Superior Mesenteric Artery Syndrome: Diagnosis and Management

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ABSTRACT

Superior mesenteric artery syndrome is a life-threatening rare acquired upper gastrointestinal disorder due to mechanical compression of third part of duodenum by the acute angulation of Superior mesenteric artery, leading to obstruction. Acute loss of intervening mesenteric fat as a result of a variety of debilitating conditions is believed to be the etiologic factor causing the reduced aortomesenteric angle. Abdominal CT angiography showed the dilatation of second part of duodenum and vascular compression of the proximal third part of the duodenum between the aorta and superior mesenteric artery. We report a case of 15 year old young boy who presented with recurrent postprandial pain in the epigastric region, accompanied by epigastric fullness, nausea, postprandial bilious vomiting and weight loss. When conservative measures were ineffective, laparoscopic retrocolic duodenojejunostomy, side to side anastomosis, was performed in the patient to relieve the obstruction. This case report is unusual as it is concerned with the description of a rare disease entity and its radiological appearances for early preoperative diagnosis, better understanding and management of the disease are discussed in the pertinent light of literature.

KEY WORDS

CT angiography, duodenal obstruction, duodenojejunostomy, superior mesenteric artery syndrome

INTRODUCTION

Superior mesenteric artery (SMA) syndrome is an uncommon cause of upper intestinal obstruction caused by trapping of the third part of the duodenum between the aorta and the SMA.[1] Acute loss of intervening mesenteric fat pad as a result of a variety of debilitating conditions like immobilization, rapid and dramatic weight loss for any reason or after scoliosis surgery, external compression due to body cast treatment of a spinal fracture is believed to be the etiologic factor causing the narrowing of aortomesenteric angle.[2] Wilkie suggested other causes, such as a congenitally low origin of the SMA, a high insertion of the duodenum at the ligament of Treitz, and compression of the duodenum caused by peritoneal adhesions after duodenal malrotation.[3,4] It poses a difficult diagnostic dilemma but high resolution ultrasound power color doppler sonography, CT angiography or MRI confirm the diagnosis. This article reports the case of SMA syndrome in a young boy who presented with a three month history of nausea, postprandial vomiting, epigastric fullness and recurrent postprandial abdominal pain.

CASE REPORT

A 15 year old young asthenic boy was admitted to the Geetanjali Medical Hospital, Udaipur, with one month history of recurrent postprandial abdominal pain at the epigastric region, epigastric fullness, nausea and postprandial bilious vomiting.He reported acute weight loss of 8 kg over three months. Pain was colicky and nonradiating in nature. The patient denied any past surgical or medical history. Abdominal examination revealed epigastric tenderness and hyper peristaltic bowel

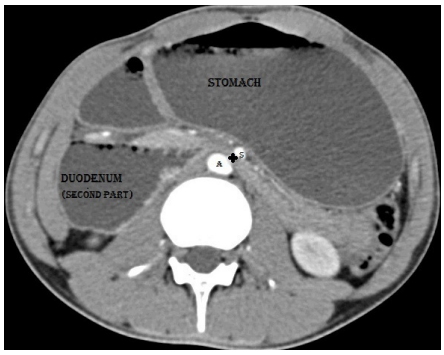

Figure 1. Axial contrast enhanced CT Scan showing compressed third part of duodenum (asterisk representing Aortomesentric distance = 4.2 mm) between aorta (A) and SMA (S), resulting in proximal upstream dilatation of proximal duodenum and stomach.

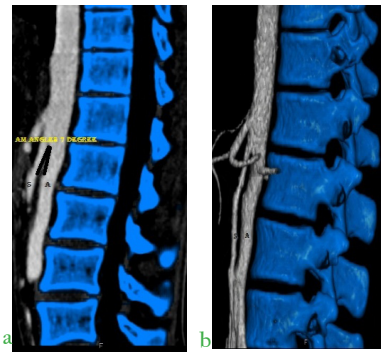

Figure 2. a. Multiplanar reformation image showing marked narrowing of aortomesentric angle (AM Angle =7°) between Aorta (A) and Superior Mesenteric Artery (S). b. Volume rendered surface shaded display signalling towards reduced aortomesenteric distance of 4.2 mm (normal: 10 to 28 mm). Superior Mesenteric Artery (S) dropping posteriorly, trapping the duodenum like a scissors.

