A Rare Case of Gastric Mantle Cell Lymphoma causing Superior Mesenteric Artery Syndrome in an Adult

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Abstract

Keywords

- artery
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Superior mesenteric artery (SMA) syndrome or Wilkie syndrome is a rare condition that arises when the distal third of the duodenum becomes trapped between the SMA and the abdominal aorta causing intestinal obstruction. It is most commonly described in adolescents and rarely in adult patients. We present an interesting case of an adult who developed SMA syndrome from an uncommon gastric mantle cell lymphoma that was recognized early and treated surgically.

Superior mesenteric artery (SMA) syndrome or Wilkie syndrome is a rare condition that arises when the distal third of the duodenum becomes trapped between the SMA and the abdominal aorta causing intestinal obstruction. It is most commonly described in adolescents and rarely in adult patients. We present an interesting case of an adult who developed SMA syndrome from an uncommon gastric mantle cell lymphoma (MCL) that was recognized early and treated surgically.

Case Presentation

A 60-year-old male presented to the emergency department with a 1-day history of abdominal pain, distension, nausea, and vomiting. He had an endoscopy and colonoscopy a day earlier before his presentation as part of his weight loss workup. He had unintentionally lost 40 lb over a period of 5 months and was complaining of intermittent episodes of vomiting after eating food for the last few weeks. His history was significant for epilepsy and skin melanoma removal few years ago. He gave no history of peptic ulcer disease. On arrival to the emergency room, the patient's physical examination revealed a distended abdomen with mild, diffuse abdominal tenderness. He had a white blood cell count of

8,700, hemoglobin of 11.4, and lactate of 1.8 with a positive fecal occult blood test.

In the emergency department, he underwent an X-ray of the abdomen (➤**Fig. 1**) showing a huge gastric bubble necessitating placement of a nasogastric tube for relief of his symptoms. His computed tomography (CT) scan of the abdomen revealed a massively dilated and thick stomach along with dilated first and second portions of the duodenum with collapsed distal bowel (➤**Fig. 2**). The CT scan interestingly also showed a narrowed aortomesenteric angle with SMA impinging on the third portion of the duodenum (➤**Figs. 3 and 4**). His previous endoscopy showed hypertrophic gastric mucosa with a dilated stomach and duodenum with questionable signs of extrinsic pressure on the third part of the duodenum, all consistent with a diagnosis of SMA syndrome. The gastric biopsy from endoscopy was highly suspicious for lymphoma.

With poor nutritional status, weight loss, and inability to tolerate any oral foods or fluids, the plan was to improve his nutritional status so that he could start chemotherapy. Starting him on parenteral nutrition was an option but that would have come with its own inherent risks and would not have improved his symptoms. Surgical placement of a feeding jejunostomy with gastric drainage would have helped but would require surgery and the resulting quality of life is not

Fig. 1 Abdominal X-ray showing a huge astric bubble.

what patient expected to live with. Feeding through a nasoenteric tube past ligament of Treitz seemed a good choice but would require a long hospital stay with decreased quality of life and sending him home with a nasoenteric feeding tube with continued symptoms was not feasible. We then decided that the best course of action was to fix his condition surgically and performed a duodenojejunostomy, where the second part of the duodenum was anastomosed to a segment of jejunum to bypass the obstructed third portion of duodenum. He tolerated this procedure very well.

He was started on clears on postoperative day 2 and on postoperative day 3 he tolerated a regular diet with less frequent episodes of vomiting. Before being discharged home, he also underwent placement of an infusaport for administration of chemotherapy. Subsequent to the surgery, he slowly started gaining weight and within a month with

Fig. 2 CT scan showing dilated stomach and duodenum. CT, computed tomography.

Fig. 3 CT scan showing (a) SMA coming off aorta at a narrow angle. CT, computed tomography; SMA, superior mesenteric artery.

Fig. 4 CT scan showing dilated (a) second part of duodenum being compressed by (b) SMA coming off (c) aorta. CT, computed tomography; SMA, superior mesenteric artery.

improved nutritional status was started on chemotherapy. During his follow-up visit, he gained back around 20 lb with resolved symptoms and was tolerating diet. His follow-up CT scan showed a remarkable improvement in gastric dilation (►Fig. 5).

Discussion

SMA syndrome comprises a constellation of intestinal symptoms that arise from compression of the third part of the duodenum by the superior mesenteric artery against the aorta. The third part of the duodenum, suspended by the ligament of Treitz, is anatomically positioned between the SMA and abdominal aorta, such that any reduction of aortomesenteric angle leads to mechanical compression of the duodenum causing intestinal obstruction (►Fig. 6). It was

Fig. 5 Follow-up CT scan showing resolution of gastric dilation. CT, computed tomography.

Fig. 6 (Left) The normal anatomy. (Right) Duodenum being compressed by SMA from loss of fat pad with acute SMA-aortic angle. SMA, superior mesenteric artery.

first described by von Rokitsanski in 1861 and later Wilkie provided a more detailed clinical and pathophysiologic description in a series of 64 patients.¹

SMA syndrome is very rare and its incidence according to some studies is reported as between 0.20 and 0.78%.^{2,3} It is found more commonly among older children and adolescents⁴ and is thought to be as a result of rapid growth in length without simultaneous weight gain.^{5,6}

The SMA originates at the level of first lumbar vertebra behind the neck of pancreas and creates an acute angle with the aorta (–Fig. 6). The normal angle between the aorta and SMA ranges from 25 to 60 degrees and the normal aortomesenteric distance is between 10 and 28 mm.^{3,7} Normally, fat and lymphatic tissues around the SMA provide protection to the duodenum against compression.⁸ In SMA syndrome, both these parameters are reduced, with values of 6 to 15 degrees and 2 to 8 mm.⁹ Our patient's aortomesenteric angle and distance were 10 degrees and 6 mm, respectively.

