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Superior mesenteric artery syndrome: spectrum of CT findings with multiplanar reconstructions and 3-D imaging

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Abstract

Objective: This article reviews the causes, clinical presentation, and CT diagnosis of superior mesenteric artery (SMA) syndrome.
Conclusion: In conjunction with an appropriate clinical history, several CT findings can suggest the diagnosis of SMA syndrome. These findings include narrowing of the aortomesenteric angle and distance, distension of the stomach and duodenum, and dilatation of the left renal vein with left-sided venous collaterals.

Key words: Computed tomography—Superior mesenteric artery syndrome—SMA syndrome—3-D reconstruction—Multiplanar reformation

An entity first described almost 150 years ago, ''superior mesenteric artery (SMA) syndrome'' represents a unique set of clinical symptoms caused by compression of the duodenum between the aorta and SMA [1, 2]. Classically described in young women, patients experience early post-prandial satiety, abdominal pain, nausea, and vomiting, often resulting in chronic anorexia and weight loss [2]. Many have historically been skeptical as to the true existence of this syndrome, partly because of its rarity, but also because of the difficulty in definitively proving the presence of true duodenal obstruction with any imaging modality [3]. Nevertheless, the development of multidetector CT (MDCT) with multiplanar reformats and 3-D technique has shown that certain CT findings can be extremely suggestive, especially in the setting of an appropriate clinical history. In this pictorial essay we present 10 cases with classic CT findings and suggestive clinical histories, some of whom had their symptoms resolve following operative intervention.

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Background

SMA syndrome is thought to result from an abnormally short distance between the aorta and SMA, which results in compression of the duodenum. Patients with SMA syndrome usually present with nonspecific symptoms, making diagnosis extremely difficult. Typically seen in young women, their chronic anorexia, nausea, vomiting, and postprandial abdominal pain are often blamed on non-anatomic, psychosocial causes, resulting in a delayed diagnosis [4]. Nevertheless, when carefully questioned, these patients often have a characteristic history, with their symptoms relieved by changes in posture, such as turning to their left side, bringing their knees up to their chest, or the prone position [4, 5]. Because of its infrequency, the exact incidence of SMA syndrome is unknown, although estimates based on fluoroscopic studies have suggested it may be as low as 0.01% [6].

It is believed that the cause of this anatomic compression is loss of intra-abdominal fat, which normally separates the SMA from the aorta. As a result, SMA syndrome has primarily been described in two patient populations: (1) Patients who have undergone surgery, resulting in loss or distortion of the normal retroperitoneal fat (such as scoliosis surgery, aortic aneurysm repairs, or any abdominal gastrointestinal surgery), and (2) Patients who have had severe, rapid weight loss for any number of reasons, including bariatric surgery, cancer, chronic immobilization, major burns, etc. [1, 2, 4, 6]. Many of these patients may be congenitally predisposed

to this condition by a lean body habitus, a short ligament of Trietz, or a low take-off of the SMA [7].

Treatment

Treatment for SMA syndrome is typically conservative, including placement of a nasogastric tube to decompress the duodenum, placing the patient in positions which minimize duodenal compression (left decubitus, prone, knees to chest), and the use of tube feeds and total parenteral nutrition (TPN) to improve nutrition. The ultimate goal of conservative therapy is to increase body weight, hopefully restoring the aortomesenteric distance to a normal value, thereby relieving the patient's obstruction [1].

In those patients in whom conservative methods have failed, surgical intervention has been advocated, including the use of a duodenojejunostomy to bypass the obstructed portion of the duodenum [1]. A historical alternative was the "Strong's Operation," where the ligament of Trietz is severed, and the bowel is repositioned to alleviate duodenal compression [7]. Although most studies of surgical intervention have been in small series, the success rates have been reported to be between 75% and 100% [8, 9].

