Superior Mesenteric Artery Syndrome; Case Report, Full Diagnostic Approach and Treatment

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ABSTRACT

Introduction: Superior mesenteric artery (SMA) syndrome is a rare cause of proximal intestinal obstruction. It is mostly seen in young and underweight females.

Case presentation: We present a case of a 25-years-old cachectic male with bi-temporal atrophy, with prominent decrease in urination, anorexia, severe nausea and vomiting and vague abdominal pain increased postprandial and were relieved in lateral decubitus or knee-elbow (squatting) position; abdominal x-ray showed double bubble sign (obstruction) and CT-scan showed compression of third part of duodenum between aorta and superior mesentery artery and reduction in aortomesenteric distance. To all these, we established diagnosis of SMA syndrome whom was successfully treated first with hemodialysis due to his high blood creatinine and then -after full resuscitation- underwent open duodenojejunostomy for his duodenal obstruction.

Discussion: Causes of SMA syndrome are both Congenital which both of them usually decrease aortomesenteric angle causes extrinsic compression of third part of the duodenum. It can be acute or chronic. Symptoms are recurrent epigastric pain, bilious vomiting, early satiety and postprandial discomfort increases in the supine position and relieves in left lateral or knee-elbow position. The diagnosis is based mostly on clinical symptoms, excluding other causes of obstruction of the duodenum and radiologic evidence of obstruction. The treatment commonly entails medical therapy and if it fails or in severe cases surgical therapy is preferred (duodenojejunostomy, gastrojejunostomy, Strong's procedure and transposition of the SMA in the infrarenal aorta in adults and Ladd's procedure in children).

Key Words: Superior mesenteric artery, Wilkie syndrome, weight loss, abdominal pain

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INTRODUCTION

Superior mesenteric artery (SMA) syndrome (Wilkie syndrome) is a rare cause of proximal intestinal obstruction [1,2]. It is due to compression of third portion of duodenum as a result of narrowed space between the SMA and aorta due to several causes [3].

It is most commonly seen in young, underweight, and female individuals.[4] Symptoms include postprandial epigastric abdominal pain, and obstructive symptoms in more advanced cases including profound nausea and

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bilious vomiting. This condition can lead to further weight loss, as well as in nutritional and electrolyte abnormalities. Rare, yet severe, complications include gastric perforation and peritonitis (can be fatal).

The treatment commonly entails medical therapy (nasogastric decompression, fluid resuscitation, correction of electrolyte abnormalities and nutritional support via parenteral or enteral (nasojejunal feeding access) means for weight loss); and if it fails or in severe cases surgical therapy is preferred to prevent further complications.

Surgical options in adults include duodenojejunostomy, gastrojejunostomy, Strong's procedure and transposition of the SMA in the infrarenal aorta and Ladd's procedure in children. Open and laparoscopic approaches have been described.

We present a case of a 25-year-old cachectic male, that we established diagnosis of SMA syndrome whom was successfully treated at our institution via open duodenojejunostomy after full resuscitation and gained weight in follow up.

Presentation of case:

A 25-year-old man admitted to Emergency Department of Emam Hussein Hospital, Shahid Beheshti University, Tehran, Iran, at February 2015, presented with chief complaint of prominent decrease in urination from 3 days prior. In addition, he complained of anorexia, severe nausea and vomiting (followed by confusion) and vague abdominal pain from 3 month before, that increased recently; but he didn't seek healthcare.

At the time of admission he had epigastric and upper abdominal, non-crampy, constant compressive, non-radiating pain. His symptoms increased postprandial and were relieved in lateral decubitus or knee-elbow (squatting) position.

Physical examination revealed a cooperative young cachectic patient with bi-temporal atrophy and mild altered mental statues (little confusion). Vital signs are: blood pressure: 90/pulse mmHg; heart rate: 120 beats/minute; respiratory rate: 22/minute; with a low grade fever (38.2℃). He had about 15 kg weight loss in last three months (weighed 45 kg, 171 cm tall and his BMI was 15.39 at admission).

