

Superior Mesenteric Artery Syndrome: A Single-institution Experience

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ABSTRACT

Background:

Superior mesenteric artery syndrome (SMAS) is a rare disease in adult. SMAS is characterized by acute, or, more commonly, chronic nonspecific symptoms due to duodenal obstruction and severe malnutrition with reduced arterio-mesenteric angle and distance. Surgical treatment may be necessary in most cases with chronic symptoms or when conservative treatment fails in SMAS.

Methods:

A retrospective chart review was performed on patients who underwent operation for SMAS from January 2008 to August 2020 in Cardinal Tien Hospital. Patients' clinical presentations, surgical intervention, and outcomes.

Results:

Data from a total of 14 patients diagnosed with SMAS were analyzed, of which seven were diagnosed with SMAS by abdominal computed tomography and upper gastrointestinal series with water-soluble barium contrast. Six of the confirmed cases underwent surgery, namely, gastric decompression using a nasogastric tube, and correction of electrolyte imbalance. The nasoduodenal tube was placed through the obstructed duodenum to provide a high-nutrient fluid supplement. After conservative treatment failure, the patients underwent surgery. Of the six patients, four underwent duodenojejunostomy, one underwent a mini-laparotomy duodenojejunostomy bypass, and the last one underwent Roux-en-Y duodenojejunal bypass with duodenal feeding tube insertion.

Conclusion:

Patients with SMAS should initially be treated conservatively. Surgical intervention should be considered in patients in whom conservative treatments were not effective. Complete resolution of all symptoms may not always be guaranteed after surgical intervention. Laparoscopy is currently widely used. In well-selected patients, minimally invasive or mini-laparotomy duodenojejunostomy is a safe and effective treatment for SMAS. The main advantages of mini-laparotomy duodenojejunostomy over other surgical approaches include half-length surgical incision and a shorter operative time. Duodenojejunostomy is rapidly becoming the standard procedure of this condition, and it has excellent outcomes comparable with those of open surgery.

Keywords:

Clinical cases, SMAS, minimal invasive duodenojejunostomy.

Background

Superior mesenteric artery syndrome (SMAS) was first described in 1842 by Rotikansky [1] and was subsequently studied by Wilkie in 1921 [2]. He also defined the pathophysiological changes of the third segment of the duodenum when obstructed following arteriomesenteric compression. Wilkie [2] used the term “chronic duodenal ileus” in 1927 [3]. Some studies have reported that SMAS occurs in 0.013%–0.3% of the general population.[4,5] Moreover, 75% of the patients with SMAS are 10–30 years old and predominantly women.[4-6] The main symptoms include postprandial abdominal pain, early satiety, vomiting, and weight loss. [7,8] There is a significant psychological overlap in many patients who present with this condition.[9]

Diagnosis of SMAS is challenging, but it may be suspected based on clinical presentation and supported by imaging studies.[7,10,11] A delay in the diagnosis of

SMAS can result in malnutrition, electrolyte imbalance, gastric perforation, pneumatosis, and hypovolemia, with a reported mortality rate of up to 33%.[12-15] SMAS symptoms do not always correlate well with abnormal anatomic findings on radiologic studies and may not resolve completely after treatment. [4,16,17] We presents patients with SMAS and compared their clinical course from our hospital. Besides, mini laparoscopic duodenojejunostomy is feasible as better method of surgery compared with other surgical approach.

Methods

A retrospective chart review, which was approved by an institutional review board, was performed in all patients with SMAS who underwent duodenojejunostomy and Roux-en-Y duodenojejunostomy from January 2008 to August 2020 at Xin-Dain Cardinal Tien Hospital (Table 1).

