Abdominal Radiology (2019) 44:3188–3194
https://doi.org/10.1007/s00261-019-02066-4

REVIEW

Superior mesenteric artery syndrome: a radiographic review

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Published online: 24 May 2019

Abstract

Purpose To provide a review of the etiology, clinical presentation, and imaging findings of superior mesenteric artery (SMA) syndrome.

Methods A literature review of 24 relevant articles regarding SMA syndrome was performed.

Results Clinicians and radiologists with a high index of suspicion based on symptomatology may pursue radiologic investigation in the form of upper gastrointestinal (GI) series and contrast-enhanced abdominal computed tomography (CT). Magnetic resonance imaging (MRI) and ultrasound (US) are less commonly utilized modalities in the work-up of SMA syndrome, but provide imaging alternatives without the use of ionizing radiation. Imaging can assist in diagnosis by demonstrating characteristic findings of reduced aortomesenteric angle, reduced aortomesenteric distance, gastroduodenal distention, bowel caliber narrowing at the takeoff of the superior mesenteric artery from the aorta, as well as delayed gastric emptying or positional obstruction observed with real time with fluoroscopy.

Conclusion SMA syndrome is a rare disease that can go unrecognized and undiagnosed, exacerbating weight loss in an already significantly malnourished patient population. The diagnosis of SMA syndrome must be based on clinical symptomatology correlated with radiographic information. Once diagnosed, SMA syndrome can be safely treated by conservative measures although occasionally requires invasive intervention in the form of enteral tube placement, percutaneous jejunostomy tube placement, total parenteral nutrition, ligament of Treitz lysis, or duodenojejunostomy.

Keywords Superior mesenteric artery syndrome · Duodenal obstruction · Radiography · Tomography · X-ray computed · Fluoroscopy

Introduction

Superior mesenteric artery (SMA) syndrome results from the extrinsic compression and symptomatic obstruction of the third portion of the duodenum by the SMA anteriorly and the aorta posteriorly. This vascular compression syndrome was first described in 1861 by Carl Freiherr von Rokitansky, a Viennese pathologist, who identified the characteristic constellation of autopsy findings [1]. Wilkie subsequently published the first, and the largest to date, case series of 75 patients with SMA syndrome in 1927 [2]. Since then, multiple publications have described SMA syndrome as Wilkie syndrome, cast syndrome, mesenteric root syndrome, chronic duodenal ileus, and intermittent arteriomesenteric occlusion [3–6]. Controversy has swirled around SMA syndrome since its discovery over 150 years ago. Some have even doubted its existence despite multiple publications well characterizing its clinical and radiographic features.

SMA syndrome is rare and as a result may not be considered by either the radiologist or clinician, often going unrecognized. The symptoms can be vague, chronic, and have significant overlap with more common gastrointestinal disorders, such as gastritis, peptic ulcer disease, irritable bowel syndrome, and gastroparesis [7]. It is important for the clinician to have a high index of suspicion of SMA syndrome in the appropriate patient population in order to perform the appropriate radiological testing and ultimately reach the correct diagnosis.

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Etiology

Anatomically, the third portion of the duodenum passes between the aorta and the SMA. A retroperitoneal fat pad that sits beneath the takeoff of the SMA from the aorta acts as a buffer and bolster, allowing the duodenum to pass unobstructed between the two vessels (Fig. 1). Severe weight loss can lead to a reduction in the size of the aforementioned fat pad in turn narrowing the angle between the SMA and the aorta [8]. This narrowing ultimately leads to extrinsic compression and obstruction of the duodenum, preventing the passage of intestinal contents [9].

Clinical presentation

The diagnosis of SMA syndrome requires a high index of suspicion. Symptoms are often vague and can be associated with other more common underlying gastrointestinal pathology such as gastritis, gastroparesis, gastroesophageal reflux disease, and irritable bowel syndrome. Signs and symptoms of SMA syndrome can include early satiety, postprandial pain or discomfort, nausea and bilious emesis often after a meal, bloating, eructation, and reflux [10–12]. Patients frequently report resolution of symptoms following an episode of emesis. Over time, as patients begin to anticipate the postprandial symptoms of pain, nausea, and emesis, they tend to avoid eating altogether. Food aversion and decreased nutritional intake perpetuate the disorder as further weight loss leads to continued decrease in retroperitoneal fat at the mesenteric root and a resultant increase in the degree of obstruction.

