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Superior mesenteric artery syndrome: Diagnosis and management

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Abstract

Superior mesenteric artery (SMA) syndrome (also known as Wilkie's syndrome, cast syndrome, or aorto-mesenteric compass syndrome) is an obstruction of the duodenum caused by extrinsic compression between the SMA and the aorta. The median age of patients is 23 years old (range 0-91 years old) and predominant in females over males with a ratio of 3:2. The symptoms are variable, consisting of postprandial abdominal pain, nausea and vomiting, early satiety, anorexia, and weight loss and can mimic anorexia nervosa or functional dyspepsia. Because recurrent vomiting leads to aspiration pneumonia or respiratory depression via metabolic alkalosis, early diagnosis is required. The useful diagnostic modalities are computed tomography as a standard tool and ultrasonography, which has advantages in safety and capability of real-time assessments of SMA mobility and duodenum passage. The initial treatment is usually conservative, including postural change, gastroduodenal decompression, and nutrient management (success rates: 70%-80%). If conservative therapy fails, surgical treatment (*i.e.*, laparoscopic duodenojejunostomy) is recommended (success rates: 80%-100%).

Key Words: Superior mesenteric artery syndrome; Wilkie’s syndrome; Cast syndrome; Aorto-mesenteric compass syndrome

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Core Tip: To summarize, superior mesenteric artery syndrome (SMAS) is a rare condition that occurs when the superior mesenteric artery compresses the third part of the duodenum. This compression can cause obstructive symptoms and weight loss. SMAS can be caused by a variety of factors, including abnormal anatomy, rapid weight loss, and previous abdominal surgery. It is most commonly seen in young, thin females, but can occur in males and people of any age or body type. Treatment may involve

dietary modifications, medications, and surgery to correct the underlying cause. If left untreated, SMAS can lead to serious complications, including malnutrition and intestinal damage.

INTRODUCTION

Superior mesenteric artery (SMA) syndrome is a rare cause of duodenal obstruction by extrinsic compression between SMA and the aorta (SMA-Ao) (Figure 1), and a morbid entity when the diagnosis is delayed[1-3]. Von Rokitansky[1] first described this entity in his textbook with a case presentation in 1842. Later, Wilkie[2] described pathological and diagnostic findings in details with 75 cases of his own in 1927, thus SMA syndrome is also known as Wilkie's syndrome. The other names are reported as cast syndrome, aorto-mesenteric compass syndrome, or mesenteric duodenal obstruction[3]. A cast, which is used to treat certain congenital deformities such as scoliosis and hip displacement, is a major cause of SMA syndrome[4-6]. By the year 2022, more than 730 articles with approximately 2400 cases of SMA syndrome had been reported (author's review in PubMed). Initial conservative therapy occasionally fails, and surgical or, more recently, endoscopic surgical duodenojejunostomy is successfully performed. This review provides clinical information of SMA syndrome in details.

EPIDEMIOLOGY AND ETIOLOGY

SMA syndrome frequently concerns young adult female[2,7-12]. Our review found the median age is 23 years old (inter-quartile range: 16-39) but any age can be affected (range 0-91 years old), with increasing trends of elder patients with SMA syndrome in recent literatures (Figure 2). The affected age seems to be related to underlying patients' conditions (*i.e.*, congenital scoliosis at children or weight loss due to chemotherapy). Whereas the affected gender is, as reported previously, predominant in females over males with a ratio of 3:2. The incidence of SMA syndrome in the general population has been estimated at 0.013%-0.78% based on radiographic studies[7,13-16], though an

accurate prevalence of the disease is unknown, depending on under- or over-diagnosis in clinical practice and each patient's condition[10,17] (Table 1)[12,18-34]. A prospective case-control study conducted by Xu *et al*[27] found the incidence of SMA syndrome is 2.67% (26/973 admissions for 9 years). Scoliosis and burn injuries are well-known etiologies of SMA syndrome and clinicians should be aware of this entity. In contrast, in functional dyspepsia patients, the incidence is much higher (10.8%) than in the general population, which is explained by clinicians under-diagnosing.

PATHOPHYSIOLOGY

Decreasing SMA-Ao angle causes compression to the third part of the duodenum (see "DIAGNOSIS" part). Decreases in the SMA-Ao angle can be either congenital or acquired (Table 2)[2,4-6,17-19,22,25,30,33-60]. The major causes of SMA syndrome involve body weight loss and resulting loss of mesenteric fat tissue between SMA-Ao, which in turn, makes a narrower angle between the vessels. The reasons for weight loss include several types of dietary conditions (eating disorders and malabsorptive diseases), hypermetabolism (drugs and burns), and cachexia causing conditions (tuberculosis and malignancy). Especially in severe injuries and burns, prolonged bedrest in a supine position increases risk of compression of the duodenum. Scoliosis treatments (surgery and cast) and scoliosis itself are well-known causes of SMA syndrome. The lengthening of the spine during scoliosis surgery is thought to be the underlying pathophysiology. Intestinal surgeries including ileal pouch-anal anastomosis and colectomy are also well-described causes, reducing the SMA-Ao angle due to pulling on the mesentery. Congenitally short or hypertrophic ligament of Treitz is a major cause in children.