Cases	Age	Sex	Symptoms	BMI (kg/m ²)	Aortic-mesenteric angle	Management
Case 1	59	Female	Vomiting, epigastric pain	19.8	22	Exploratory laparotomy with enterolysis and side-to-side duodenojejunostomy
Case 2	20	Male	Vomiting, epigastric pain, bloating	15.5	18	Exploratory laparotomy with lysis of adhesion and duodenojejunostomy
Case 3	27	Male	Nausea, Vomit, epigastric pain, reflux	13.5	11	Exploratory laparotomy with lysis adhesion, Roux-en-Y duodenojejunostomy
Case 4	28	Female	Nausea, epigastric pain, bloating	17.5	15	Exploratory laparotomy with Roux-en-Y duodenojejunostomy with mild partial obstruction
Case 5	20	Female	Nausea, vomiting, epigastric pain, bloating	20.5	8	Minimal exploratory laparotomy with lysis of adhesion and duodenojejunostomy
Case 6	71	Male	Nausea, epigastric pain, bloating	16.8	21	Conservative treatment without operative intervention
Case 7	40	Male	Epigastralgia, constipation, dysphagia	17	11	Exploratory laparotomy with enterolysis and side-to-side duodenojejunostomy

Table 1: Patients' background, disease course, and their clinical management.

Case Report

A 20-year-old Taiwanese female patient presented to a general surgery outpatient clinic with complaints of epigastric pain, nausea, vomiting, and weight loss (4 kg) for more than 6 months. Postprandial tenderness was noted and worsened in the supine position. Vomiting (six times a day) was noted 3–4 h after meals and consisted of undigested food. She denied a significant medical history. On physical examination, the patient was extremely emaciated and had a distended abdomen and fullness over the epigastrium. She had undergone an upper endoscopy, which revealed that a greenish solid debris impacted the second segment of the duodenum (Figure 1 A-B).

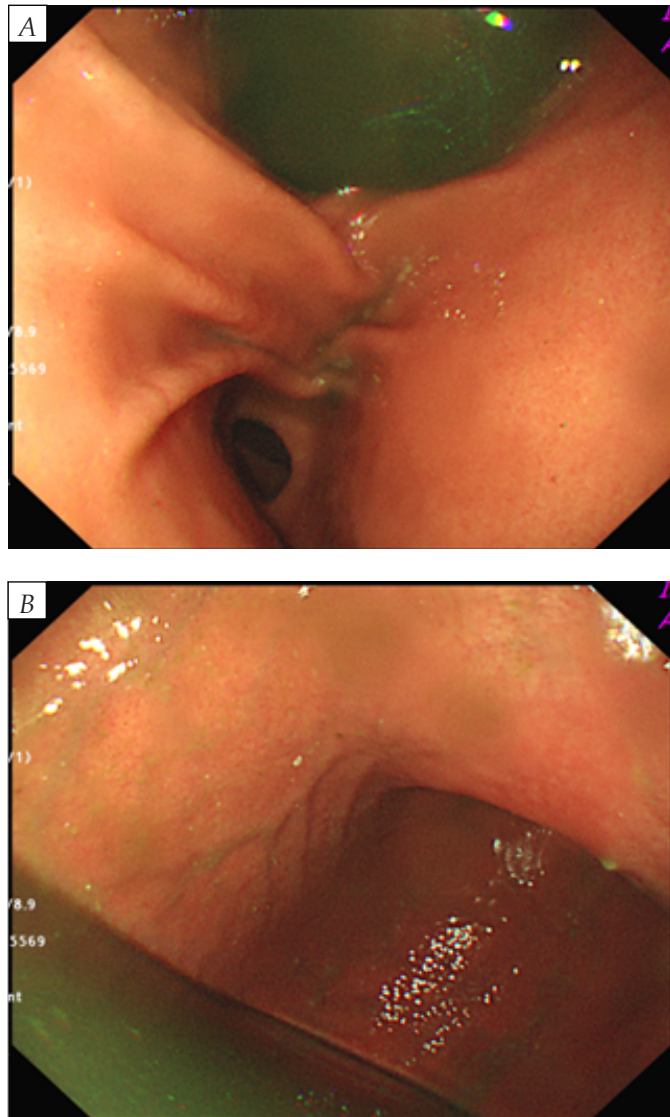


Figure 1:

- A. Intestinal obstruction at least below the second segment.
B. Greenish solid debris impacted the second segment.