CT diagnosis

In a normal patient, the distance between the aorta and SMA ("aortomesenteric distance") should range from 10 to 34 mm, and the normal angle between the aorta and SMA ("aortomesenteric angle") should be between 28° to 65° (Fig. 1) [6]. Angiographic studies have shown that patients with SMA syndrome clearly have an abnormal aortomesenteric angle (6°–22°), and a shortened aortomesenteric distance (2–8 mm) compared to normal patients (Figs. 2, 3, 4, 5, 6, 7, 8, 9, 10, 11) [10]. In another study utilizing MDCT, the mean aortomesentric angle in SMA syndrome patients was 13.5°, with an aortomesenteric distance of 4.4 mm [6]. Clearly, any absolute numerical cut-off is arbitrary, but some have advocated that an aortomesenteric distance <8 mm and an angle $<22°$ should be suggestive of the diagnosis in the right clinical setting [6]. In our experience, because the SMA is often anterolateral relative to the aorta, simply using sagittal multiplanar reformats can result in erroneous measurements. Therefore, 3-D reconstructions are helpful in providing the most accurate measurements by placing the aorta and SMA in a parallel plane [11].

Despite some studies suggesting that these numerical cut-offs are relatively specific for the diagnosis, a great deal of caution should be used before using these measurements in isolation [6]. Ancillary findings of duodenal obstruction should be sought, including distension of the stomach and proximal duodenum, with abrupt narrowing of the duodenum at the level of the SMA [10]. Some have observed that in true cases of duodenal obstruction, the gastric antrum is often displaced anterosuperiorly relative to the main portal vein, up into the porta hepatis [11]. At the same time, duodenal distension alone is not diagnostic—a "megaduodenum" can be idiopathic, or can be secondary to diabetes, pancreatitis, peptic ulcer disease, neoplasm, or a number of other chronic underlying disorders [10].

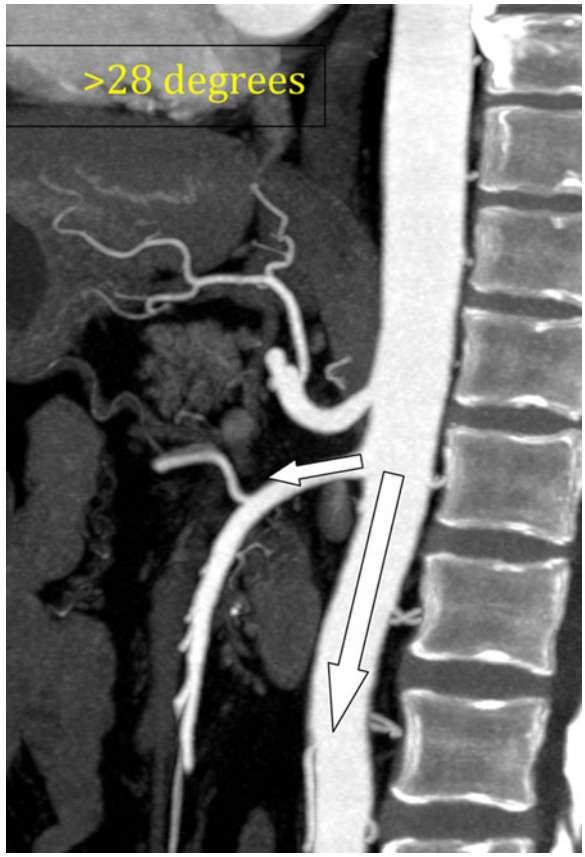

Fig. 1. Sagittal contrast-enhanced MIP CT image demonstrating a normal aortomesenteric angle.

In addition to narrowing of the duodenum, a small aortomesenteric distance can result in compression of the left renal vein between the aorta and SMA. Patients can present with isolated left renal vein thrombosis, or alternatively, enlargement of the left gonadal vein or other left-sided venous collaterals as a result of chronic renal vein compression ("nutcracker phenomenon") (Figs. 2, 6, 7, 10) [6]. This finding can be an important clue that a narrowed aortomesenteric distance is physiologically significant, rather than an incidental finding.

