Recalling Superior Mesenteric Artery Syndrome

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Key Words
Superior mesenteric artery syndrome • Arteriomesenteric duodenal compression • Wilkie's syndrome • Small bowel obstruction • Strong's procedure

Abstract
Background: Superior mesenteric artery syndrome is uncommon and characterized by postprandial epigastric pain, nausea, vomiting, anorexia and weight loss. The syndrome is caused by compression of the third part of the duodenum in the angle between the aorta and the superior mesenteric artery. This review updates etiology, epidemiology, diagnosis, treatment and outcome of the superior mesenteric artery syndrome.

Methods: Review of the literature.

Results: Frequently, predisposing medical conditions associated with catabolic states or rapid weight loss result in a decrease of the aortomesenteric angle and subsequent duodenal obstruction. External cast compression, anatomic variants and surgical alteration of the anatomy following spine surgery or ileoanal pouch anastomosis can also precipitate the syndrome. Once radiologic studies have established diagnosis, first-line treatment is usually conservative with jejunal or parenteral nutrition for restoration of the aortomesenteric fatty tissue. If conservative management fails, surgical options include open or laparoscopic duodenojejunostomy or duodenal mobilization and division of the ligament of Treitz.

Conclusion: Superior mesenteric artery syndrome is clearly defined and frequently associated with a wide range of predisposing conditions and surgical procedures; clinicians have to consider this syndrome in such a setting. Larger studies are needed to better define the optimal treatment for this disease.

Introduction

The superior mesenteric artery syndrome is a rare medical condition that describes the clinical symptoms resulting from vascular compression of the third part of the duodenum in the angle between the aorta and the superior mesenteric artery. The complex of clinical signs reflecting duodenal obstruction includes postprandial epigastric pain, nausea, vomiting, anorexia and weight loss and has been termed aortomesenteric artery compression [1], arteriomesenteric duodenal compression [2], duodenal vascular compression [3], superior mesenteric artery [4–6], Wilkie’s [7, 8] or cast syndrome [9]. In contrast, megaduodenum characterizes a dilatation of the duodenum without primary or secondary duodenal stenosis which has been ascribed to intestinal myopathy and which can be familial [10–12]. In fact, the existence of the superior mesenteric artery syndrome has been con-
troversial because of confusion with other causes of megaduodenum [13].

The entity was first described by the Austrian professor Carl von Rokitansky in his anatomy textbook in 1842 [14]. Since then, a number of case reports and reviews appeared and discussed the syndrome and its treatment [15]. Subsequently, Wilkie [16] published the first comprehensive series of 75 patients in 1927 and his name has become a common eponym for the superior mesenteric artery syndrome. By the year 1989, more than 400 patients with the syndrome had been reported [4].

Today, a PubMed search (www.ncbi.nlm.nih.gov/ PubMed, 1950 to September 2006) using a combination of the MeSH term ‘superior mesenteric artery syndrome’ with additional text words (Wilkie’s syndrome, aortomesenteric compression, arteriomesenteric duodenal compression, duodenal vascular compression, excluding biliary cast syndrome) yielded more than 330 articles including case reports, original articles and reviews. However, still most clinicians are not aware of this disease and patients are often diagnosed only after a long history of abdominal complaints. Thus, recent cases were presented as rare clinical images with educational value to recall the disease entity [7, 17, 18].

Surgical progress has brought along new operative options (e.g. laparoscopic repair) but also additional precipitating factors (e.g. bariatric surgery, ileoanal pouch surgery). A recent case of superior mesenteric artery syndrome at the Department of Surgery, University of Heidelberg, stimulated the authors to review the literature based on a PubMed search. Reference lists of the retrieved literature were manually cross-searched for additional publications. It was noticed that most of the comprehensive studies and large series dated back to 1960–1980.

This review summarizes current etiology, anatomy and epidemiology, clinical and radiographic signs, treatment and outcome of the superior mesenteric artery syndrome as reported in the literature. It further provides a case report of a superior mesenteric artery syndrome following ileoanal pouch anastomosis.

### Anatomy and Etiology

The superior mesenteric artery syndrome is caused by a low aortomesenteric angle resulting in vascular compression of the third part of the duodenum which is relatively immobile and crossed by the mesenteric root (fig. 1). The superior mesenteric artery originates behind the neck of the pancreas at the level of the first lumbar vertebra and leaves the aorta at an acute angle (fig. 2). The left renal vein crossing the vertebral column, the uncinate process of the pancreas and lymphatics are embedded in retroperitoneal fat tissue sustaining the physiologic aortomesenteric angle. In parallel to compression of the duodenum, compression of the left renal vein in the aortomesenteric angle associated with venous hypertension is known as the ‘nutcracker syndrome’ [19]. The mean angle formed by the superior mesenteric artery and the aorta varies between 38 and 56° but angles ranged from 20 to 70° in an analysis of 64 human aortas [20–22]. The mean radiographic aortomesenteric distance was 10–28 mm [23].