SMA syndrome can result due to abnormal congenital anatomical factors or can be acquired. Some anatomical factors described are hyperextension of the spine with

increased lumbar lordosis and prolonged supine position as seen in quadriplegic patients.⁶ Congenital factors include short intestinal mesentery, anomalous SMA origin, hypermobility of right colon, malrotation of intestines, low take off of SMA, or an abnormally high origin of ligament of Treitz pulling the duodenum toward the root of the mesentery.^{7,10} No genetic causes have so far been reported.

Acquired factor could be any condition that results in weight loss causing reduction in retroperitoneal fat that under normal circumstances serves as a cushion holding the SMA off the spine and protecting the duodenum from compression.^{6,11} This syndrome has been described following cancer, burns, and bariatric surgeries.¹² It has also been reported after surgeries such as restorative proctocolectomy with ileoanal anastomosis that decreases the aortomesenteric angle owing to the tension placed on the small bowel when stretched to the anal anastomosis.¹³ It has also been reported following scoliosis surgery as a result of a relative lengthening of the spine postoperatively.⁸

The symptoms of SMA syndrome are those of proximal intestinal obstruction. They can also present with nonspecific symptoms such as intermittent abdominal pain, nausea, bilious vomiting, early satiety, and postprandial bloating.¹⁴ As a result, the diagnosis of SMA syndrome is often delayed and made through a process of exclusion. The symptoms are usually aggravated by meals and relieved by the patient lying in a knee-chest or prone position as the SMA falls off the duodenum resulting in temporary release of the obstruction.^{15,16}

Diagnosis is based on clinical suspicion and is confirmed with radiological evidence of obstruction. Plain radiographs usually reveal presence of dilated, fluid-filled or gas-filled stomach and duodenum indicating duodenal obstruction.¹⁰ Upper gastrointestinal (GI) contrast study shows duodenal dilatation, retention of barium in the duodenum, and a sharp vertical cutoff from extrinsic pressure at the third portion of the duodenum with collapsed small bowel distal to SMA.^{3,17} If contrast flows in to the jejunum when patient is placed in a left lateral position it clinches the diagnosis.¹² Endoscopic examination may sometimes visualize a pulsatile extrinsic compression suggestive of this condition.¹⁸ However, currently CT scan and magnetic resonance angiography are considered to be the best studies as not only they provide greater anatomic details but also can show other pathology. Several authors therefore report that the diagnosis of SMA syndrome is best confirmed using CT angiography, which can measure the aorta-SMA distance and visualize the amount of intra-abdominal and retroperitoneal fat.¹⁸

Initial treatment of SMA syndrome is nonsurgical. GI decompression with nasogastric tube placement to deflate the dilated stomach and fixing electrolyte abnormalities are the first goals of therapy.⁸ Therapy is then aimed toward treating the underlying cause which consists of increasing the aortomesenteric angle by reconstituting the fat pad. This can be done by either providing nutrition via enteric nasojejunal feeds or via hyperalimentation. Surgery is indicated for patients who do not improve from conservative management or in conditions where enteral feeding and/or parenteral nutrition is not feasible such as in our

patient who needed prompt chemotherapy for his intestinal lymphoma.

The three common surgical procedures to treat SMA syndrome described in literature are gastroduenojejunostomy, duodenojejunostomy, and dissection of ligament of Treitz called Strong procedure, which have been described laparoscopically too.¹⁶ Strong technique involves displacement of the duodenum downward and away from the apex of the aortomesenteric angle.¹⁰ It is advantageous since it helps to avoid enterotomy but has high recurrence rates unlike the bypass techniques, probably due to the postoperative adhesions, which tether the bowel into a similar position as the ligament of Treitz.¹⁹

Thus, most surgeons prefer duodenojejunostomy as the procedure of choice for SMA syndrome.³ The latter technique has been extensively used over the years, with Wilkie¹ alone reported to have successfully performed it on 64 patients with SMA syndrome. Gastroduenojejunostomy may be ideal when SMA syndrome patients present with severe duodenal dilatation, associated peptic ulcer disease, or when it is difficult to mobilize the duodenum.²⁰

Delay in diagnosis can result in dehydration, electrolyte imbalance, and even death.⁶ Other complications include acute respiratory distress syndrome due to aspiration of gastric contents²¹ and gastric perforations.²²

Gastric MCL causing SMA syndrome has never been reported as both conditions are uncommon. Approximately 6% of lymphomas are classified as MCL and they are described in adults with a median age of 60 with a male predominance. Our patient fits the exact description.^{23,24}

MCL usually responds poorly to conventional therapeutic regimens and is associated with short median survival because of early relapse. Median survival with standard treatment for MCL patients' remains between 3 and 4 years.²⁵ As survival rates are poor, our patient needed prompt chemotherapy and redressal of his symptoms. Duodenojejunostomy was therefore a good choice in our patient that allowed him to eat a regular diet, abated his symptoms, and allowed him to receive chemotherapy sooner.

Conclusion

To our knowledge this is the first case of MCL presenting with SMA syndrome. SMA syndrome is a rare condition that affects patients after substantial weight loss and should be considered as one of the differentials in a patient who presents with vomiting and history of rapid weight loss. Our patient did not qualify for conservative management and underwent duodenojejunostomy with excellent recovery and weight gain.

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