The initial precipitating weight loss that leads to SMA syndrome can be severe and rapid in onset secondary to acute etiologies such as trauma and extensive burns. Other subacute causes include cancer, chronic inflammatory states, and anorexia nervosa when the weight loss is gradual. The etiologies of the initiating weight loss are also loosely organized into four general categories: nutritional or eating disorders, catabolic states, prolonged bed rest, and mechanical or structural causes. Anorexia nervosa, malabsorption, and starvation are included with nutritional or eating disorders as risk factors for SMA syndrome. Catabolic states with greater metabolic demands include cancer, burns, and chronic inflammatory states such as AIDS. Prolonged bed rest is also associated with SMA syndrome and can be seen with use of body casts, neurologic injuries, and loss of muscle tone [13]. Finally, mechanical or structural risk factors for SMA syndrome include asthenic build, accelerated linear adolescent growth, scoliosis, exaggerated lumbar lordosis (pathologic or iatrogenic/surgical), high ligament of Treitz, low SMA takeoff, and peritoneal adhesions [6, 10].

Duodenal obstruction in SMA syndrome can be complete or incomplete based on the degree of narrowing of the duodenal lumen, partial obstruction being more common. This narrowing is largely thought to be due to a decrease in the aortomesenteric angle (AMA), formed by the takeoff of the superior mesenteric artery off of the abdominal aorta. Patient positioning can also affect the degree of duodenal obstruction. Duodenal obstruction is maximal in the supine position as this decreases the distance between the SMA and aorta, also known as the aortomesenteric distance (AMD). Hence, supine position characteristically exacerbates symptoms. Symptoms of SMA may be relieved by prone or left lateral decubitus positioning as the SMA falls away from the aorta, increasing the AMD and thereby lessening the degree of obstruction [9, 11]. Other postural methods of decreasing SMA obstruction include a forward leaning sitting position and even bringing the knees to the chest [6].

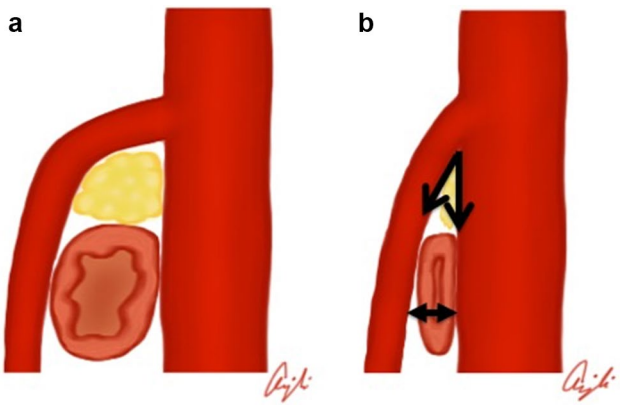

Fig. 1 **a** Normal aortomesenteric relationship and **b** the aortomesenteric relationship in SMA syndrome where the proximal SMA compresses the mid-transverse duodenum against the abdominal aorta. Extrinsic compression of the mid-transverse duodenum obstructs emptying, resulting in dilation of the stomach and proximal duodenum. The aortomesenteric angle (lines with open arrows) and aortomesenteric distance are demonstrated (line with closed arrows)

Imaging findings

Plain films

An abdominal radiograph is often the first radiologic study performed on a patient with symptoms concerning for SMA syndrome. Typical radiographic findings include gastric or gastroduodenal distention and a paucity of distal bowel gas (Fig. 2) [11, 14]. In the proper clinical context, these findings may prompt the clinician to suspect SMA syndrome and investigate further with single contrast upper GI barium