Although it is not pure SMA syndrome, aortic artery aneurysm (AAA) and surgery near or around the SMA and 3rd duodenum induces "pseudo-" SMA syndrome. AAA-related SMA syndrome was first reported by Dr. Osler as aortoduodenal syndrome[57-59].

SYMPTOMS

Patients with SMA syndrome suffer from vague and nonspecific symptoms, such as nausea, vomiting, epigastric pain, early satiety and post-prandial discomfort, bloating (abdominal distension), and weight loss, which can mimic anorexia nervosa and functional dyspepsia[8,12,61]. The epigastric pain and discomfort are more severe in a supine position and relieved in the lateral decubitus position (positioning knees to the chest) which reduces tension on the small bowel mesentery[61]. Especially in acute phase, severe duodenal obstruction leads to severe symptoms and life-threatening dilatation of the stomach[8,12,61]. In contrast, in chronic phase, the recurrent nausea and vomiting leads to inadequate food intake, resulting in severe weight loss and thus, aggravation of the syndrome[8,12,61].

COMPLICATIONS AND COMORBIDITIES

Various complications of SMA syndrome have been reported (Table 3)[35,62-86]. Notably, unrecognized or severe cases may progress to life-threatening complications, such as hypovolemic shock, aspiration pneumonia, and sudden death, even in young patients. Mechanisms of sudden death remain unclear, and several hypotheses, however, can be raised based on published cases including autopsies - arrhythmia by severe hypokalemia, severe compression of the inferior vena cava by dilated duodenum, or severe pulmonary depression induced by alkalosis and increased abdominal pressure. Thus, immediate corrections of blood election and volume and early reduction of intestinal pressure should be required in severe cases. The most frequent complication is gastrointestinal injury caused by retained or refluxed peptic acid and bile acid as well as elevated intraluminal pressure. The incidence of mucosal injury has been reported as 25%-59% in patients with SMA syndrome[62,73]. Inadequately treated or chronic mucosal injuries may progress to emphysema, necrosis, portal venous gas, and pneumoperitoneum. Elevated intraluminal pressure at the second portion of the duodenum disturbs the flow of pancreatic juice, occasionally resulting in elevated pancreatic enzymes and acute pancreatitis. Vomiting itself can increase serum amylase (mainly from salivary glands), so pancreatic amylase isozyme and lipase should help to

recognize pancreatic abnormalities. Recurrent vomiting also leads to aspiration pneumonia, dehydration, electrolyte abnormalities, and severe malnutrition. SMA syndrome sometimes co-exists with other vascular compression diseases (Table 3)[87-90]. Of these, nutcracker phenomenon is the most frequent based on anatomic location to the SMA. It is a condition that occurs when the left renal vein becomes compressed between the aorta and SMA with similar symptoms as SMA syndrome.

DIAGNOSIS

Due to its non-specific symptoms, SMA syndrome might be overlooked in clinical practice[26,91]. Even in radiologists, the duodenum seems to be a neglected segment in the intestine[92]. SMA syndrome requires a high degree of clinical suspicion and few teaching methods have been reported[93,94]. The diagnosis is based on clinical symptoms supported by radiological evidence of duodenal obstruction. Traditional criteria for SMA syndrome are based on barium X-ray studies (Figure 3): (1) Dilatation of the first and second parts of the duodenum with or without gastric dilatation; (2) abrupt vertical or oblique compression of the third part of the duodenum; (3) reverse flow of contrast proximal to the obstruction; (4) significant delay (4-6 h) in gastroduodenal transit; and (5) relief of obstruction after postural changes (the prone knee-chest or lateral decubitus position)[17,18,95]. Recently, in addition to barium studies, various imaging modalities have been used to confirm SMA syndrome, such as computed tomography (CT), abdominal ultrasound (US), magnetic resonance imaging (MRI), endoscopy and endoscopic ultrasonography (EUS), *etc.* (Table 4). Many cases are diagnosed by these modalities, which can directly visualize the SMA compression of the duodenum without barium studies. The standard diagnostic modality is CT scan (Figure 4) which allows for both diagnosis of SMA syndrome with measurement of the SMA-Ao angle and distance as well as detection of complications, such as gastric necrosis, portal vein gas, acute pancreatitis *etc.* 3D-CT is more helpful in recognizing the anatomy of SMA, the aorta and duodenum[96]. The normal SMA-Ao angle is between 38 to 65 degrees and has a distance of 10 to 33 mm[5,97]. Unal *et al*[97] reported the cutoff value is 22 degrees on