Progression from irrelevant compression to complete obstruction is related to a number of factors that diminish the aortomesenteric angle to approximately 6–16° and the aortomesenteric distance to about 2–8 mm [5, 22, 23]. Although a thin, asthenic habitus may predispose an individual to the syndrome, an additional stimulus is probably necessary for manifestation [8]. It has been observed that the superior mesenteric artery syndrome is more likely precipitated by an acute change in patient status that complicates a chronic debilitating disease [5]. Rapid weight loss and certain metabolic states lead to a depletion or loss of mesenteric and retroperitoneal fat and subsequent decrease of the aortomesenteric distance. In ad-
dition, anatomical variants like a short or a high insertion of the ligament of Treitz at the duodenojejunal flexure can result in a dislocation of the duodenum to a more cranial position into the vascular angle. It was hypothesized that hypertrophy of the suspensory muscle of Treitz is responsible for the cranial dislocation [24]. However, muscle hypertrophy in the ligament of Treitz could not be confirmed on biopsy [25].

Numerous predisposing conditions for the superior mesenteric artery syndrome with potential impact on the aortomesenteric angle have been identified and can be summarized into the following three categories: severe weight loss in catabolic states, external (e.g. belts and body spica casts) and intra-abdominal compression or mesenteric tension (table 1).

In addition, surgical alterations or correction of the anatomy with relationship to the structures forming the aortomesenteric angle constitute risk factors. Tension and a caudal pull of the small bowel mesentery after ileoanal pouch anastomosis can diminish the aortomesenteric angle resulting in duodenal compression (see presented case). The superior mesenteric artery syndrome is further a well-known complication following scoliosis surgery (see also table 1) due to a relative lengthening of the spine postoperatively. Asthenic habitus, sagittal kyphosis and postoperative weight loss have been identified as specific risk factors for development of superior mesenteric artery syndrome after scoliosis surgery [42, 43, 45].

The frequent association of superior mesenteric artery syndrome with psychiatric disorders like anorexia nervosa raised the question long ago whether anorexia nervosa is the cause or complication of the superior mesenteric artery syndrome [49].

**Epidemiology**

Case reports of superior mesenteric artery syndrome most frequently concern patients following spine or scoliosis surgery. The prevalence of superior mesenteric artery syndrome after scoliosis surgery has been reported to range from 0.5 to 2.4% [42, 43, 50]. In comparison, of 1,687 severely burned patients admitted from 1966 to 1970 at the Brooke Army Medical Center in Texas, 19
cases with superior mesenteric artery syndrome (1.1%) were documented [31]. The mean burned area was 48.5% of the total body surface (range 32–64.5%) which implies an extreme catabolic condition.

In the general population, the prevalence of superior mesenteric artery syndrome was estimated at around 0.013–0.3% on the basis of upper gastrointestinal barium studies [4, 51, 52], underlining its rarity. Interestingly, the prevalence in a chronic-care hospital (0.965/1,000 admissions) was significantly higher than that in acute general hospitals (0.0108–0.0520/1,000 admissions) [6]. However, superior mesenteric artery syndrome might be overdiagnosed and a strict definition with precise clinical and radiographic criteria for diagnosis may substantially reduce reported prevalence rates. Hines et al. [5] confirmed only 14.6% of 44 patients that had been diagnosed with superior mesenteric artery syndrome after critically reviewing the cases according to strict radiographic signs (see Diagnosis).

Overall, females are more commonly affected by the syndrome and two thirds of patients are in the age group between 10 and 39 years [4, 16, 27, 32, 53, 54]. The affected age is of course closely linked to the underlying predisposing condition (e.g. scoliosis surgery at children or adolescents’ age) but the syndrome has also been described in an 84- and an 86-year-old man [18, 29].

**Symptoms**

Patients predominantly present with a history of chronic abdominal complaints with intermittent exacerbations depending on the cause and grade of duodenal compression. In rare cases, the syndrome may be acute with rapid evolving upper intestinal ileus [34, 55]. Whereas patients that underwent scoliosis surgery often show symptoms within the first postoperative week [27, 42, 43] and burn patients after 2 weeks [31], other patients suffer from non-specific symptoms for months [10, 56] to years [1, 7, 8, 17].