Abdominal computed tomography (CT) was performed on the following day. It demonstrated distension of the first and second segments of the duodenum and severe distension of the stomach and proximal segment of the duodenum. Constriction of the third segment of the duodenum between the abdominal aorta and SMA, with a reduced angle (8°) and shortened distance (4–5 mm) between these two arteries (Figure 2 A-B), collapsed the

fourth segment of the duodenum and jejunum.

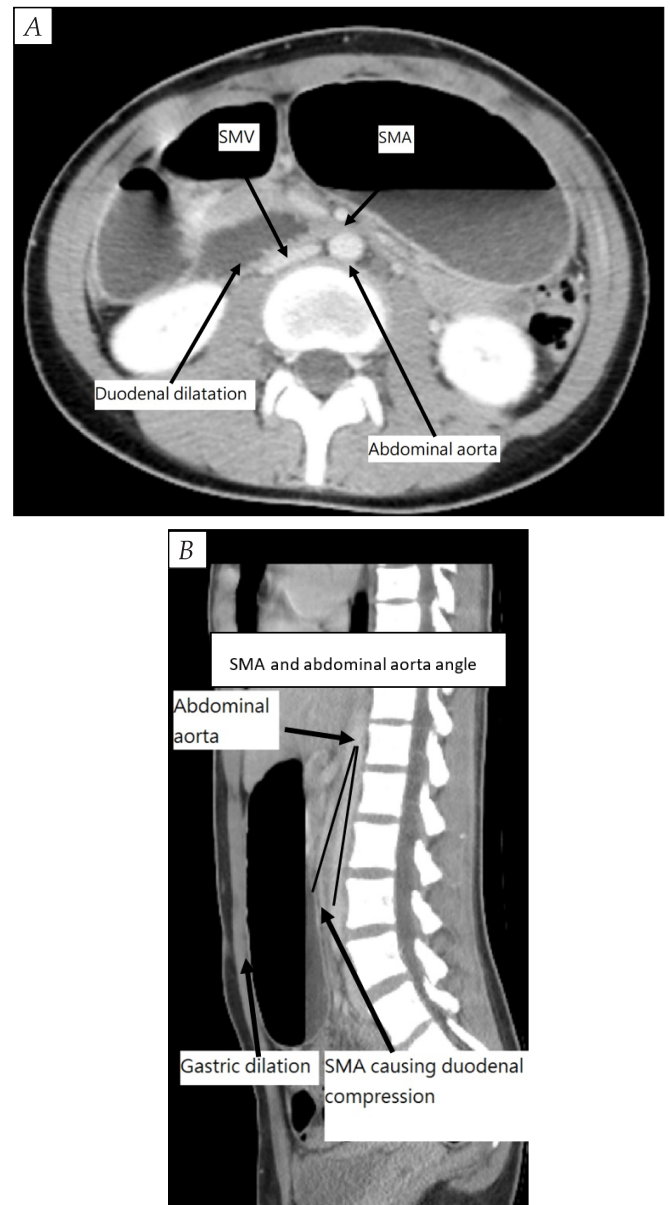


Figure 2:

- A. Axial CT section demonstrating compression of the third segment of the duodenum between the SMA and abdominal aorta, with proximal duodenal and gastric dilation
B. Sagittal CT section showing compression of the third segment of the duodenum between the SMA and abdominal aorta with an angle of 8° , resulting in proximal duodenal and gastric dilation. (CT, computed tomography; SMA, superior mesenteric artery; SMV, superior mesenteric vein).

An upper gastrointestinal (GI) contrast study was performed, which revealed a distended stomach with delayed gastric emptying and lagging of contrast at the third segment of the duodenum (Figure 3 A-B). These findings were suggestive of an aortomesenteric clamp. Hence, based on known findings, the diagnosis of Wilkie syndrome (SMAS) was established.