Abdominal examination revealed a mild epigastric and upper abdomen tenderness (with rebound tenderness); without distension, hypoactive bowel sounds, palpable mass and organomegaly. DRE was fecal stained.

Primary laboratory data was CBC: WBC:16500-nut:90%, Hb:14.9, plt:402000; Bs:160, Urea:190, Cr:5.9, Na:124, k:3.7, uric acid:13.9; LFT: normal; VBG: ph:7.47, pCO2:30, HCO3:38.3; U/A: SG:1027, Keton:1+; these showed leukocytosis, mildly high hemoconcentration (due to severe dehydration), prerenal azotemia, hyponatremia and alkalosis.

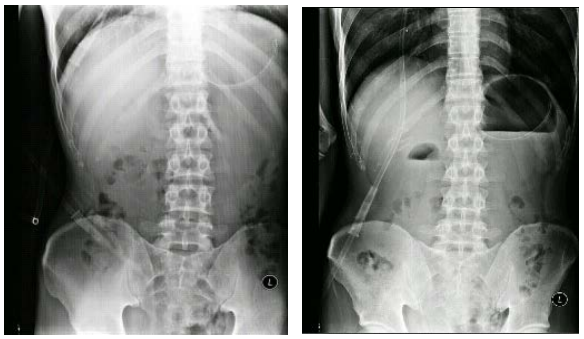
Because of severe decrease in urination and high blood creatinine, the patient was admitted to Nephrology floor for hemodialysis (via double tunnel jugular catheter).

After 3 days creatinine level became normal and he was completely conscious, but nausea remained, and he developed intensive constant abdominal pain. There was no defecation from the day of admission, but minimal gas passing. He couldn't tolerate per mouth - Solid or liquid - due to severe nausea and pain.

His second Laboratory data (after early management) was CBC: WBC: 12500-nut:90%, Hb:11.1, plt:224000; Bs:160, Urea:113, Cr:1.6, Na:140, k:3.2; VBG: ph:7.41, pCO2:39.5, HCO3:19; and except mild anemia and Hypokalemia, all others were normal.

At this stage, we perfumed radiologic assessments; Abdominal X-ray showed no gas in bowels in supine position (figure 1) and double bubble sign in upright position (figure 2); Contrast enhanced abdominal CT-scan showed dilated stomach, first and second part of duodenum and compression of third part of duodenum between aorta and superior mesentery artery, the aortomesenteric distance was clearly reduced, and there is high dilution of oral contrast in proximal duodenum, due to high accumulation of fluid (figure 3). These radiological features supported the diagnosis of SMA syndrome. (Due to severe obstruction, angiography was not performed to assess aortomesenteric angel.)

Figure 1 **Figure2**



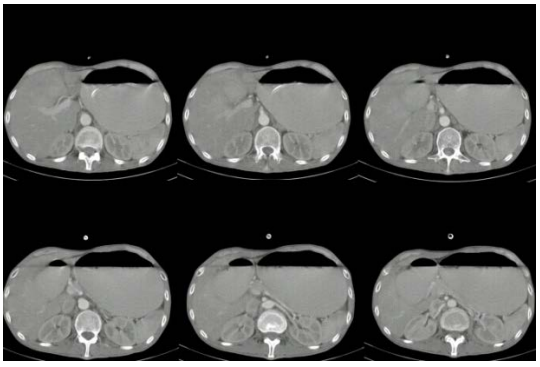


Figure 3

Firstly, the patient was managed with intravenous fluid (Normal saline); after adequate urination, KCl was added. Nasogastric suction and gastric irrigation was performed, dark bilious secretion was came out and results in reduction of nausea. He was on parenteral nutrition for about 24 hours before surgery. At this Stage, vital signs are: blood pressure: 105/70 mmHg; heart rate: 110 beats/minute; respiratory rate: 17/minute; Temperature: 37.6℃.