The most typical and most frequent clinical finding is intermittent or postprandial abdominal pain (59–81%) followed by vomiting, nausea and anorexia resulting in weight loss [4, 27]. At diagnosis, most patients are underweight with calculated mean weights of 48.3 (15 patients [6]) and 52 kg (16 patients [4]) in patients older than 15 (range 16–71) years. The pain is often described as epigastric and is characteristically relieved by a prone, knee-chest or left lateral decubitus position that all reduce small bowel mesentery tension at the aortomesenteric angle [5]. It has been well established that duodenal obstruction may lead to retrograde duodenal stasis, dilatation of the proximal duodenum and gastric reflux with occasional bile emesis and with an association of peptic ulcer disease in up to 15–25% [2, 4, 16, 18, 46, 57]. In a series of patients with major burns, 5 of 19 patients (26%) with superior mesenteric artery syndrome developed duodenal ulcerations [31].

**Diagnosis**

Patients that present with a history of symptoms suggesting superior mesenteric artery syndrome 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 [21, 23, 26, 47, 54, 58, 59]. Conventional barium studies still play an important role for diagnosis and the classic but unspecific picture is that of a dilated proximal duodenum with an abrupt termination of the barium column in the third portion. This radiographic appearance might also be seen in cases of megaduodenum (intestinal pseudo-obstruction) [10]. The following strict radiographic criteria have been established for diagnosis of the superior mesenteric artery syndrome: (i) dilatation of the first and second parts of the duodenum, with or without gastric dilatation, (ii) abrupt vertical and oblique compression of the mucosal folds, (iii) antiperistaltic flow of contrast medium proximal to the obstruction, (iv) delay in transit of 4–6 h through the gastroduodenal region, and (v) relief of obstruction in a prone, knee-chest or left lateral decubitus position [1, 5].

Contrast-enhanced CT scan additionally demonstrates the aortomesenteric angle, distance and fat tissue, obstruction of the duodenum and a potential culprit for compression, e.g. local neoplasia or an aneurysm. Contrast-enhanced CT scan and MR angiography seem to be equivalent in evaluating the exact angle and distance [26]. In the past, angiography has been suggested as the ‘gold standard’ procedure for assessment of the aortomesenteric angle and distance [54, 60]. An aortomesenteric angle of <22–25° and a distance of <8 mm correlated well with symptoms of superior mesenteric artery syndrome [23, 58]. Unal et al. [58] reported cutoff values of 22° (42.8% sensitivity and 100% specificity) for the aortomesenteric angle and 8 mm (100% sensitivity and specificity) for at least one symptom of the superior mesenteric artery syndrome.
Recently, the use of ultrasound power color Doppler imaging was advocated for detection of a reduced aortomesenteric angle in suspected cases [23]. Because of superior information content and non-invasiveness, contrast-enhanced CT or MR angiography is more valuable if the cause for the superior mesenteric artery syndrome is unclear. In addition, upper gastrointestinal endoscopy should be performed to rule out intestinal intraluminal obstruction and gastric or duodenal ulcer disease that might be secondary to reflux or that might constitute a primary pathology mimicking superior mesenteric artery syndrome.

**Treatment and Outcome**

In absence of displacement by an abdominal mass, an aneurysm or another pathologic condition that requires immediate surgical exploration, the treatment of the superior mesenteric artery syndrome usually begins with conservative approaches.

Nasogastric tube placement for duodenal and gastric decompression and mobilization into the prone or left lateral decubitus position often is effective in the acute setting [1]. Acute superior mesenteric artery syndrome with a history of a month and less seem to be more apt to conservative treatment [21, 30]. Medical treatment pursues correction of the fluid and electrolyte balance, a positive nitrogen balance and an increase in body weight promoting restoration of the retroperitoneal fat tissue with consecutive increase of the aortomesenteric angle. Both enteral jejunal tube feeding and parenteral nutrition have been successful [1, 30]. Patients seem to benefit from nutritional support with hyperalimentation irrespective of the disease history [26].

Advances in both enteral and parenteral nutrition in the last years have led to a substantial shift towards medical treatment. In the comparison of two similar pediatric cohorts that were published in 1974 and in 2006, the need for surgical treatment decreased from 70 to 14% [27].

Surgery is indicated in symptomatic patients when conservative treatment fails. There is no clear time limit for duration of medical treatment, since relief of symptoms has been observed from 2 to 12 days, but treatment of up to 169 days has also been reported [6, 27, 42]. Duodenal atony after massive dilatation may persist even after duodenal decompression and delay normal gastrointestinal function.