Initial conservative management was adopted to improve the patient's nutrition status. Surgical treatment was recommended to prevent disease recurrence. Our surgical approach was a mini-laparotomy duodenojejunostomy with a side-to-side stapled anastomosis between the jejunum (30–40 cm from

Treitz's angle) and the second segment of the duodenum (Figure 4 A-B). The patient recovered with less pain but with a delay in diet tolerance. She was discharged home after 6 days and was on a liquid diet, and digestive transit was restored. After 2 months of follow-up, the patient gained 3kg and remained asymptomatic.

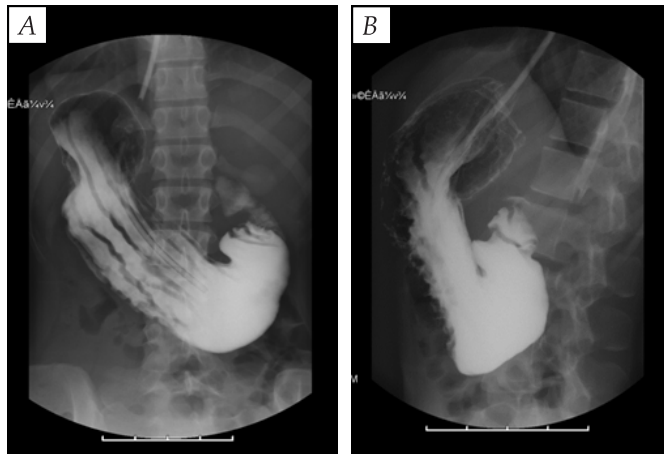


Figure 3:

- A. Dilated stomach and straight-line cut-off of the third segment of the duodenum.
 B. Minimal contrast medium runs into the fourth segment of the duodenum and proximal jejunum.

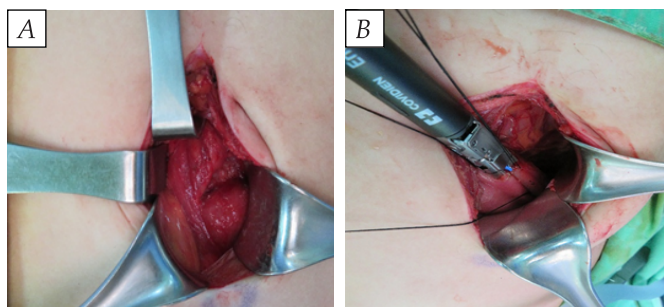


Figure 4:

- A side-to-side duodenojejunosomy performed using a 60-mm linear stapler advanced to 40 mm through an open approach.
 A. Open view of the duodenum and jejunum approximated prior to anastomosis.
 B. Enterotomies made in the jejunum and duodenum; a 60-mm linear stapler was advanced to at least at the 40-mm mark, then the enterotomy was closed with an interrupted suture.

Surgical technique

Under general anesthesia, the patient was placed in the supine position. The skin was prepared and cleansed with Betadine + 75% alcohol. In this procedure, the midline incision of the abdomen was made between the xiphoid and the umbilicus, measuring approximately 7 cm. The fascia was incised with a knife, and the fascial incision was lengthened with a cautery pencil. The linea alba was separated with cautery pencil, the peritoneum was picked with hemostasis, and then elevated. The peritoneum was nicked with a deep-point knife, and the incision was extended with Metzenbaum scissors. The abdominal contents were exposed. Sequentially, the duodenum and jejunum were identified at the ligament of Treitz. The peritoneum overlying the junction of the second and third segments of the duodenum was divided through the transverse mesocolon. The duodenum was gently mobilized off the retroperitoneum at this

level. A segment of the jejunum approximately 40 cm was closed with absorbable stay suture. Immediately adjacent to this, a 2-0 absorbable suture was placed. Enterotomies were performed in both the duodenum and jejunum between the sutures placed earlier. A side-to-side duodenojejunosomy was performed using a 60-mm linear stapler advanced to 40 mm through a mini-laparotomy approach (Figure 4 A-B). A fully hand-sewn technique was used.

The common enterotomy was then closed with a 2-0 absorbable interrupted suture. The abdomen was closed layer by layer. The patient underwent postoperative observation for 4 days with diet progression from a clear liquid diet to a semi-liquid diet, as she tolerated. Later, she was discharged with follow-up in the outpatient department.