After full resuscitation the patient was prepared for urgent exploratory laparotomy through a midline incision (with general anesthesia). Intraoperative findings were severely dilated stomach distended to pelvic, extrinsic

obstruction of third part of duodenum by SMA, with distension of first and second parts of duodenum. (Figure 4) Jejunum and the rest of the abdomen were clubbed. Gut continuity was restored via a side-to-side duodenojejunostomy (from about 20 cm of Treitz ligament) -looped anterior to superior mesenteric vessels- by a 4 cm longitudinal incision; separated gambi suture with monocryl 3-0 was done. Drain was inserted near anastomosis site and surgery finished.

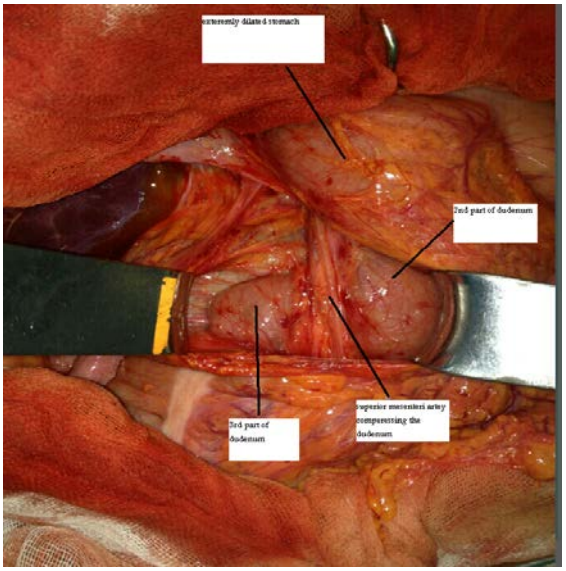


Figure 4

Post operation, patient was on Short course of TPN, and then had NG tube for 4 days; and it was removed when oral liquid diet started. Soon after, patient tolerated full Oral diet. There was no fever and infection. Creatinine level and electrolytes remain normal and there is no need for hemodialysis any more.

In first 10 days follow-up visits, he was completely symptom free and sutures were pulled; and in second one month follow up, had gained 8 kg in weight (weighed 53 kg).

DISCUSSION

The SMA syndrome was first described by von Rokitanski in 1861.[5] Wilkie later published one of the largest series, a total of 75 cases in 1927. The incidence of this condition varies between 0.013 and 0.3 % of barium series of the upper gastrointestinal tract.2

The third part of duodenum is in a fixed compartment bound anteriorly by the mesenteric root and the SMA and posteriorly by the aorta and the lumbar spine.[6] Normally, retroperitoneal fat and lymphatic tissues serve as a cushion around the SMA, protect the duodenum from vascular compression and holding it off the spine.

Causes of SMAS is likely multifactorial. Congenital factors are the high and fixed suspensory of the ligament of Treitz and low origin of SMA. Acquired factors include thin body form, high lumbar lordosis, visceroptosis, abdominal wall laxity, depletion of the mesenteric fat by rapid and severe weight loss as the most common cause (due to wasting conditions: AIDS, malabsorption, cancer and cerebral palsy; that associated with cachexia), prolonged bed rest owing to severe injuries, dietary disorders (anorexia nervosa and drug abuse), and catabolic states (burns), and complication of spinal surgery, aortic aneurysm repair, ileoanal pouch anastomosis and body cast usage (in scoliosis and vertebral fractures); which all of above usually decrease the aortomesenteric angle causes extrinsic compression of the third part of the duodenum (SMA is anterior and aorta is posterior). [7, 8,9,10]

It can manifest itself as either an acute or chronic episode. Patients often present with chronic symptoms of recurrent pain in epigastrium, vomits (bilious), early satiety and weight loss and postprandial discomfort. These symptoms are aggravated post-prandial or in the supine position and are relieved by lying in a left lateral or knee-elbow position.

