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Superior Mesenteric Artery Syndrome: Diagnosis and Treatment Strategies

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Abstract
Introduction Superior mesenteric artery (SMA) syndrome is an unusual cause of vomiting and weight loss resulting from the compression of the third part of the duodenum by the SMA. Various medical and psychiatric conditions may result in the initial rapid weight loss which causes narrowing of the aortomesenteric angle. The vomiting and obstructive syndrome is then self-perpetuated regardless of the initiating factors. The young age and nonspecific symptoms often lead to a delay in diagnosis.
Discussion A series of eight cases is presented reviewing the presentation, investigations, surgical treatment by division of duodenum and duodenojejunostomy, and outcomes.
Conclusion SMA syndrome is a well-described entity which must be considered as a cause of vomiting associated with significant weight loss in young adults. Surgical treatment should be allied with psychological assessment to treat any underlying psychosocial abnormality.

Keywords Superior mesenteric artery syndrome · Cast syndrome · Wilkie's syndrome · Duodenal obstruction · Strong's procedure · Laparoscopy · Duodenojejunal bypass

Introduction

Duodenal outlet obstruction may result from variations in the anatomical relationship between the aorta, mesenteric vessels, and duodenum. In young and otherwise healthy patients presenting with abdominal pain, nausea, anorexia, weight loss, and vomiting, the diagnosis of superior mesenteric artery (SMA) syndrome should be considered. SMA syndrome differs from conditions with similar symptoms including familial neuropathic diseases, such as megaduodenum, in that it is a true obstructive condition without any underlying myopathy. In recent years, there have been numerous case reports of this condition, but in spite of this, diagnosis of this condition is frequently delayed resulting in ineffective symptomatic therapies and inappropriate investigations. This manuscript presents a series of eight SMA syndrome patients treated at a single institution being the largest series reported in over 30 years and a literature review to raise awareness of the condition and try and redress this problem of late diagnosis.

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Patients and Methods

From January 2002 to March 2007, eight patients presented to the Upper Gastrointestinal Surgical Unit (UGI) of Bankstown and Liverpool Hospitals. The unit is a tertiary referral center with a local drainage population of 1.5 million, and two of the patients were referred from outside the normal drainage area. A prospective collated database was then retrospectively reviewed for demographic data, clinical presentation, diagnostic workup, treatment, and outcomes. Long-term follow-up was obtained from patient review for all patients.

Eight patients (one male/seven females) presented over the period (Table 1). Mean age was 27 years (range 18–32 years).

Table 1 Patient Characteristics

Patient	Age/sex	Comorbidities	Symptoms	Length symptoms (months)	Weight loss (kg)	Preoperative weight (kg)	Aortosuperior mesenteric artery angle (°)	Other radiology	Postoperative weight gain (6 months) (kg)
1	32/M	Drug use	Vomiting reflux	24	25	49	17	Ba meal: dilated to D3	9
2	18/F	Binge ETOH	Vomiting	8	30	45	12	Ba meal: obstructed D3	12
3	22/F	Intentional dieting	Vomiting	12	23	48	14	Ba meal: dilated stomach	15
4	28/F	Nil	Vomiting/reflux	18	22	42	18	CT: dilated to D4	10
5	32/F	Pancreatitis	Vomiting/reflux/ulcer/electrolyte abnormality	24	40	32	9	Ba meal: obstructed D3	10
6	32/F	Obstructive defecation	Vomiting/reflux/ulcer/electrolyte abnormality	18	30	35	15	CT dilated to D3	12
7	31/F	Domestic abuse/ social issues	Vomiting/reflux	28	32	38	12	CT dilated to D3	15
8	23/F	Drug abuse	Vomiting and reflux	18	20	35	17	CT dilated to D3	5

One patient had a previous laparoscopic cholecystectomy but no other patient had a history of abdominal surgery.

Presentation consistently involved persistent chronic vomiting, esophageal reflux, and epigastric pain. In six patients, this was associated with severe reflux, and in two patients, significant electrolyte abnormalities, particularly hypokalemia requiring intravenous replacement. The symptoms were chronic with length of symptoms ranging from between 8 to 28 months (mean 18 months) prior to diagnosis. All patients had sought medical advice during this period and had specialist gastroenterologist review and endoscopies. All had been diagnosed with reflux and treated with proton pump inhibitors. In all cases, there was a prolonged period of medical review, investigation, and treatment from the development of symptoms to final diagnosis.

Confounding the ability to accurately diagnose the condition were comorbidities including psychosocial conditions. These included drug and alcohol abuse, domestic violence, eating disorders as well other illnesses such as pancreatitis.

All patients had extreme weight loss from the development of symptoms ranging from 20 to 40 kg (mean 29 kg) with the weight loss ranging from 33% to 55% of their body weight.