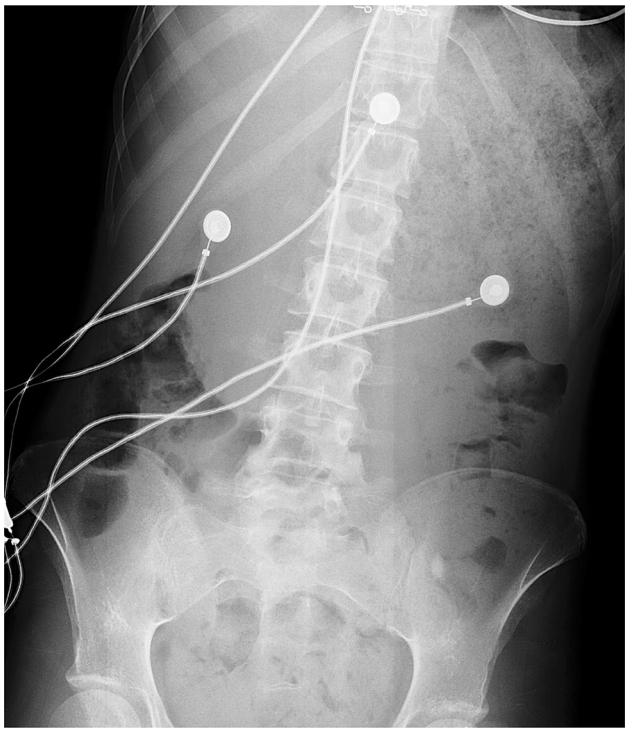

Fig. 2 Abdominal radiograph showing a markedly dilated stomach containing food debris. In the proper clinical context, this finding may prompt the clinician to consider work-up for possible SMA syndrome

study or contrast-enhanced abdominal computed tomography (CT).

Upper GI Barium study

Fluoroscopic imaging allows for dynamic, real-time evaluation of clinical history and imaging findings suspicious for SMA syndrome and is classically the imaging modality of choice in evaluation of SMA syndrome. The fluoroscopic findings of SMA syndrome include gastroduodenal dilation, delayed gastroduodenal emptying, and a vertical, linear, band-like defect across the third portion of the duodenum due to vascular compression by the SMA (Fig. 3). Other findings include a characteristic 'to and fro' pattern of peristalsis, commonly described as antiperistaltic flow, of the enteric contents proximal to the obstruction. Finally, patients with true vascular compression of the duodenum often demonstrate decreased obstruction with postural changes [11, 15–17]. For example, obstruction is characteristically the greatest in the supine position and the least in the prone and left lateral decubitus positions thereby allowing passage of enteric contrast (Fig. 4).

Computed tomography

While the upper GI barium study is classically most useful in diagnosis of SMA syndrome, complementary CT findings can aid in corroborating the diagnosis. CT with intravascular contrast allows for evaluation of aortomesenteric vasculature as well as gastroduodenal caliber, and hence, it is a valuable imaging modality to evaluate patients with suspected SMA syndrome. The CT findings of SMA syndrome include a

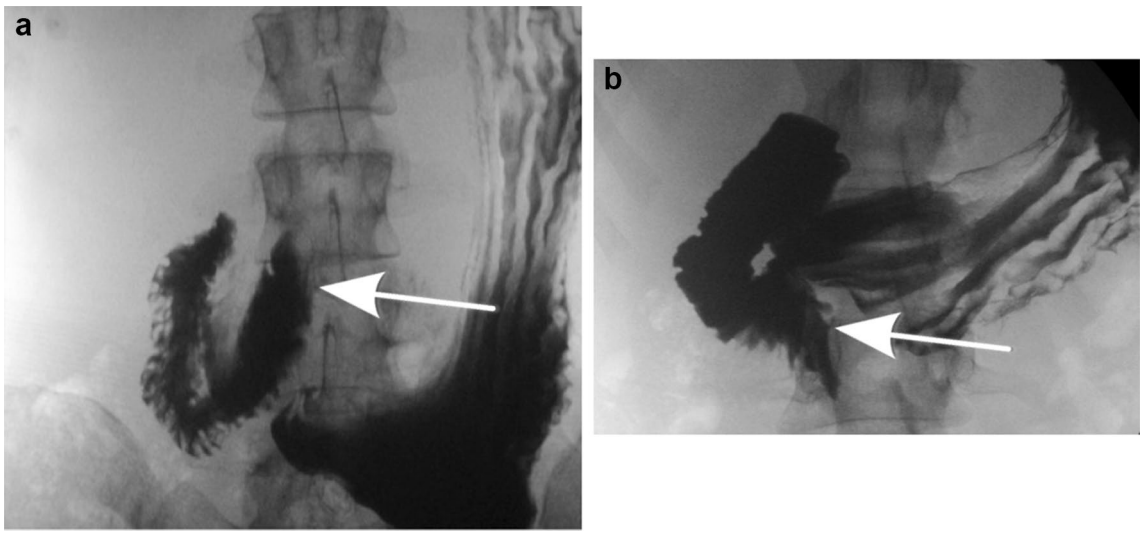

Fig. 3 **a**, **b** Images from upper gastrointestinal barium studies demonstrating the typical vertical defect (arrow) across the mid-transverse duodenum from extrinsic compression of the overlying SMA