sounds. There was no relief of obstruction in the left lateral decubitus or prone position. His vitals were stable. Routine blood and urine examination were unremarkable. Erythrocyte sedimentation rate (ESR) and Mantoux test were negative. Oesophagogastroduodenoscopy (OGD) showed no evidence of gastric pyloric obstruction. The upper gastrointestinal contrast study with a hypotonic duodenography revealed a distended proximal duodenum and narrowing in the third part of duodenum,hence excluding incomplete rotation of midgut and duodenal stenosis. A subsequent helical CT angiography (Fig. 1) and abdominal CT scan was performed that showed distension of the stomach with dilated first and second part of the duodenum, abrupt vertical and oblique compression of the 3rd part, where it is abruptly tapered and completely clamped between the SMA and the abdominal aorta. Fig. 2(a) and Fig. 2(b) revealed the marked narrowing of the aortomesenteric angle to 7° (normal: 25° to 60°) and reduced aortomesenteric distance of 4.2 mm (normal: 10 to 28 mm) respectively.

These findings in conjunction with clinical symptoms and signs were highly suggestive of SMA syndrome. A prokinetic conservative treatment was tried over three weeks with peripheral parenteral nutrition and nasogastric drainage for gastric decompression, followed by post-pyloric feeding by a Dobhoff catheter and mobilization into the prone, modified knee-chest or left lateral down position. Metaclopromide 0.5 mg/kg/day along with fluid resuscitation by inserting a nasojejunal feeding tube under endoscopic and fluoroscopic guidance, restored total enteral nutrition to 80 ml/h. The stenting effect of Nasojejunal tube also helped to drain the gastric and duodenal fluid to jejunum and distal bowel. Enteral jejunal feeding and parenteral nutrition support was tried for 3-4 weeks to maintain the nutrition and correct the weight loss, with monitoring the Ryle's tube aspirate output. After four weeks, he started tolerating oral diet in the form of fractionated meals and was discharged with weight monitoring and follow-up. However, the conservative treatment failed a few months later and the patient presented again with bilious vomiting and diffuse abdominal pain. There was relapse syndromic obstruction and pronounced duodenal dilatation with stasis, so at this point, subsequent surgical consultation was indicated. After adequate nutritional optimization pre-operatively, laparoscopic duodenojejunostomy under general anesthesia with endotracheal intubation was planned. The abdomen was entered via a left subcostal incision in the midclavicular line using a 10-mm optiview trocar under direct vision.

After insufflation of the abdomen and establishment of carbon-dioxide pneumoperitoneum of 12 mmHg, a 10 mm, 30 degree laparoscope was used to maximize visualization. Three additional trocars were placed under direct laparoscopic vision: a 5 mm right subcostal port, a 5 mm left lateral port and a 12 mm port was made on the left lower quadrant abdomen. After retraction of the transverse mesocolon superiorly, small bowel or colon was decompressed and the ligament of Treitz was identified. Intra-operative findings confirmed the distension of first and second part of duodenum with extrinsic obstruction of the distal part, immediately to the right of SMA. The site of obstruction was further confirmed by nasogastric air-insufflations. A duodenojejunostomy was performed using proximal jejunum approximately 30 cm distal to the ligament of Treitz. A side-to-side anastomosis was created with a 45 mm, 2.5 endoscopic linear stapler, and 2–0 vicryl sutures for closure of the enterotomy. The abdomen was irrigated and the trocar were removed. A methylene blue test showed no leak, and a nasogastric tube was left in the stomach. Operative time was 140 minutes with minimal blood loss. An upper gastrointestinal contrast study performed three days after surgery demonstrated emptying of gastric contrast through the anastomosis into the jejunum and suggested no leak with a patency of

the duodenojejunal anastomosis. The patient recovered uneventfully, as his occlusive symptoms resolved and he started to gain weight. He was well when examined at his 2 month, 4 month and 6 month post-operative follow-up visit.

