Superior Mesenteric Artery Syndrome: A Rare Cause of Intestinal Obstruction. Diagnosis and Surgical Management

Savas Yakan · Cemil Caliskan · Hasan Kaplan ·
Ali Galip Denech · Ahmet Coker

Received: 14 December 2010 / Accepted: 2 March 2012 / Published online: 15 March 2012
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Abstract Superior mesenteric artery syndrome is a rare but well-known clinical entity characterized by compression of the third or transverse portion of the duodenum against the aorta by the superior mesenteric artery, resulting in chronic, intermittent, or acute, complete or partial, duodenal obstruction. The treatment for this arteriomesenteric compression includes conservative measures and surgical intervention. The aim of the study was to evaluate our surgical management and outcomes of the patients with superior mesenteric artery syndrome. The cases with superior mesenteric artery syndrome admitted between January 2000 and January 2010 were retrospectively investigated from the patients' records. All six patients had a history of chronic abdominal pain, nausea, postprandial early satiety, vomiting, and weight loss. Diagnostic methods included barium esophagogastrroduodenography, upper gastrointestinal endoscopy, and computed tomography. Medical management was the first step of treatment in all cases before surgery. Of those, four underwent Roux-en-Y duodenojejunostomy and two underwent gastroenterostomy. Postoperative periods were uneventful and mean duration of hospitalization after the operations was 7 days. Conservative initial treatment is usually followed by surgical intervention for the main problem that is the narrowing of the aortomesenteric angle in patients with superior mesenteric artery syndrome. This syndrome should be considered in the

differential diagnosis in patients with chronic upper abdominal pain. Duodenojejunostomy is the most frequently used procedure with a high success rate.

Keywords Superior mesenteric artery syndrome · Intestinal obstruction · Diagnosis · Surgical management

Introduction

Superior mesenteric artery syndrome (SMAS) is caused by external compression of the third portion of the duodenum between the superior mesenteric artery (SMA) and the abdominal aorta. It is also known as Wilkie's syndrome, cast syndrome, and aortomesenteric syndrome [1]. It is thought that narrowing of the aortomesenteric angle results in a decrease in distance between the aorta and the SMA with consequent compression of the intervening segment of the duodenum. Normally, the third portion of the duodenum passes between the SMA and the abdominal aorta; the mean angle between the SMA and the abdominal aorta is approximately 45° (range of 38–56°). Many precipitating factors that narrow the aortomesenteric angle by approximately 6–25° can cause this syndrome [2, 3]. The true incidence of this syndrome is unknown, but it has been estimated to be approximately 0.013–0.3% [4]. Thin body habitus may predispose to SMA syndrome, but an additional cause is required for the condition to manifest. Catabolic states and malnutrition predispose to this condition by a reduction in mesenteric and retroperitoneal fat tissue, so patients with burns are at increased risk. All patients requiring prolonged bed rest following severe injury are at risk. SMA syndrome can present itself as acute small bowel obstruction or intermittent compression with chronic symptoms. The classical presentation is recurrent postprandial pain, nausea, vomiting, bloating, abdominal discomfort, or pain and tenderness.

S. Yakan (✉) · A. G. Denech
Department of Surgery, Izmir Bozyaka Education and
Research Hospital Ministry of Health,
9207 sokak No:4 Daire:2 Maliyeciler sitesi,
Karabağlar Izmir, Turkey
e-mail: savasyakan@gmail.com

C. Caliskan · H. Kaplan · A. Coker
Department of Surgery,
Ege University Faculty of Medicine,
Izmir, Turkey

We present our experience with six operated patients over the past 10 years with SMAS and discuss current literature highlights.

Methods

We performed a retrospective analysis of all medical records of patients operated with SMAS between January 2000 and January 2010. Data were extracted from our computerized database and patients' hospital records for demographics, clinical presentation, diagnostic workup, type of intervention, and outcomes.

