duodenum but no obvious mass was seen. Biopsy for the second part of the duodenum showed chronic inflammatory changes but no evidence of celiac disease or any other pathology. The patient was diagnosed with SMA syndrome based on clinical presentation and findings of the investigations. NJ feeding was declined by the patient based on previous experience and hence total parental nutrition was initiated with dietician input and after conservative management after a month of inpatient hospital stay, her symptoms improved.

Her BMI improved from 11.2 to 13.1. She was discharged on nutritional supplements and dietary modifications with an intake of fatty and protein meals in small quantities. She was followed up after 2 weeks and had no symptoms anymore, furthermore, she continued to gain weight.

Discussion

Superior mesenteric artery (SMA) syndrome also called Wilkie’s syndrome is a rare vascular disorder, caused by extrinsic compression of the third part of the duodenum between the origin of SMA and the aorta [3]. This can lead to chronic, intermittent, or acute complete or partial obstruction of the duodenum. It is a rare cause of vomiting, abdominal pain, weight loss, and food intolerance. Normal aortomesenteric angle is 25-60° and aortomesenteric distance is 10-28 mm. Numerous medical and psychiatric conditions contribute to the early rapid weight loss which results in decreasing the aortomesenteric angle and distance [4]. Reduction in aortomesenteric angle to approximately 6-25° and aortomesenteric distance to 2-8 mm would cause SMA syndrome. Merrett et al. described that there can be many causes due to which SMA can be left undiagnosed which includes the nonspecific symptoms, additionally young age often leads to a delay in diagnosis [4]. Underlying causes can be divided into congenital or acquired. The congenital low position of the SMA, the narrow superior mesenteric artery-aorta distance, and intestinal malrotation can lead to SMA syndrome [2,5,6]. Acquired causes of Superior mesenteric artery (SMA) syndrome comprise malabsorption, anorexia nervosa, malignancy, burn injuries, and recent upper abdominal gastrointestinal surgeries [3]. Patients who have severe neurological defects, like cerebral palsy, may increasingly hyperextend their spine, thus being at risk of having a reduced distance between the aorta and the SMA [3]. Oguz et al. stated that patient post scoliosis surgery is prone to the increased tautness of the mesentery leading to SMA syndrome [6].

In addition to clinical symptoms, performing a full preoperative inspection; contrast imaging, upper gastrointestinal...
endoscopy, and cross-sectional angiography are suggested for both confirmation and early diagnosis. We believe that assessment by a vascular surgeon and radiologist together can help report vascular imaging essential in determining the diagnosis [7]. These examinations can demonstrate the following findings: greater delay of the gastroduodenjejunal transit time, dilatation of the proximal part of the duodenum, and failure of a contrast passaging beyond the compressed part of the duodenum [6].

Initial treatment is usually conservative, nonoperative medical management. Acute setting management involves fluid resuscitation for electrolyte correction, nasogastric tube insertion, gastric decompression, and enteral or total parenteral nutrition. It is recommended to eat small quantities and participate in posture therapy (i.e., knee-to-chest position, prone, left lateral position, or Goldthwaite maneuver) after eating can lead to improvement in symptoms. Dietary support hyperlinking with conventional therapy is important to increase the mesenteric fat pad, thereby increasing the angle and improving the symptoms. Many patients fail conventional treatment and eventually require surgical involvement [8].

Numerous surgical procedures have been considered like duodenojunostomy and gastrojejunostomy. Traditional open bypass surgery was the standard care of treatment until the first successful laparoscopic duodenojunostomy was performed in 1998. This technique has since surpassed open bypass and is the most commonly recommended surgical treatment [9].

**Conclusion**

SMA syndrome is a rare cause of duodenal obstruction and can provide a real challenge to diagnosis. A high index of suspicion should be kept in mind to diagnose it based on clinical presentation. While an active effort should be made to rule out other causes of weight loss and duodenal obstruction, CT imaging is a useful investigation, which could help in diagnosis while OGD should be considered to rule out other mechanical causes of duodenal obstruction and as part of work up to investigate weight loss.

**Human subjects**

Consent was obtained by all participants in this study.

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**Patient consent**

Informed and written consent was obtained by the patient for publication of this study.

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