DISCUSSION

In 1861, von Rokitanski first described SMA syndrome. Subsequently, Wilkie reported a more detailed clinical and pathophysiologic description of seven instances of SMA syndrome in 1927.[4] Due to this exceedingly rare occurrence, an incidence of 0.013–0.3% in the general population with a mortality rate of 33% have been reported in the world literature.[5,6] The defining feature of this entity is mechanical compression of the third part of the duodenum, between the aorta and the origin of superior mesenteric artery, leading to marked narrowing of aorto-mesenteric angle and reduced aorto-mesenteric distance with values of 6° to 15° and 2 to 8 mm respectively.

In our case, aorto-mesenteric angle and aorto-mesenteric distance are 7° (normal: 25° to 60°). and 4.2 mm (normal: 10 to 28 mm) respectively. The cause of compression of third part of duodenum is loss of retroperitoneal and periduodenal fat, which normally acts like cushion around the SMA. Acute loss of intervening mesenteric fat pad as a result of a variety of debilitating conditions like immobilization, external compression due to body cast treatment of a spinal fracture, catabolic and postoperative states, trauma, rapid and dramatic weight loss for any reason or after scoliosis surgery, bariatric surgery, ileoanal pouch anastomosis and aortic aneurysm repair is believed to be the etiologic factor causing the narrowing of aortomesenteric angle and reduced aortomesenteric distance.[1,2]

Wilkie suggested other causes, such as a congenitally low origin of the SMA, a high insertion of the duodenum at the ligament of Treitz, and compression of the duodenum caused by peritoneal adhesions after duodenal malrotation and an abnormal high, fixed position of ligament of Treitz.[3,4,7] In general, most cases of SMA syndrome clinically presents with nausea, vomiting, postprandial abdominal pain, epigastric fullness and weight loss.[8] However, the classic diagnostic upper gastrointestinal series with the following strict radiographic criteria should be established for diagnosis of SMA syndrome: (i) duodenal dilatation of the first and second part with or without gastric dilatation, (ii) characteristic vertical linear extrinsic pressure on one of the most fixed parts of the duodenum (third) with abrupt vertical and oblique compression of the mucosal folds (iii) antiperistaltic flow of contrast medium proximal to the duodenal obstruction, (iv) continous active peristalsis and delay in transit of four to six hours through the gastroduodenal region, and (v) relief of duodenal obstruction in the left lateral decubitus, knee-chest (Hayes maneuver), or prone position, so that the drag on the small-bowel mesentery get diminished.[1]

Once identified, the initial treatment approach should be conservative with advanced intensive nutritional strategies. The aim for conservative treatment is to correct/ break the vicious cycle of weight loss with implementation of enteral or parenteral feeding. With adequate fluid resuscitation, mobilization into the prone, modified knee-chest (Hayes maneuver) or left lateral decubitus position, gastric decompression via nasogastric drainage and/or post-pyloric feeding when possible, followed by oral diet as tolerated, most patients with SMA syndrome do not require surgery.[1,5] However, if chronic refractory gastroparesis persists and conservative measures are ineffective; surgery is indicated because refractory obstruction may led to peptic ulcer disease.[9]

Surgical treatment options, including open or laparoscopic duodenojejunostomy, duodenoduodenostomy and Roux-en-Y bypass, gastrojejunostomy and division of the ligament of Treitz with duodenal mobilization for lowering the duodenojejunal flexure (Strong's operation), are normally indicated in symptomatic patients to resolve or bypass duodenal compression.[5,9] Gastrojejunostomy, a previously reported surgical technique, has been abandoned because of increased postoperative complications like blind loop syndrome, especially when patient has megaduodenum (like in SMA syndrome). It will further exacerbate duodenal dysmotility and bacterial overgrowth. Studies reported Strong's operation to be less invasive, however, this surgical option had a failure rate of 25%.[6] In a retrospective study, Pottorf et al. concluded that laparoscopic enteric bypass is safe and effective and should be considered the optimal treatment for patients presenting with duodenal obstruction from superior mesenteric artery syndrome.[10] The authors add if a surgical drainage procedure is inevitable, duodenojejunostomy is the operation of choice to relieve the obstruction, with a success rate up to 90%.[9,10]

The present case is a rare entity that requires a high index of suspicion in patients with long standing abdominal complaints. Early diagnosis may allow appropriate management by interrupting the cycle of weight loss and duodenal obstruction, resulting from SMA syndrome. If conservative measures are ineffective, surgical treatment (either laparoscopic or open method) is the only accepted method of managing SMA Syndrome.

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