Case series

Imaging studies were consistent with SMAS with an impression of an abrupt obstruction at the level of the SMA and dilatation of the proximal duodenum, which reflected clinical symptoms during physical examinations. In some cases, primary evaluation with gastroenterologists for endoscopic examination was performed. Imaging studies include CT of the abdomen with coronal reconstructions. In addition, CT angiography further delineated the vascular anatomy and relationship with the duodenum. All study patients had clinical characteristics and confirmatory radiologic findings consistent with the clinical features of SMAS. Data collected from the charts included patient clinical presentation, operative data, and outcomes such as morbidity, mortality, and further therapies.

Surgical techniques

Surgery was performed under mini-laparotomy duodenojejunosomy as described previously in case presentation above (as case 5). Two patients underwent exposed laparotomy with Roux-en-Y duodenojejunosomy (cases 3 and 4). One patient underwent tube enterostomy. Duodenojejunosomy reestablished bowel continuity with a success rate of >90%, and duodenojejunal continuity was the preferred surgery. Surgical complications include bleeding, leakage, or stricture at the anatomical site.[18]

Long-term concerns included small bowel bacterial overgrowth in the "blind loop" created in the bypassed third and fourth segments of the duodenum.

Another approach for duodenojejunosomy was the division of the fourth segment of the duodenum. This was not recommended because leaving the duodenojejunal continuity intact was the preferred approach.

Discussion

The diagnosis of SMAS should be considered in patients with clinical features of duodenal obstruction, especially with imaging studies revealing duodenal obstruction in the third segment of the duodenum with active retrograde peristalsis. The average mean angle formed by the SMA and the aorta varied ranged from 38° to 56°, and the mean radiographic aortomesenteric distance was 10–28 mm. Further imaging studies should be pursued to establish the diagnosis when the aortomesenteric

angle is $\leq 25^\circ$, and the aortomesenteric distance is ≤ 8 mm. Other clinical features of SMAS are high fixation of the duodenum by the ligament of Treitz, abnormally low origin of the SMA, or anomalies of the SMA.

The diagnosis, treatment, and indications for surgery are always challenging when dealing patients with SMAS. Surgeons must follow a strict algorithm before proceeding with surgical intervention. The following strict radiographic criteria have been established for the diagnosis of SMAS: (1) dilatation of the first and second segments of the duodenum, with or without gastric dilatation, (2) abrupt vertical and oblique compression of the mucosal folds, (3) retrograde flow of contrast medium proximal to the obstruction, (4) delay in transit by 4–6 h through the gastroduodenal region, and (5) relief of obstruction in a prone, knee-chest, or left lateral decubitus position.[5,19,20]

The initial management of SMAS should always be conservative. Fluid and electrolyte imbalance must be corrected, the stomach decompressed with a nasogastric tube, and nutritional support instituted with either nasojejunal feeding or total parenteral nutrition. Gastric prokinetic agents such as metoclopramide may be helpful for a short period of time. Patients with acute SMAS often respond to and benefit from conservative treatment. [5,19,21,22] However, patients with chronic disease and malnutrition may require surgical intervention.

Several surgical procedures including gastrojejunostomy, duodenojejunostomy, and Strong's operation have been performed to resolve or bypass duodenal compression in SMAS [5,23]. Both Strong's procedure and loop duodenojejunostomy have been performed using the laparoscopic approach, with shorter recovery time and hospital stay than conventional approaches by laparotomy.[19,21,23–25]

Strong [26] first described the division of the ligament of Treitz with mobilization of the transverse and ascending duodenum for caudal displacement of the duodenum. The advantages of this procedure are as follows: does not damage the bowel wall, less likely to have complications, shorter operative time, and shorter postoperative recovery course. As disadvantages, the procedure can be aggravated or impossible to carry out due to dense adhesions, and caudal displacement of the duodenum cannot always be achieved because of interference with short vessels from the inferior pancreaticoduodenal artery to the duodenum.[5,22] However, this procedure is now largely of historic interest and has a higher failure rate to relieve duodenal obstruction.[22]

A loop gastrojejunostomy has been shown to provide adequate gastric decompression but fails to completely release duodenal obstruction, leading to persistence of symptoms. Persisting obstruction may lead to blind-loop syndrome, gastric bile reflux, and ulceration, which necessitated further surgical intervention in some cases.[5,22] This may be a potential consideration when adhesions from previous surgeries prevent adequate access to create the duodenojejunal anastomosis.