Endoscopies were performed in all patients. Nonspecific findings of reflux esophagitis were described with only one endoscopist suspecting a possible obstruction of the third part of the duodenum (D3).

Suspicion of SMA syndrome occurred late in the disease course and only after the patients had a second opinion with the UGI surgical team. All patients then underwent diagnostic imaging aimed at confirming the suspected diagnosis. All patients had either formal Barium meal (Ba meal) studies (Fig. 1) or computerized axial tomography (CT) scans with oral contrast (Fig. 2) which showed a dilated stomach and duodenum with an obstructive lesion in D3.

In spite of the marked weight loss, all patients had serum albumin and serum proteins within normal limits. Two patients had electrolyte abnormalities as previously noted, and in one of these patients, we elected to give 3 weeks of total parental nutrition (TPN) prior to surgery

CT angiography was performed on all patients to assess the aortosuperior mesenteric artery angle (Figs. 3 and 4). The angle ranged from 9° to 18° with a mean of 12° (normal 38–68°). Scans also indicated reduction of the aortomesenteric distance with compression of the left renal vein seen in one case (Fig. 4).

Hence, all patients met the diagnostic criteria of SMA syndrome with radiological evidence of obstruction of D3, reduced aortosuperior mesenteric artery angle, and reduced aortomesenteric space.

Figure 1 Ba meal showing dilated stomach and cut off at third part of duodenum.

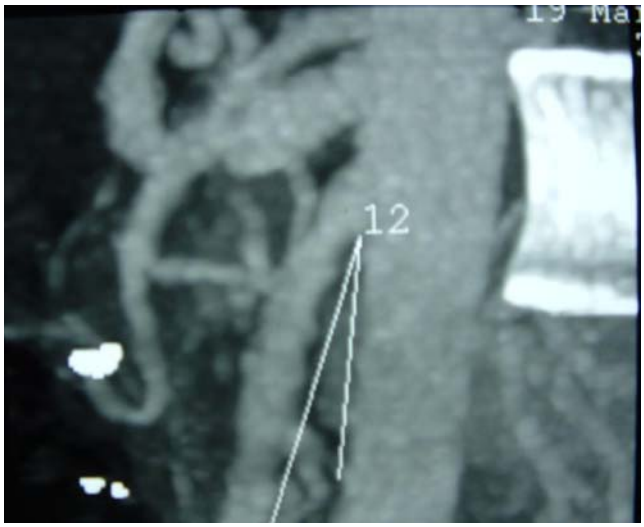

Figure 3 CT angiography showing aortoduodenal angle.

All patients were treated with mobilization and division of the fourth part of the duodenum with the end portion of the jejunum then being placed through the avascular portion of the right mesocolon and a side to side anastomosis between the third part of the duodenum and jejunum. All patients had uneventful postoperative recoveries. All patients had oral nutrition reinstated after 4 days. There were no wound infections or anastomotic breakdowns and the average length of stay was 10 days (range 7–14 days).

All patients had an excellent clinical outcome gaining between 5 and 15 kg at 6 months review. Postoperative imaging with Ba meal studies and endoscopy showed good emptying from the stomach and duodenum and no evidence of further obstruction. The patient's weight gain stabilized at the 6-month period with no patient recording any major weight gain after this period. One patient developed an incisional hernia for which she has since had a laparoscopic repair. Three of the patients continued to require long-term psychological support for their underlying psychological disorders and one patient committed suicide at 9 months. All other patients remain well (follow-up range 5–68 months).

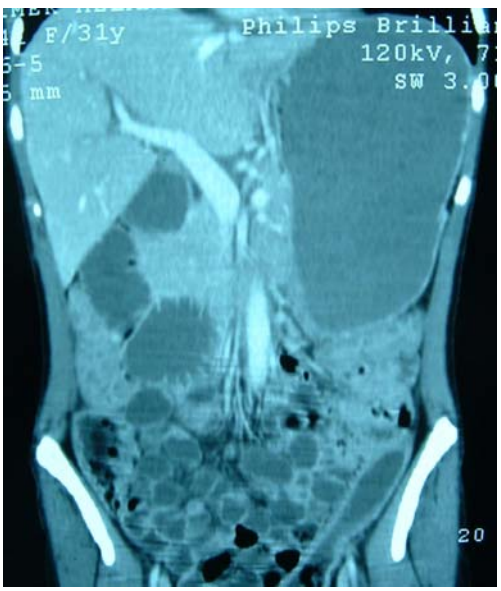

Figure 2 CT scan showing dilated stomach and duodenum with obstruction by SMA.