In our experience, multiplanar reconstructions and 3-D technique are critical in best delineating many of these ancillary findings of SMA syndrome—the presence of a large venous collateral, or the asymmetric dilatation of the left renal vein, are often more apparent with 3-D technique (Figs. 2, 6, 7, 10). Moreover, the full extent of duodenal obstruction is often best appreciated with multiplanar reformations.

Ultimately, it is dangerous to make the diagnosis of SMA syndrome based on CT imaging features alone. The CT findings can certainly be suggestive, but can be

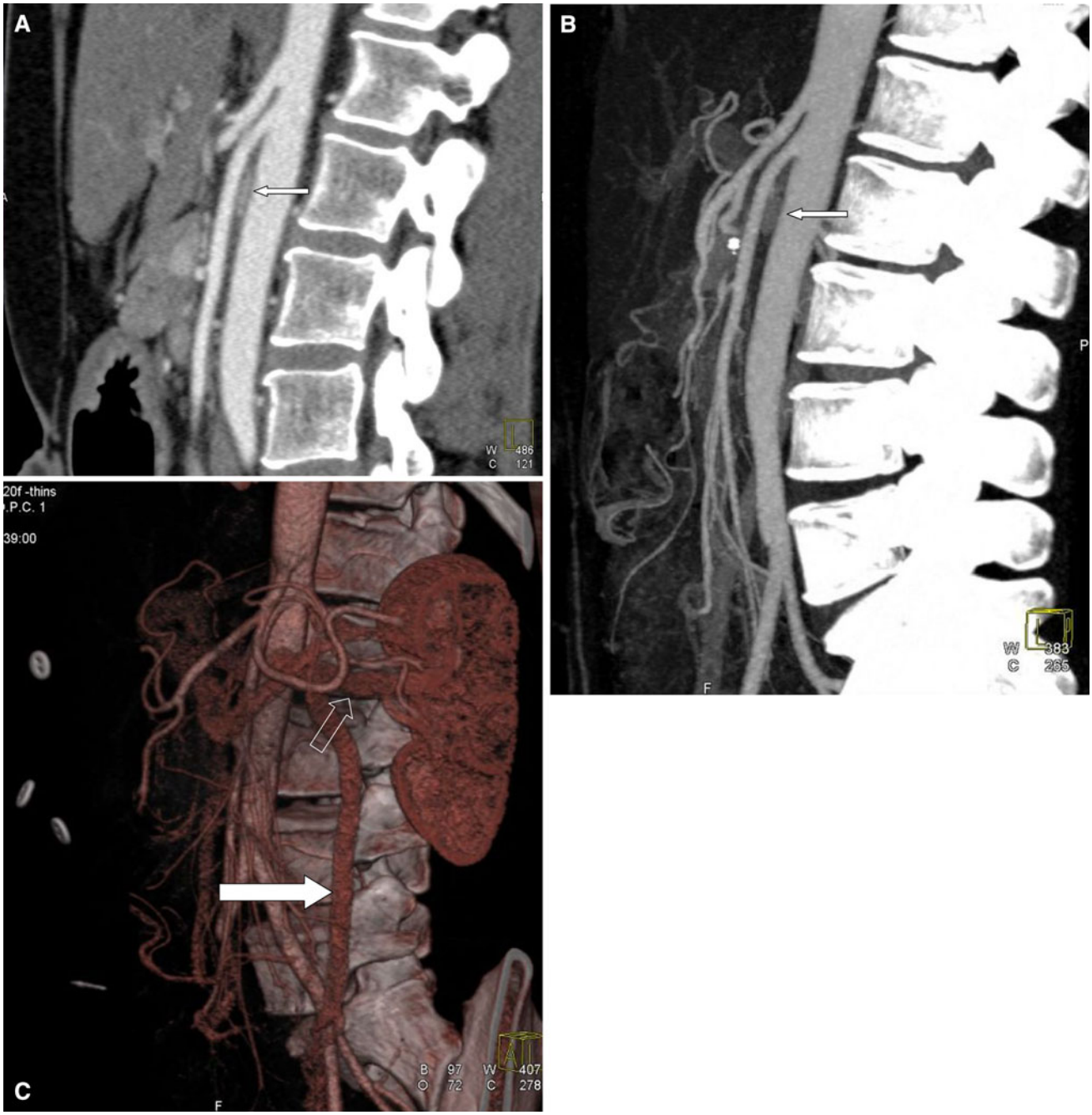

Fig. 2. A 46-year-old female with debilitating post-prandial abdominal pain, which had led to severe weight loss. Sagittal contrast-enhanced (**A**) and sagittal maximum intensity projection (MIP) (**B**) CT images demonstrate severe narrowing of the aortomesenteric angle, with an aortomesenteric distance of only 5 mm, and resultant compression of the duodenum (*arrow*). Volume-rendered CT image (**C**) demonstrates a compressed, dilated left renal vein (*open arrow*) and marked dilatation of the left gonadal vein (*solid arrow*). The patient underwent duodenojejunostomy, and subsequently gained weight.

present in normal subjects as well. CT findings, even when severe, must be interpreted in the context of the patient's symptoms, given the rarity of the disorder. SMA syndrome is not simply a constellation of CT findings, but rather, represents a group of distinct clinical symptoms.