Loop duodenojejunostomy as a treatment option for this condition is generally accepted as having superior results to both Strong's procedure and loop gastroenterostomy, with good results achieved in $\geq 79\%$ of patients with SMAS.[5,22] Surgical complications include bleeding,

leakage, or stricture at the anatomical site. Long-term concerns include small bowel bacterial overgrowth in the "blind loop" created in the bypassed third and fourth segments of the duodenum.[18]

Roux-en-Y duodenojejunal bypass is the only procedure in which no blind loop is left, with free drainage of not only the duodenum proximal but also distal to the compression site at the SMA.[27] This operation has also been reported to successfully treat recurrence in a patient 1 year after duodenojejunostomy.[28] SMAS caused by Roux-en-Y duodenojejunal bypass includes branches of the SMA that supply the transverse colon, may pass through the peritoneum that covers the duodenum, and should not be severed during the dissection to prevent compromising the blood supply to the colon.[29] Roux-en-Y duodenojejunostomy should be considered an alternative procedure, especially when duodenal obstruction occurs beyond the second segment of the duodenum.

Long-term outcomes in patients who developed SMAS following surgery are limited in the literature. One series of 16 patients showed significant weight gain, but most symptoms remained unchanged except for decreased vomiting.[30] Addressing eating disorders, bulimia, and underlying psychiatric issues is a key aspect of post-surgery management. Surgical morbidity and mortality can be affected by other comorbidities such as diabetes mellitus and end-stage renal disease.[31]

Conclusion

SMAS has a spectrum of symptoms, which can be referred to as "great mimickers" of a GI motility disturbance. The main GI motility conditions include delayed gastric emptying; dilated duodenum suggesting intestinal pseudo-obstruction; and unexplained nausea and vomiting and abdominal pain, suggesting a cycle of vomiting syndrome and irritable bowel syndrome. This is a great challenge for physicians in practice.[32]

The clinicians should pay attention to patients with unexplained abdominal pain provoked by eating and accompanied by nausea and vomiting; endoscopic evidence of retained food in the stomach and a dilated proximal duodenum; and a slow scintigraphic gastric-emptying result.

Patients with SMAS should initially be treated conservatively, by which clinical symptoms may potentially resolve spontaneously, and adequate nutrition and hydration are the main treatment focus. Surgical intervention should be considered in patients in whom conservative treatments were not effective. Complete resolution of all symptoms may not always be guaranteed after surgical intervention.

Laparoscopy is currently widely used. To prevent unnecessary complications during laparoscopy operation, the following are absolute contraindications: hypovolemic shock, large pelvic or abdominal mass, inadequate experience of the operator, inadequate equipment, and severe cardiac disease. In well-selected patients, minimally invasive or mini-laparotomy duodenojejunostomy is a safe and effective treatment for SMAS, with excellent short-term outcomes. The main advantages of mini-laparotomy duodenojejunostomy over other surgical approaches include half-length

surgical incision and a shorter operative time. Duodenojejunostomy is rapidly becoming the standard procedure of this condition, and it has excellent outcomes comparable with those of open surgery. However, we still needed to better define the optimal methods of diagnosis and treatment of the SMAS.

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CHC, VTKC, SJC, HCJ, THC, SCW, HYL, YHH conceptualized and designed the study, acquired, and analyzed data, interpreted the study results, drafted the manuscript, and critically revised the final version of the manuscript.

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Competing interests

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

Availability of data and materials

Further information is available from the corresponding author on reasonable request.

Ethics approval

The ethics review board approved this study and did not require informed consent from study participants since this was a strictly registry-based study.

Provenance and peer review

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