Discussion

The SMA syndrome was first described in 1842 in an anatomy text by Rokitansky.[1] It involves the entrapment and obstruction of the third part of the duodenum between

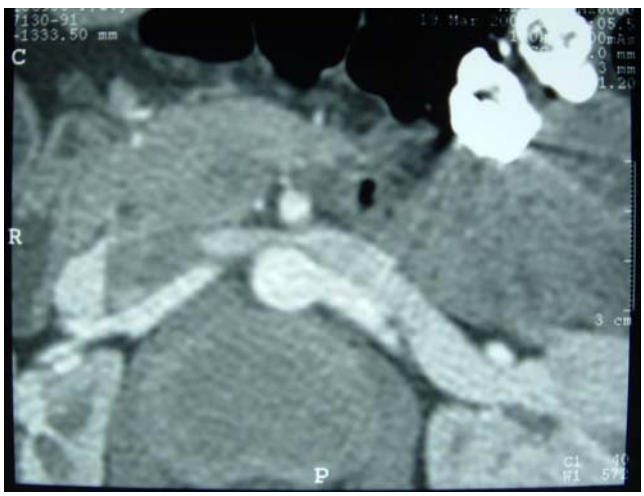

Figure 4 CT showing reduction of aortoduodenal distance with compression of left renal vein.

the SMA and the aorta. Wilkie published the first large series of 75 patients in 1927 and his name is frequently ascribed to this condition.[2] The majority of reports of this condition are limited to case reports and small series.

The main anatomical feature of this syndrome is the narrowing of the angle between the SMA and aorta, normally 38° to 65°.[3] The angles in our series were between 9° and 22°. This narrowing results in the compression of the third part of the duodenum as it crosses between the aorta and the SMA and may even result in compression of the left renal vein. The aortomesenteric distance is reduced from the normal 10–28 to 2–8 mm.[4]

The cause of this narrowing, rather than a congenital abnormality, is postulated to be related to loss of intra-abdominal adipose tissue. In the normal individual, this adipose tissue displaces the SMA anteriorly away from the aorta so avoiding duodenal compression by increasing the space for the duodenum to pass. Loss of this intraabdominal fat narrows the aortosuperior mesenteric artery angle resulting in functional obstruction. In our series, all patients had been asymptomatic until an initial significant weight loss, either related to intentional dieting or illness, followed then by a continuation and worsening of symptoms. A similar etiology is seen in patients who develop this condition following scoliosis and other surgery,[5] the condition developing from alteration of the relationship of the structure which forms the aortosuperior mesenteric artery angle with the relative lengthening of the spine following surgery increasing the tension on the mesentery and narrowing this angle.

This syndrome has been described in patients falling into two broad categories: those who develop it following surgery or compression and the second associated with severe weight loss. This first group includes those following corrective spinal surgery which traditionally has been the most frequently described cause.[6] The second includes wasting conditions such as AIDS,[6] malabsorption, cancer,[7] cerebral palsy, and other conditions associated with cachexia; catabolic conditions such burns;[8] and with eating disorders such as anorexia nervosa and drug abuse.[9,10] Similarly, it has been described following surgery-associated rapid weight loss such as bariatric surgery, esophagectomy, and abdominal trauma.[11,12] Regardless of the etiology, once the condition has become established, it becomes self-perpetuating with a cycle of vomiting leading to further weight loss and thus further vomiting.

Females and young adults (18–35 years) are more likely to be affected by the condition, though it can occur at any age.[2,7,13] This age and sex distribution may simply reflect the predisposing cause of the condition and, in particular, eating disorders.

The classical features include chronic food intolerance with nausea and vomiting, weight loss, and epigastric pain. The pain is classically described as being relieved by lying prone or in the left lateral decubitus position, maneuvers which release tension on the small bowel mesentery, and thus releasing the aortomesenteric angle. The patients are usually significantly underweight at the time of diagnosis. Patients may also complain of severe reflux and endoscopy may demonstrate severe esophagitis and gastritis associated with stasis and chronic obstruction. It usually occurs after an episode of weight loss and can manifest during puberty, possibly due to changes in lean body mass with some reduction in intraabdominal fat. Food intolerance promotes ongoing weight loss, which may further reduce intra-abdominal adipose tissue and exacerbate the problem, resulting in a vicious cycle and deterioration in clinical condition.[5,9,13,14]

Diagnosis is frequently delayed, relies on a high index of suspicion, and is often made by a process of exclusion. The differential diagnosis includes megaduodenum[15] and other more common conditions including chronic pancreatitis and peptic ulcer disease. A thorough investigative process, including gastroscopy with biopsy and contrast imaging, is recommended before arriving at the diagnosis of SMA syndrome.