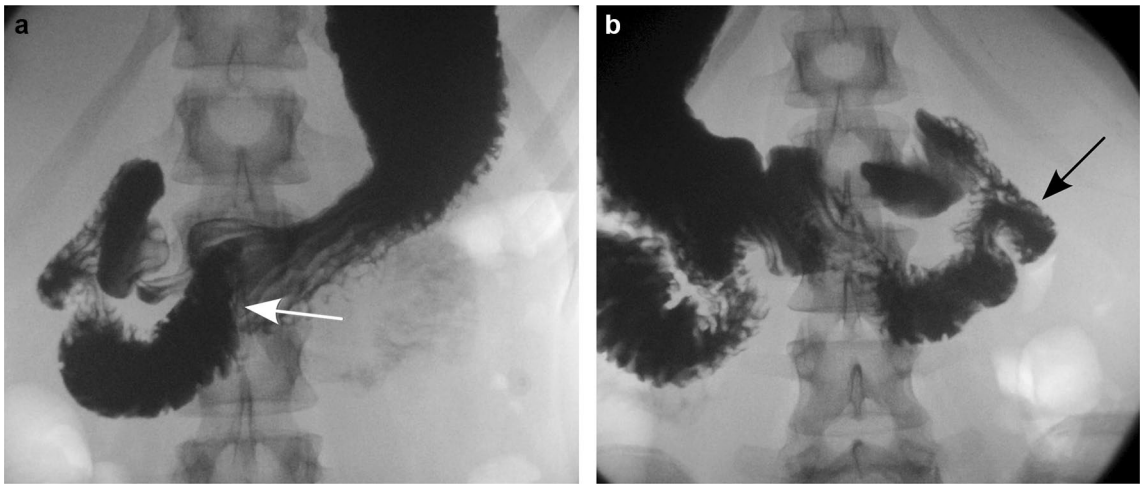

Fig. 4 SMA syndrome in a 22-year-old female with a history of eating disorder. **a** Image from an upper gastrointestinal barium study demonstrates the typical vertical defect (white arrow) across the mid-transverse duodenum in the supine position. **b** As a result of patient repositioning into a prone position, there is effective passage of contrast into the distal duodenum (black arrow)

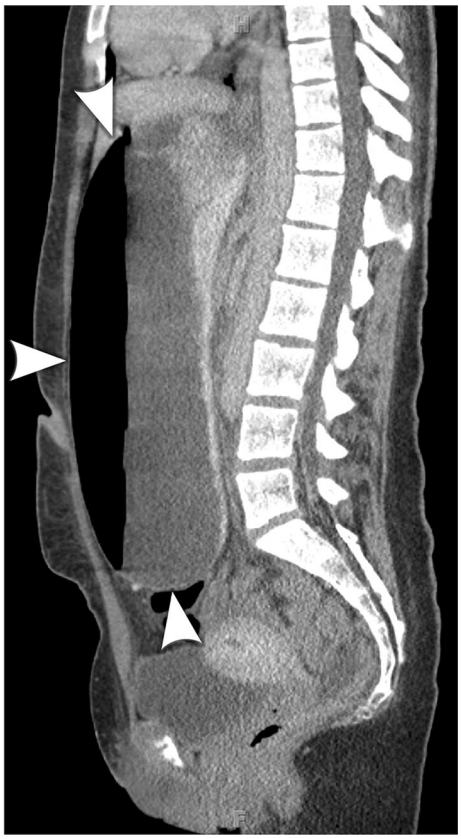

Fig. 5 Midline sagittal reformatted abdominal CT image demonstrating severe gastric dilation (arrowheads) due to SMA syndrome

narrowed AMA, a decreased AMD, and gastroduodenal dilation with an abrupt narrowing in bowel caliber at the SMA takeoff from the aorta (Fig. 5) [5, 15].