Results

Six patients with SMAS were operated between January 2000 and January 2010. Mean age of the patients was 37 years (20–70 years). Three (50%) patients were female. Chronic epigastric pain, nausea, vomiting after meals, and weight loss were seen in all patients. The symptoms were chronic with length of symptoms beginning from 6 to 18 months (mean 12 months) prior to diagnosis. All patients had weight loss from the beginning of symptoms to diagnosis ranging from 16 to 30 kg (mean 21 kg), which is 23–43% of their body weight. The predisposing factor was severe burn in three patients, anorexia nervosa in two patients, and abdominal aortic aneurysm in one patient. Endoscopy was performed in all patients. Nonspecific findings of reflux esophagitis were described, and in only two patients a possible obstruction of the third part of the duodenum was suspected. All patients had either formal barium meal (Ba meal) studies (Fig. 1) or computerized abdominal tomography (CT) scans with oral contrast (Fig. 2), which showed a dilated stomach with an obstructive lesion in the third part of the duodenum. CT angiography was performed in all patients to assess the aortosuperior mesenteric artery angle (Fig. 3). The angle ranged from 11° to 21° with a mean of 17° (normal 38° – 56°). Although there was a prominent weight loss, there was no laboratory abnormality other than mild electrolyte imbalance in two patients. Nutritional support was the first-line therapy for all patients. Patients were directed to have multiple small frequent meals as much as six to eight times, and total parenteral nutrition was started in addition to a peristalsis-stimulating agent such as metoclopramide. Two burn patients benefiting from conservative methods were excluded from the study, whereas six patients were treated surgically due to insufficiency of conservative measures. Surgical treatment was duodenojejunostomy in four (66%) and gastroenterostomy in two (34%) patients. Preoperative underlying etiology was evident in all patients, so any histopathological diagnosis was not attempted. There was no postoperative complication, and patients well tolerated oral

Fig. 1 Barium meal showing distended stomach and dilated duodenum (150-min appearance)

feeding started at the third postoperative day. Mean duration of hospitalization was 7 (5–9) days. All patients had good clinical outcomes, weight gaining between 8 and 17 kg in 6 months.

Fig. 2 The third portion of the duodenum is compressed at the aortomesenteric angle on cross-sectional computed tomographic views (D, duodenum; A, aorta; SMA, superior mesenteric artery)

Fig. 3 Sagittal reconstruction of CT angiography demonstrated an aortomesenteric angle of 15°

There was no additional problem during the follow-up period (2 months to 6 years, mean 4 years), and none of the patients required reoperation. The patient with aortic aneurysm was followed together by the cardiovascular surgery department. Details of the patients' features, diagnostic workup, and surgical procedures are shown in Table 1.

Discussion

SMA syndrome was first described by Rokitsansky in 1842. Wilkie published the first series in 1927; therefore, SMA syndrome is also called Wilkie's syndrome [5]. Cast syndrome, arteriomesentric duodenal compression, aortomesentric artery compression, and duodenal vascular compression are the other terms used to describe the same condition. Structural and acquired factors are offered in etiology. Thin body habitus, advanced lumbar lordosis, previous trauma, spinal deformity or trauma, diseases like anorexia nervosa or malabsorption, anatomical abnormalities like insufficient rotation of duodenum, superior and short Trietz ligament, SMA originating inferiorly than normal or SMA aneurysm are some of these factors. Initial factors are weight loss causing loss of mesenteric and retroperitoneal fat tissue due to catabolic states like cancer or burns, traumas causing severe damage, anorexia nervosa, or malabsorptive states [6–9]. In our series, three patients had weight loss and prolonged bed rest due to severe burn, and two patients had anorexia nervosa. In another patient, abdominal aortic aneurysm was diagnosed by imaging techniques and blamed for SMAS by causing a decrease in SMA/aorta angle. Females and young adults (18–35 years) are more likely to be affected by the condition, though it can occur at any age [7, 9]. This age and sex distribution may be reflecting the predisposing cause of the condition and, in particular, eating disorders.

Symptomatology varies between acute onset to a gradually progressive course of early satiety and postprandial abdominal pain. Typical features include pain aggravated by a supine position and relieved by prone or left lateral decubitus, or even by a knee–chest position. Other symptoms include bilious

Table 1 Details of the patients' features, diagnostic workup, and surgical procedures

Number of cases	Age (years)	Sex	Symptoms	Etiological factor	Preoperative diagnostic workup	Aortosuperior mesenteric artery angle (°)	Surgical procedure
1	70	Male	Postprandial pain, nausea, emesis	Abdominal aortic aneurysm	UGIE, UGIB, CT	11	Retrocolic gastroenterostomy
2	39	Male	Early satiety, bloating, emesis	Severe burn	UGIE, UGIB, CT	21	Roux-en-Y duodenojejunostomy
3	23	Female	Postprandial nausea, emesis, weight loss	Anorexia nervosa	UGIE, UGIB, CT	18	Roux-en-Y duodenojejunostomy
4	21	Female	Postprandial nausea, emesis, weight loss	Severe burn	UGIE, UGIB, CT	17	Roux-en-Y duodenojejunostomy
5	50	Male	Postprandial pain, nausea, emesis	Severe burn	UGIE, UGIB, CT	15	Retrocolic gastroenterostomy
6	20	Female	Postprandial pain, early satiety, bloating, emesis	Anorexia nervosa	UGIE, UGIB, CT	19	Roux-en-Y duodenojejunostomy

