

Case Report

Superior Mesenteric Artery Syndrome: Late Complication of Ileal Pouch-Anal Anastomosis



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ABSTRACT

Superior mesenteric artery syndrome (SMAS) is an uncommon condition which is difficult to diagnose due to non-specificity of symptoms. The most common causes of SMAS are severe weight loss secondary to severe medical conditions, surgical history, and cancer. A 31-year-old male with a history of ulcerative colitis status-post proctocolectomy with ileal pouch-anal anastomosis 10 years prior, presented with progressively worsening weight loss and abdominal pain. Radiographic imaging was consistent with SMAS, which was subsequently confirmed intraoperatively during an emergency surgery in which a Roux-En-Y gastrojejunostomy was performed. Clinicians should be aware that SMAS is a rare but possible complication of ileal pouch-anal anastomosis. Although rare, there should be a low threshold for this diagnosis when obstructive symptoms present.

Keywords: proctocolectomy, surgery, superior mesenteric artery syndrome, ulcerative colitis

Introduction

Superior mesenteric artery syndrome (SMAS) is a rare cause of small bowel obstruction caused by the compression of the duodenum between the aorta and the superior mesenteric artery (SMA) [1]. Clinical symptoms of SMAS include nausea, vomiting, postprandial fullness, anorexia, and weight loss [1-2]. Diagnosis of the SMAS is often challenging but should be suspected based on clinical presentation and radiological imaging. Due to the difficulty of diagnosis, there is often a delay in medical management which consists of gastric decompression, nutritional support, and electrolyte imbalance correction [1]. SMAS refractory to conservative management should be considered for surgical intervention [2].

A total proctocolectomy with ileal pouch-anal anastomosis (IPAA) is the gold standard surgical procedure performed in patients with familial adenomatous polyposis or ulcerative colitis (UC) [2-3]. The SMA is the main vascular pedicle in the creation of the ileal pouch during an IPAA which has the potential to be stretched and cause SMAS. A rare association

Table 1. Onset time of Superior Mesenteric Artery Syndrome occurring after restorative proctocolectomy.

Author (y) [ref]	Disease	Onset of SMAS (Post operative day)
Ballantyne et al (1987) [2]	UC	2
Christie et al (1988) [3]	UC	6
Goes et al (1995) [4]	UC	3
Ravindra et al (1999) [5]	UC	7
Essadel et al (2001) [6]	UC	5
Matheus et al (2005) [7]	FAP	3
Welsch et al (2007) [8]	FAP	7 y
Truong et al (2013) [9]	FAP	15
Kitade et al (2015) [10]	UC w/ descending colon cancer	17
Hamid et al (2017) [11]	FAP	3
Present case	UC	10 y

FAP = familial adenomatous polyposis; SMAS = superior mesenteric artery syndrome; UC = ulcerative colitis.

with SMAS is a complication of the IPAA, with a few reported cases in the literature (Table 1) [2-11]. We herein describe a rare case of SMAS that developed 10 years after IPAA in a patient with a history of UC managed with proctocolectomy.

Case Report

A 31-year-old male with a significant history of UC status-post proctocolectomy with IPAA 10 years prior, presented to the emergency department with a 3-day history of progressive and colicky epigastric abdominal pain associated with nausea, and vomiting. A detailed inquiry into the patient's history established that his symptoms of UC had largely resolved, but he still reported mild intermittent abdominal pain. All prior abdominal imaging within the past 10 years, including a computed tomography (CT) scan 3 months prior to presentation, were negative for pathology. However, the patient now reported a new-onset abdominal bloating and anorexia with weight loss of up to 40 pounds in more recent years.

Upon presentation, the patient's vitals were stable; however, his body mass index (BMI) was 16.8 kg/m^2 . Our differentials were bowel obstruction as a result of adhesions from prior surgery, versus ileus adhesions due to a slight electrolyte abnormality given the patient's BMI. Computed tomography (CT) revealed significant distension of the stomach and proximal duodenum with transitional narrowing of the duodenum as it courses between the SMA and aorta. The CT also showed an aortomesenteric angle calculated to be 10 degrees, with

diminished mesenteric fat, and compression of the duodenum (Figure 1). We then shifted our focus to the diagnosis of bowel obstruction secondary to SMAS given the patient's surgical history, anatomical location of the obstruction, and the paucity of intra-abdominal fat on imaging. At that time, the obstruction was managed conservatively with nasogastric decompression and intravenous fluids. The patient declined placement of a jejunostomy tube beyond the narrowing point and requested in-home total parenteral nutrition for conservative management of SMAS.