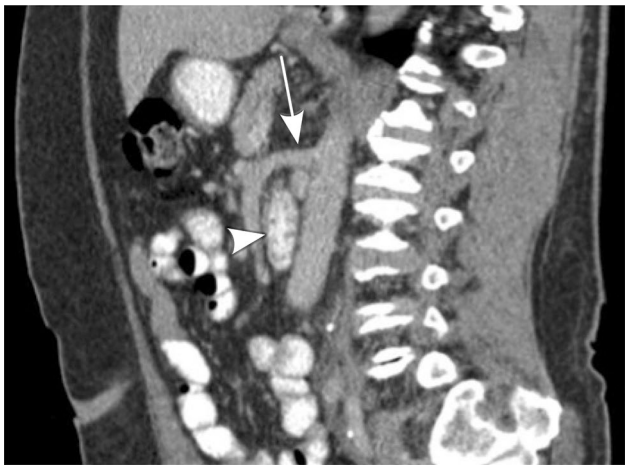

Fig. 6 Midline sagittal reformatted abdominal CT image demonstrating the normal relationship between the superior mesenteric artery (white arrow), aorta, and mid-transverse duodenum (arrowhead). The aortomesenteric angle (between 28° and 65°) and the aortomesenteric distance (> 10 mm) are both normal, allowing unobstructed passage of the duodenum

The vascular angles and distances are measured on the CT sagittal and axial reformations, respectively. The normal AMA formed by the SMA and aorta is between 38 and 56° (Fig. 6) [6, 12]. When the AMA decreases to 6-22°, extrinsic compression of the duodenum can occur. The AMD also decreases from a normal interval of 10-28 mm to 2-8 mm in SMA syndrome (Fig. 7a, b) [10, 12]. While these measurements can be performed on routine portal venous phase CT, CT angiography provides the added benefit of thin (1-mm) slice thickness and multiplanar 3D reconstructions which allow for more precise evaluation of the AMA and AMD

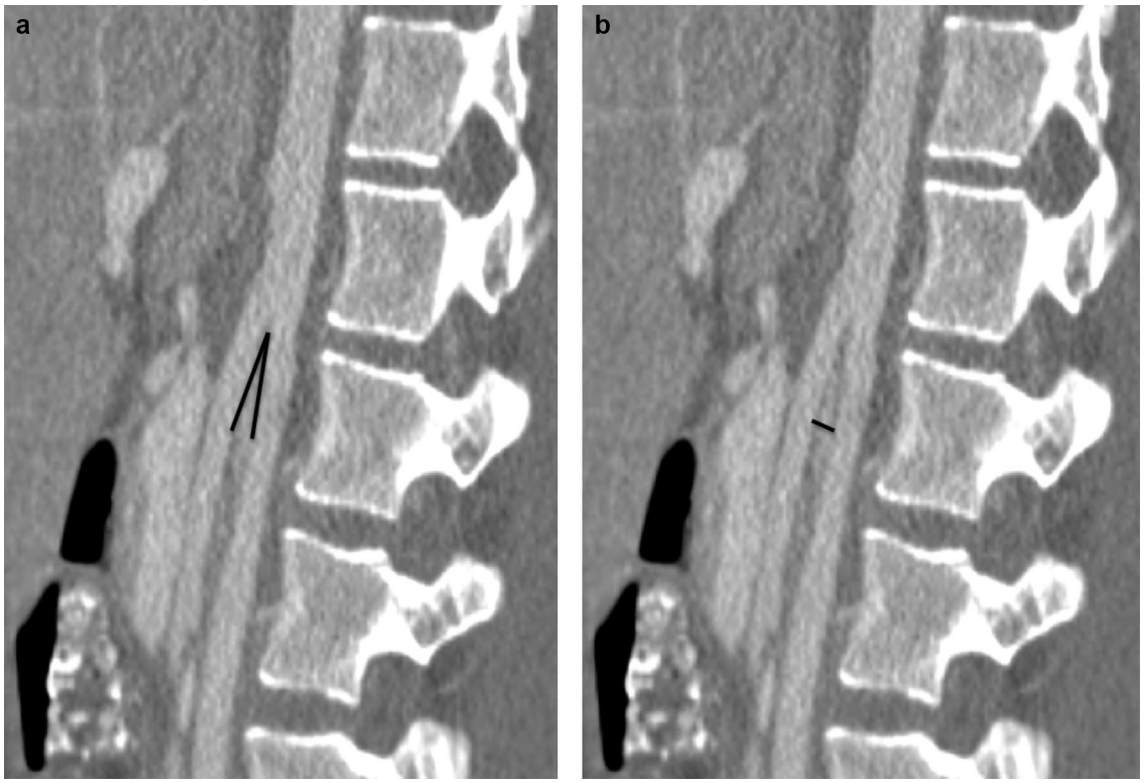

Fig. 7 Midline sagittal reformatted abdominal CT image. **a** narrow aortomesenteric angle measuring 28° degrees, the lower limit of normal; and **b** a short aortomesenteric distance < 10 mm consistent with SMA syndrome

(Figs. 8, 9) [4]. In addition, vascular compression of the duodenum is often more easily elucidated on CT angiography reformations compared with portal venous phase CT.