UGIE, upper gastrointestinal endoscopy; UGIB, upper gastrointestinal barium series; CT, contrast-enhanced abdominal computed tomography

vomiting with loss of weight and appetite, which can occur with advanced obstruction [10]. These patients might present with manifestations of electrolyte imbalance or with gastrointestinal obstructive complications including massive gastric dilatation with gastric wall pneumatosis and portal venous gas [11]. Physical examination is nonspecific, with findings including a thin body build, abdominal distension, and signs of intestinal obstruction.

Patients with a history of symptoms suggesting SMAS should undergo further radiographic studies to establish the diagnosis. Upper gastrointestinal series, computed tomography (CT) scan or CT angiography, magnetic resonance (MR) angiography, conventional angiography, ultrasonography, and endoscopy have all been used for diagnosis [1]. The clinical diagnosis can be confirmed by radiologic studies in 95% cases [12]. The following radiologic criteria have been established for the diagnosis of SMA: (i) dilatation of the first and second portions of the duodenum, with or without gastric dilatation; (ii) abrupt vertical and oblique compression of the mucosal folds; (iii) antiperistaltic flow of barium proximal to the obstruction; (iv) delay of 4–6 h in transit through the gastroduodenal region; and (v) relief of obstruction in a prone, knee–chest, or left lateral decubitus position.

CT is also useful in demonstrating duodenal distention, the anatomy, and the relationships of the superior mesenteric vessels and excluding other pathologies such as tumor. By the use of selective SMA angiography against a contrast-filled duodenum, it is possible to demonstrate extrinsic compression and to measure the aortomesenteric angle and the distance from the aorta.

Therapeutic options for SMAS are conservative management or surgical bypass of the obstruction. The goal of medical therapy is to induce weight gain, which would presumably result in an increase in fat at the mesenteric root. Left lateral decubitus, knee–chest, or prone positioning decreases the mesenteric pressure on the duodenum and allows temporary alleviation of the duodenal obstruction. The patient may then be able to tolerate frequent small meals to increase caloric intake. Alternatively, enteral nutrition can be provided by a nasogastric feeding tube positioned distal to the ligament of Treitz, or total parenteral nutrition can be provided for patients with contraindications to enteral nutrition [6, 13, 14]. Failure of the treatment, although a certain time period can not be determined, is decided with vomiting as the chief symptom and recurrence of the other symptoms, and constitutes indication of surgical treatment for SMAS [6, 14].

Conventional open surgical techniques or laparoscopic techniques can be used. Several surgical procedures including gastroduodenostomy, duodenojejunostomy, and Strong's operation (duodenal mobilization for distal duodenojejunal flexure) have been performed to resolve or bypass duodenal compression [1]. A duodenojejunostomy remains the operation of choice to relieve the obstruction, with success rates

up to 90% [4], though it may cause blind loop syndrome. Gastroduodenostomy has been shown to provide adequate gastric decompression but failed to completely release duodenal obstruction, leading to persistence of symptoms that necessitated duodenojejunostomy in some cases [15]. Persisting obstruction may lead to blind loop syndrome, gastric bile reflux, and ulceration. Strong's procedure has the advantage of maintaining bowel integrity, and thus it is the less invasive, quicker, and safer procedure. It has been correlated with an earlier postoperative recovery [1]. The disadvantages are that the procedure can be aggravated or impossible due to adhesions and that caudal displacement of the duodenum cannot always be achieved because of interference with short vessels from the inferior pancreaticoduodenal artery to the duodenum [15]. With the advancement in laparoscopic surgery, laparoscopic Strong's procedure and duodenojejunostomy became applicable in a number of hospitals. In our series, duodenojejunostomy was the operation performed in four patients and retrocolic gastroenterostomy in two patients, and none of them was laparoscopic.

Conclusion

SMAS is a rare cause of proximal small bowel obstruction. The diagnostic workup includes abdominal CT scan, upper gastrointestinal endoscopy, upper gastrointestinal barium series, CT angiography, and conventional angiography, if necessary. Whatever the underlying condition is, duodenal obstruction is a mechanical problem and physicians should not insist on conservative treatment if symptoms persist, and surgical treatment should be applied in a timely manner with considering underlying etiology.

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