the SMA-Ao angle and 8 mm on a distance with a 42.8% sensitivity and 100% specificity. Abdominal US is another modality that provides a convenient, quick, noninvasive tool to diagnose SMA syndrome[97-99]. The sensitivity of abdominal US in diagnosis of SMA syndrome has been confirmed in a comparison study with CT findings[97]. Abdominal US benefits from an improvement of image resolution and can clearly visualize the duodenum and SMA-Ao angle (Figure 5 and Video 1). Endoscopy can detect gastrointestinal complications, such as mucosal injury, bleeding, and bezoar, *etc.* (Figures 6A and B). It can also reveal extrinsic compression (by SMA) at the 3rd portion of duodenum (Figure 6C). Further, EUS with mini-probes can be used to confirm the compression by SMA and also measure SMA-Ao distance[36,100]. Recently, linear EUS has been used for measuring the SMA-Ao angle and also endoscopic duodenojejunostomy (see TREATMENTS). Laboratory tests are not diagnostic, but they are necessary to identify the presence of electrolytic complications and pancreato-biliary abnormalities. As for differential diagnosis, almost all disorders mimicking SMA syndrome are summarized in Table 5[14,26,44,55-59,86,101-127]. Patients with eating disorders, functional dyspepsia, and peptic ulcer disease present non-specific symptoms masquerading as SMA syndrome. In addition, many diseases that potentially involve or compress duodenum should be suspected of and ruled out by CT, abdominal US, or other modalities.

TREATMENTS

The therapeutic options are summarized in Table 6. The initial treatment is usually managed conservatively by decompression of dilated stomach and duodenum by postural change and/or nasal gastric tube suction[128,129]. Positioning the patient in the left lateral or sitting position should be helpful[95,130]. However, the best position for each patient may vary because recent studies revealed there is a variation of the SMA position and movement[131-133]. In addition to gastric tube suction, intravenous metoclopramide can enhance gastrointestinal motility and help decompression[134,135]. After decompression therapy, gaining weight to increase adipose tissue between SMA

and aorta should be considered. Nasal gastric feeding is effective, but the jejunal tube is more ideal while endoscopic assistant should be considered[136]. Total parenteral nutrition is a useful option for initial nutrient treatment and if the intestinal feedings are impossible. These nutrition managements contribute to the restoration of adipose tissue to increase the angle at the origin of the SMA[28].

Surgical therapy, however, can be recommended if conservative therapies fail especially in elder patients with multiple abdominal operation histories, immobility (bed rest), long history of SMA syndrome and arteriosclerosis of SMA. Surgical therapy might be considered earlier before a patient's conditions worsen and complications occur[129]. The best timing for transition to surgical options is not clear. Shin *et al*[137] recommend 6 wk at least of conservative therapy based on the average response rate to this method. There is currently no randomized study conservative *vs* surgical therapy. A recent large cohort with 80 patients with SMA syndrome by Lee *et al*[128] revealed the overall success and recurrence rates of conservative therapy were 71.3 and 15.8%, respectively. The need for surgical therapy was 18.7% of patients (15/80 cases), which is similar to other recent cohorts 11.5%-22.2%[8,138,139]. These recent operation rates are lower than previously reported (70%) in 1974[140], likely due to advances in nutritional therapies and medications[128]. Various surgical procedures include laparoscopic, laparotomic, or robotic gastrojejunostomy, gastroduodenostomy and duodenojejunostomy, Strong's procedure (a division of the ligament of Treitz), anterior transposition of the third part of duodenum, duodenal lowering, Ladd's procedure, and transabdominal duodenojejunostomy[141-145]. Since 1998, when the first successful laparoscopic duodenojejunostomy was performed, most surgeons prefer laparoscopic duodenojejunostomy because of its safety and effectiveness (success rates: 80%-100%)[141-143]. Laparoscopic approach has been reported to shorten post-operative length of hospital stay[129,142]. Most recently, a new technique of endoscopic gastrojejunostomy (so called lumen-apposing metal stent) has also been reported in several case reports as a safe and effective therapeutic option[146-148].

CONCLUSION

In conclusion, superior mesenteric syndrome is a serious condition that requires prompt diagnosis and treatment to prevent long-term complications. Diagnosis can be challenging and may involve imaging studies, such as CT or MR, and upper gastrointestinal endoscopy. Due to the non-specific nature of clinical obstructive presentations, recognition of risk factors such as rapid weight loss, previous abdominal surgery (typically bariatric surgery), trauma or congenital anomalies can predispose patients toward the development of SMA syndrome. These conditions are typically driven by a reduction in the mesenteric fat pad or an abnormal angle between the SMA and duodenum. Early diagnosis and treatment are essential to prevent complications and ensure a successful outcome.

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