Gastroduodenal dilation and the obstructive transition point at the SMA are best demonstrated on the CT axial images and coronal reformations. Although oral contrast can be helpful to determine the degree of the obstruction, patients may not tolerate oral intake. Also, because extrinsic compression of the duodenum may result in only partial obstruction, transit of enteric contrast beyond the site of vascular compression does not exclude SMA syndrome from the differential diagnosis. As SMA syndrome is rare and considered a diagnosis of exclusion, CT provides the added benefit of potentially identifying other etiologies to explain symptomatology [15].

Other imaging alternatives

Magnetic resonance imaging (MRI) provides cross-sectional imaging without ionizing radiation; however, it is a less commonly utilized modality for the evaluation of SMA syndrome. Expected findings are identical to CT findings and include a narrowed AMA, a decreased AMD, and gastroduodenal dilation with an abrupt narrowing in bowel caliber at the takeoff of the SMA. The increased acquisition time of

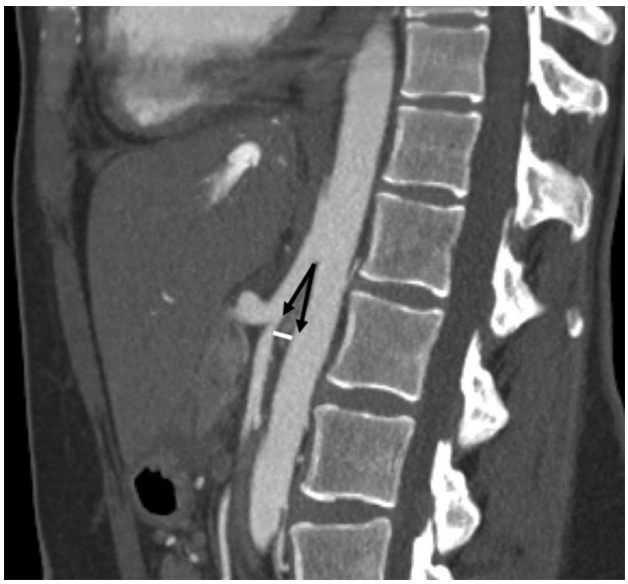

Fig. 8 41-year-old female with SMA syndrome, initially presenting with early satiety, abdominal pain, and weight loss. Midline sagittal CT angiogram reformatted image demonstrates abnormal aortomesenteric angle measuring 14 degrees (black lines) and abnormal aortomesenteric distance measuring 6 mm (white line). Variant anatomy with common origin of the celiac artery and superior mesenteric artery is incidentally noted

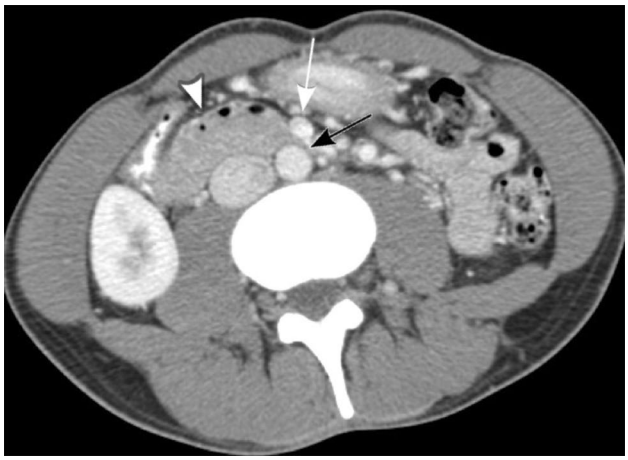

Fig. 9 Axial CT image depicting SMA syndrome with proximal duodenal dilation (arrowhead) transitioning to normal caliber mid-transverse duodenum (black arrow) just distal to the obstructing SMA (white arrow)

MRI compared with CT allows for differentiation between peristalsis and fixed narrowing of the duodenum at the level of the takeoff of the SMA. Cine images also allow for direct visualization of peristaltic activity. Cicero et al described a correlation between Crohn's disease and SMA syndrome using magnetic resonance enterography (MRE) findings of SMA in patients while also evaluating Crohn's disease bowel lesions, a chronic inflammatory condition associated with the catabolic state etiology of SMA syndrome [18].