Several surgical procedures including gastrojejunostomy, duodenojejunostomy and Strong’s operation (duodenal mobilization for lowering the duodenojejunal flexure) have been performed to resolve or bypass duodenal compression [6].

The first duodenojejunostomy for the superior mesenteric artery syndrome was done by Stavely in 1908 and has become the most frequent surgical procedure with a success rate of about 80% [53, 61]. Lee and Mangla [6] concluded after reviewing 146 cases operated after 1963 that duodenojejunostomy revealed the best results in severe cases and was significantly better compared to gastrojejunostomy and Strong’s procedure.

Gastrojejunostomy 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 [6]. Persisting obstruction may lead to blind loop syndrome, gastric bile reflux and ulceration.

Strong [62] 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 that it does not violate the bowel and thus is the less invasive, quicker and safer procedure. It has been correlated with an earlier postoperative recovery [22, 63]. 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 [6, 21].

Laparoscopic techniques of Strong’s or bypass procedures have been propagated during the last 10 years. In 1995, Massoud [64] reported his experience after laparoscopic division of the ligament of Treitz in 4 cases which was successful in 3 cases. Gersin and Heniford [65] presented the first laparoscopic duodenojejunostomy in a female with superior mesenteric artery syndrome in 1998. The feasibility of laparoscopic duodenojejunostomy has been confirmed in 2 more uncomplicated cases [66]. Here, a retrocolic duodenal bypass was performed with an endoscopic gastrointestinal anastomotic stapler in 152 and 75 min and the hospital stay was 4 and 2 days, respectively.

Positive response to medical treatment as documented in larger series has been observed in 83% (38/46) [6] and in 86% (19/20) of pediatric cases [27]. Most of the patients with superior mesenteric artery syndrome following scoliosis surgery also recovered under conservative therapy [42, 53].

Many of the published surgical cases were first treated medically and then only after treatment failure underwent surgery. Good results after surgical duodenojejunostomy
were achieved in 79% (161 patients, studied by Barner and Sherman [53] in 1963), 90% (50 cases reviewed by Lee and Mangla [6] in 1978) or even 100% of the operated patients (7 cases reported by Lee and Mangla [6] in 1978).

Case

A 28-year-old female patient complained of intermittent, postprandial epigastric pain with anorexia and weight loss of 6 kg over 8 months. On admission her body weight and height were 52 kg and 153 cm, respectively. Pain intensity was aggravated or alleviated in certain positions. She had undergone proctocolectomy with ileoanal pouch anastomosis because of familial adenomatous polyposis at the age of 21. MR imaging demonstrated moderate duodenal dilatation with compression of the superior mesenteric artery consistent with superior mesenteric artery syndrome (fig. 3). Conservative management failed to achieve improvement of quality of life with persistent symptoms and the patient underwent explorative laparotomy with mobilization of the duodenum (Strong’s procedure). The postoperative course was uneventful with quick recovery and the patient was discharged on postoperative day 6. Six weeks later, she reported a significant improvement of her symptoms with continuous weight gain and absence of anorexia since surgery.

Conclusion

Superior mesenteric artery syndrome is clearly defined, uncommon, but frequently associated with a wide range of predisposing conditions and surgical procedures. Clinicians have to consider this syndrome in such a setting in patients presenting with postprandial abdominal pain, anorexia, vomiting, or weight loss. Ultrasound findings of an aortomesenteric angle of <25° or an aortomesenteric distance of <8 mm support the diagnosis. CT or MR scans can establish definite diagnosis and can further provide information about the underlying cause giving the basis for appropriate treatment. The results reported in the literature advocate conservative management as first-line treatment to restore the correct aortomesenteric angle resulting in duodenal decompression. Advances in diagnosis, nutritional and other medical treatment options have substantially improved the success rate of conservative treatment over the last decades.

Today, surgery has a role when medical treatment fails or if the underlying cause requires surgical exploration itself. The data available in the literature does not provide enough statistical evidence to identify any surgical procedure as superior. Due to the paucity of the superior mesenteric artery syndrome, randomized controlled trials are not available. Although duodenojejunostomy has proved to be the superior surgical procedure in the past [6], Strong’s procedure is less invasive and might be as effective in selected cases. Both laparoscopic duodenojejunostomy and duodenal mobilization (Strong’s procedure) have the potential to reduce invasiveness and improve recovery of surgical treatment in the future. Since most data on large series of superior mesenteric artery syndrome were published over 30 years ago, new studies of larger series will demonstrate if current medical progress influences prevalence, diagnosis and improves therapeutic outcome.

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References

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