The diagnosis of SMA syndrome is usually difficult and delayed because of its nonspecific symptoms, resulting in many complications (abnormalities in hydration, nutrition and electrolyte). Also, compression of the duodenum in a true SMA syndrome is often intermittent and incomplete, so patients are usually asymptomatic between the episodic attacks, and even during the attacks, gastric and duodenal contents can still slowly pass through the aortomesenteric angle.[9,10,11,12,13]

The diagnosis is based mostly on clinical symptoms, excluding other causes of obstruction of the duodenum (exclusion of other differential diagnosis) and radiologic evidence of obstruction by Plain radiography (demonstrates a dilated, fluid and gas-filled stomach), Hypotonic duodenography Barium studies(gold standard diagnostic modality; characteristics include dilatation of the first and second portions of the duodenum with an abrupt vertical or linear cutoff in the third portion), Esophagogastroduodenoscopy (reveal a normal mucosal folds, dilated proximal duodenum with gastric fluid retention and narrowing of the third part of the duodenum due to extrinsic compression), abdominal Doppler ultrasound, angiography (with a lateral aortography to measure the aortomesenteric angle; however, this procedure is invasive, expensive, and time-consuming) and multidetector CT-scan (better modality because it provides more diagnostic information than conventional angiography, including aortomesenteric distance, duodenal distention, visualization of vascular compression of the duodenum and the amount of intra-abdominal and retroperitoneal fat) or MRI.[9,12,14,15]

Normally, SMA angle is 45 degrees (25-60 degrees) while the aortomesenteric distance is 10 to 28 mm. In the SMA syndrome, the aortomesenteric angle is narrowed to 6 to 15 degrees and the aortomesenteric distance is reduced to 2 to 8 mm.[2,16,17]

The differential diagnosis includes mega duodenum, chronic pancreatitis, irritable bowel syndrome, duodenitis, cholelithiasis, peptic ulcer disease and aneurysm of the abdominal vessels.[18]

Surgical intervention has indication for failed conservative treatment (NG decompression, dehydration correction, electrolyte imbalance prevention and hyper alimentation followed by oral feeding with frequent small meals, posturing maneuvers during meals and prokinetic drugs and motility agents: cisapride and metoclopramide),

for longstanding disease with progressive weight loss and duodenal dilatation with stasis, complicated peptic ulcer disease secondary to biliary stasis and reflux.

Surgical options are Duodenojejunostomy (most common and most effective, with a success rate of 90 %)[6,19], gastrojejunostomy (has been largely abandoned due to several complications including blind loop syndrome, dumping syndrome, and marginal ulceration; leading to possible reoperation of duodenojejunostomy),(As with any anastomotic procedure however, leakage and stricture of the site may present as major complications requiring reoperation. In addition, a non-physiologic loop is created);Strong's procedure (duodenal derotation by means of lysis of the Ligament of Treitz, with mobilization of the fourth portion of the duodenum; it has advantage of avoidance a gastrointestinal anastomosis, thus earlier postoperative recovery); and transposition of the SMA in the infrarenal aorta (without disrupting the continuity of the small bowel). Both open and laparoscopic approaches have been described. [7,11,13,18,20]

To date, there is no data to an optimal period or indications for preoperative or postoperative nutritional support either enterically or by TPN. Although it is well recognized that significant preoperative weight loss is associated with an increased risk of postoperative complications. [14, 18]

CONCLUSIONS

SMA syndrome is a life-threatening upper gastrointestinal disorder due to vascular compression of duodenum. It is uncommon, the diagnosis is usually difficult and delayed because of its nonspecific symptoms, resulting in complications. It should be taken into consideration in all patients with important weight loss and gastric distension and treated as soon as possible. Conservative treatment can be given especially in children but surgery is the treatment of the choice.

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