Ultrasound also provides a low-cost imaging alternative for evaluation of SMA syndrome without the use of ionizing radiation. Neri et al demonstrated that ultrasound is a useful imaging technique in demonstrating the reduced AMA and AMD [7]. Unal et al also describe the successful acquisition of the AMA and AMD in the sagittal and transverse planes, respectively, and demonstrated a statistically significant relationship between sonographic and CT measurements [15]. Additional sonographic findings include gastroduodenal dilation and narrowing of bowel caliber between the aorta and SMA [7, 19]. While not a commonly practiced imaging modality for the work-up of SMA syndrome in the United States, Mauceri et al demonstrated a statistically significant correlation between ultrasound and CT findings. The author group therefore suggests that ultrasound with color doppler is efficacious in the evaluation of reduced AMA when SMA syndrome is suspected as the cause of inexplicable abdominal pain [20].

Treatment

As SMA syndrome is a diagnosis of exclusion, additional work-up with upper endoscopy, colonoscopy, or functional gastrointestinal studies, may be considered in an effort to exclude other more common etiologies. SMA syndrome treatment typically begins with conservative measures such as small meals, liquid diet, and postural therapy to promote passage of intraluminal contents [6, 12, 14]. The results of an upper GI barium study may aid in guiding treatment by demonstrating the positions that allow the effective passage of enteric contents. Diet consistency can be advanced as tolerated with weight gain as the retroperitoneal fat pad increases in size, alleviating the degree of duodenal luminal narrowing [17]. To date, there are no data suggesting how much weight must be gained to reconstitute retroperitoneal fat and alleviate symptoms. More invasive management includes enteral tube placement, percutaneous jejunostomy tube placement, and total parenteral nutrition [12, 21]. In cases of intractable symptoms, surgical consultation for ligament of Treitz lysis with mobilization of the ascending duodenum or duodenojejunostomy can be pursued [6, 22–24].

Conclusion

SMA syndrome is a rare disease that can go unrecognized and undiagnosed, exacerbating weight loss in an already significantly malnourished patient population. Clinicians and radiologists with a high index of suspicion based on symptomatology may pursue radiologic investigation in the form of upper GI series, often complemented by contrast-enhanced abdominal CT or CT angiography. MRI and ultrasound are less commonly utilized modalities in the work-up of SMA syndrome but provide imaging alternatives without the use of ionizing radiation. Ultimately, the diagnosis of SMA syndrome must be based on clinical symptomatology correlated with radiographic information. Once diagnosed, SMA syndrome can be safely treated by conservative measures although occasionally requires invasive intervention in the form of enteral tube placement, percutaneous jejunostomy tube placement, total parenteral nutrition, ligament of Treitz lysis, or duodenojejunostomy.

Take-home points

1. SMA syndrome is a rare diagnosis and can go unrecognized and undiagnosed by clinicians and radiologists.
2. Upper GI barium study is classically the preferred imaging modality and can aid in diagnosis and be effective in demonstrating the degree of obstruction and effect of positioning on the passage of intraluminal contents. CT and CTA provide the added benefit of quantitating both the AMA and AMD, as well as assessing for vascular compression of the duodenum. MRI and ultrasound, though less commonly utilized in the work-up of SMA syndrome, provide alternatives without the use of ionizing radiation.

3. Management of SMA syndrome is typically conservative, although invasive intervention may be pursued in more severe or intractable cases.

Acknowledgements The authors thank Anjuli Cherukuri, MD for the medical illustrations.

Compliance with ethical standards

Funding There are no relevant sources of funding or financial interest to disclose.

Conflicts of interest There are no potential conflicts of interest. The retrospectively collected images were obtained in the course of treatment and have been anonymized with no personal identifying information.

Ethical approval The article was approved by the Colorado Multiple Institutional Review Board. This article does not contain any studies with human participants or animals performed by any of the authors.

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