Two months later, the patient was readmitted due to an unstable presentation. He was hypotensive (80/55mmHg) and tachycardic (108 bpm) with a rigid, diffusely tender abdomen. The patient underwent an exploratory laparotomy in which there was a significantly dilated proximal duodenum with confirmed obstruction caused by the SMA over the third part of the duodenum. There also appeared to be another point of obstruction in the jejunum by presumably adhesive disease. A portion of the jejunum was resected with pathology revealing hemorrhagic walls with focal perforation, and a Roux-en-Y gastrojejunostomy was performed for management of his SMAS and obstructive symptoms.

Discussion

SMAS, also known as Wilkie's Syndrome, was first described in 1861 by Austrian professor, Von Rokitsansky as the obstruction of the 3rd part of the duodenum due to aortomesenteric compression [12]. SMAS is a rare diagnosis with an estimated incidence ranging between 0.013% to 0.3% [13]. Normal angulation between the aorta and the SMA (aortomesenteric angle) is between 28° and 65° with a normal SMA to aorta distance (aortomesenteric distance) between 10 and 34 mm [14]. SMAS is characterized by an aortomesenteric angle below 22° and a distance between 8-10 mm [15]. Clinical symptoms of SMAS are dependent on the degree of angulation but common symptoms include abdominal pain, bilious vomiting, nausea, vomiting and anorexia [1-2].

SMAS is a rare cause of duodenal obstruction that has been found to occur in multiple scenarios. Most commonly, SMAS occurs in young patients who undergo significant weight loss, and in patients who have a low BMI. This leads to a reduction of the retroperitoneal fatty tissue between the anterior SMA and the posterior aorta that normally acts as a cushion for the 3rd part of the duodenum. Etiologies of weight loss may include cachexia due to chronic disease and cancer, eating disorders, bariatric surgery, drug abuse, and burns [16]. Studies have also found SMAS to be associated with scoliosis corrective surgery due to a proposed pathogenesis that increased in spine length may increase tension on the mesentery [17]. Other



Figure 1. CT findings. Sagittal CT image of the abdomen and pelvis demonstrates the duodenum (asterisk) compressed between the superior mesenteric artery (arrow) and aorta (arrowhead). Aortomesenteric angle is calculated to be 10° . CT = computed tomography

scenarios seen with SMAS include extra-abdominal trauma or compression and anatomical variants of the SMA or ligament of Treitz that lead to an acute angulation of the SMA to aorta and subsequent compression of duodenum [17].

An IPAA is the gold standard to provide sphincter preservation after a total proctocolectomy, an operation commonly performed in patients with UC or familial adenomatous polyposis. During an IPAA, the SMA is used as the main vascular pedicle for the constructed ileal pouch. Goes et al therefore proposed that SMAS occurs after an IPAA because of the stretching and undesired tension of the SMA created after placement of the ileal pouch deep into the pelvis [4]. This mechanical stretching of the mesenteric root subsequently narrows the aortomesenteric angle and distance, causing duodenal compression. Extensive search of literature revealed that SMAS occurred acutely 3-17 days post-operatively after IPAA creation. Our case suggests that SMAS may be a late complication of IPAA, with the patient presenting with the longest time-to-disease reported in literature of 10 years post-IPAA.

SMAS can be diagnosed radiologically by performing upper gastrointestinal series CT, CT angiography, magnetic resonance angiography, or ultrasonography [18]. When identified in the acute setting, the first-line treatment of SMAS begins with conservative approaches including nasogastric tube placement for gastroduodenal decompression, jejunal or parenteral nutrition to increase retroperitoneal fatty tissue, and positioning into prone or left lateral decubitus position [18]. In the case of emergent settings or in symptomatic cases where conservative treatment is insufficient, surgery is indicated. Surgical options include laparoscopic approach, robotic or open gastrojejunostomy, duodenojejunostomy, and Strong's operation which consists of the division of the ligament of Treitz to mobilize the duodenum [16]. In our patient, the combination of jejunal obstruction with intraoperative confirmation of SMAS resulted in a Roux-En-Y gastrojejunostomy.

Clinicians should be aware that SMAS is a rare but possible complication of IPAA. Our case suggests that SMAS does not only present as an early and acute postoperative complication as seen in the overwhelming majority of reported cases, but may present as a late complication 1 decade later. There should be a low threshold in the diagnosis of SMAS in a patient who presents with obstructive symptoms and a history of restorative proctocolectomy. Further, SMAS with concomitant adhesive disease and downstream obstruction is an exceedingly rare presentation that may present with a closed-loop obstructive picture and should be identified early to prevent a surgical emergency.

Author Contributions

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Conflict of Interest

The authors declare that they have no competing interests.

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Ethical Statement

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Data Availability

All relevant data are included in this manuscript.

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