The diagnosis of duodenal obstruction is made with contrast X-ray studies, either barium studies or CT imaging with oral contrast, and may demonstrate dilatation of the proximal duodenum with failure of contrast passage beyond the third part of the duodenum with a cut off.[16]

The vascular abnormalities are well-delineated by fine slice CT imaging with vascular reconstruction[17] measuring the aortosuperior mesenteric artery angle. An aortosuperior mesenteric artery angle of less than 25° is regarded as being the most sensitive measure of diagnosis, particularly if the condition is associated with diminution of the aortomesenteric distance to less than 8 mm.[4,18]

Treatment is either conservative or surgical. Fluid resuscitation, bowel rest, TPN, and enteric feeding with a nasojejunal tube inserted past the obstruction have all been advocated. In children and in adults with a short history, this may have a reasonable prospect of success, but in the chronic adult patient, conservative treatment is often a prolonged in-hospital therapy with a low success rate.[8,13] Thus, in the fit adult patient, after correction of electrolyte abnormalities and a period of refeeding, surgery is indicated.

To date, there is no data to guide as 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, all of our patients had uneventful recoveries as indicated by the length of stay and only one long-term incisional hernia. Only one patient had TPN given preoperatively and this would indicate that, in the presence

of normal serum proteins, surgery may be safely performed with the use of preoperative nutritional supplementation being used on a individualized basis. Similarly, the possibility of refeeding syndrome in the postoperative period may need to be considered, but again it did not occur in our series. We can only surmise that the prolonged period of vomiting and weight loss in the community setting differs from other situations of malnutrition in that good quality food, nutritional supplements, and vitamin supplements were frequently used by the patients and their treating clinicians in the months leading up to their eventual diagnosis, limiting the nutritional and healing effects.

Surgical options that have been proposed include mobilization of the duodenum by division of the ligament of Trietz, allowing the duodenum to fall away from the aorta (Strong's procedure);[19] duodenojejunostomy[20] with or without division of the fourth part of the duodenum; and gastrojejeunostomy.[21]. Strong's procedure has the advantage of maintaining bowel integrity, but has a failure rate of 25% presumably due to short branches of the inferior pancreaticoduodenal artery not permitting the duodenum to fall inferiorly. Gastrojejunostomy allows gastric decompression but the failure to relieve the duodenal obstruction may result in recurrent symptoms requiring a second procedure. As well, the unrelieved obstruction may result in blind loop syndromes and continuing peptic ulceration.[1,21] Duodenojejunostomy as a treatment for this condition was first described by Stavely in 1908 and is generally accepted as having superior results to both Strong's procedure and gastroenterostomy, but duodenojejunostomy without division of the fourth part of the duodenum also carries a risk of blind loop syndrome. Based on this, our preference has been for mobilization and division of the fourth part of duodenum with the proximal jejunum being passed through the right part of the mesocolon and a side to side duodenojejunostomy performed. This overcomes the obstructive problem and returns the bowel continuity to as normal as possible with minimal possibility of blind limb syndromes.

Advances in laparoscopic surgery have seen laparoscopic Strong procedures and laparoscopic duodenojejunostomy reported by several centers.[22,23] These techniques are certainly feasible and we performed a mobilization of the ligament of Trietz, division of the fourth part of the duodenum, and duodenojejunostomy laparoscopically on our last case.

Some authors have promoted resection of the abnormal duodenum in patients thought to have SMA syndrome rather than bypassing the duodenal third part, as they have postulated that it is a variant of a motility disorder rather than a true mechanical obstruction.[24] Certainly, patients with motility disorders such as intestinal neuronal dysplasia type B and familial megaduodenum may have similar clinical features of SMA syndrome. However, there is little evidence, either pathological or physiological, to support this postulate. In our cases, four patients had mechanical obstruction demonstrated on contrast studies and all six had critical narrowing of the aortomesenteric angle on CT scan. Two cases also had mechanical obstruction demonstrated at endoscopy. All patients had bypass surgery rather than resection with good improvement of their symptoms. All cases had postoperative contrast studies showing normal emptying of the duodenum which further supports a mechanical rather than motility disorder in patients with SMA syndrome. Furthermore, all of our patients have had good long-term results with weight gain and resolution of all of their preoperative symptoms of reflux and vomiting which would not be expected in a primary motility disorder. It remains somewhat puzzling, however, that our patients have not gained more weight over the period of follow-up.

There is undoubtedly a significant psychological overlay in many patients who present with this condition. Its association with anorexia nervosa, drug abuse, and other eating disorders is well-documented[9,10] and the psychological well-being of the patient must be considered in those diagnosed with this condition. It is, therefore, essential that the surgical correction is allied on a multidisciplinary basis with psychologists and dieticians to ensure optimal long-term outcomes once the physical obstruction has been relieved.

Conclusion

The SMA syndrome should be considered as a potential diagnosis in young adults who present with a history of persistent postprandial vomiting and weight loss. In its modern incarnation, it may be more commonly related to psychological and eating disorders rather than the traditional paradigm of weight loss from surgical and other physical causes and its relief includes treating of the psyche as well as surgical intervention.

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