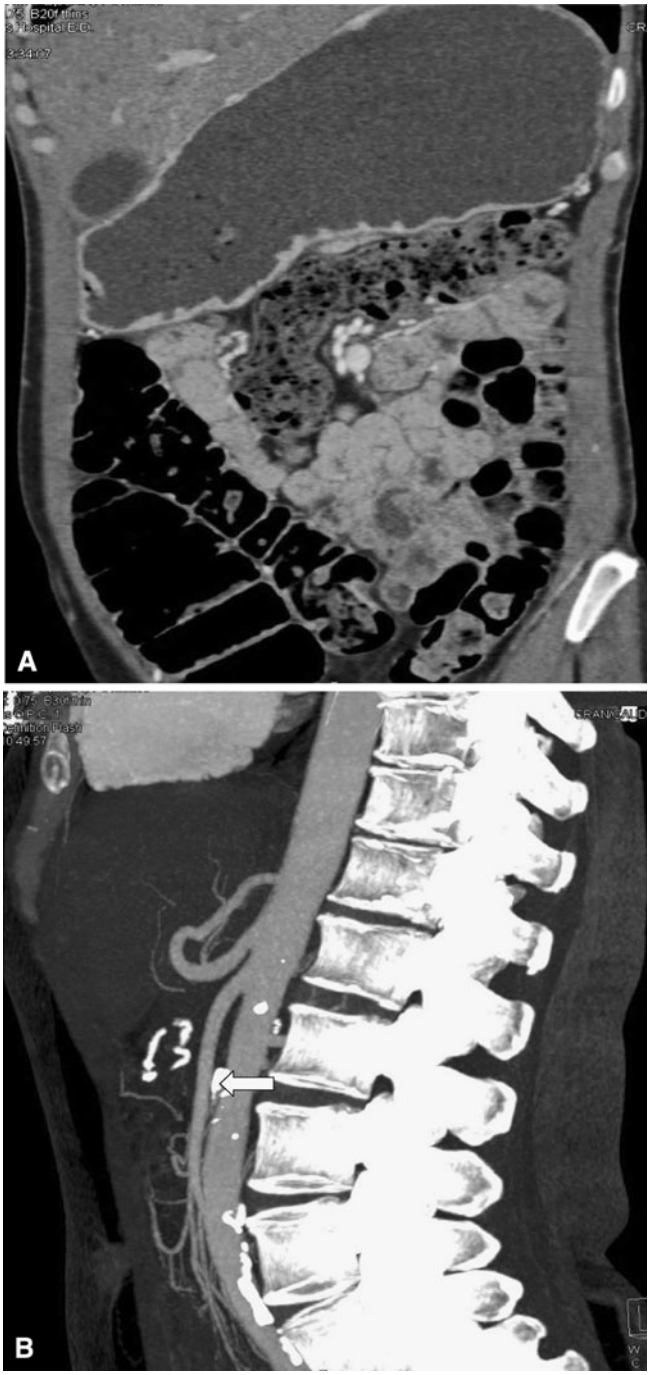

Fig. 3. A 45-year-old female with a roughly 18-month history of progressive abdominal pain, bloating, and anorexia, with fear of eating. Coronal CT image (**A**) demonstrates a markedly dilated stomach, while a sagittal MIP image (**B**) demonstrates a narrowed aortomesenteric angle and distance, with compression of the duodenum (*arrow*). She subsequently underwent a laparoscopic side-to-side gastrojejunostomy, with resolution of her symptoms.

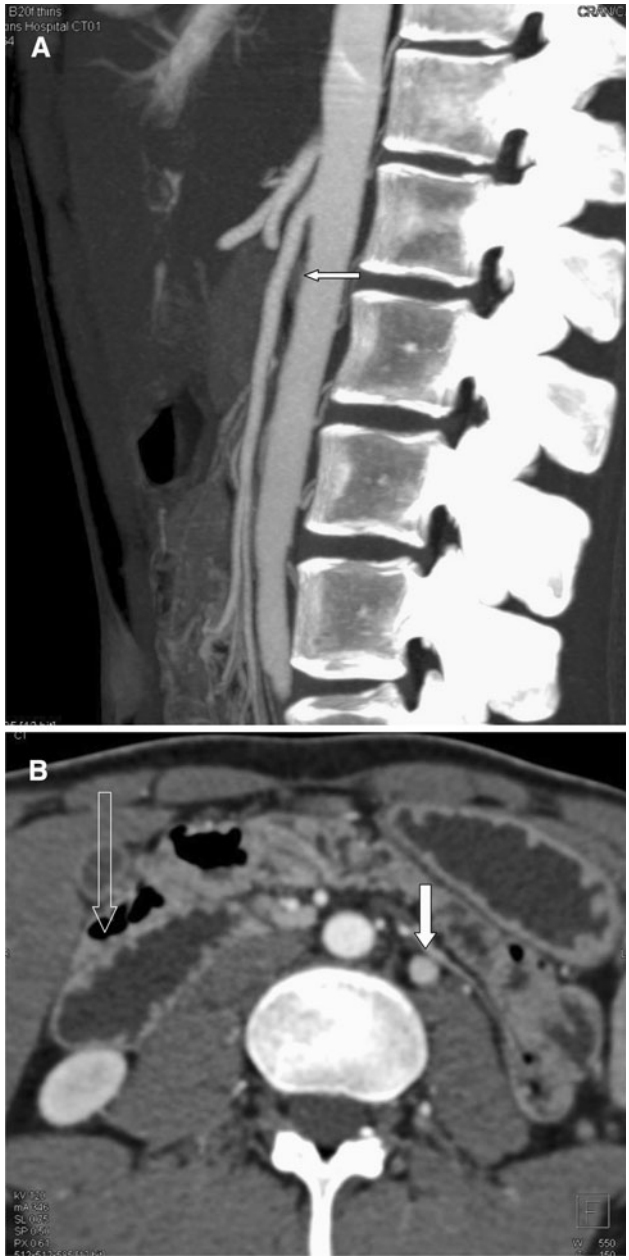

Fig. 4. A 20-year-old female with nausea, vomiting, and weight loss, as well as post-prandial abdominal pain since the age of 14. She had lost roughly 30 pounds over the last few months. Sagittal CT MIP image (**A**) demonstrates narrowing of the aortomesenteric angle and distance, with compression of the duodenum (*arrow*). Axial CT image (**B**) demonstrates a dilated duodenum (*open arrow*) proximal to where it crosses beneath the SMA. The left gonadal vein is dilated (*closed arrow*), an ancillary finding of SMA syndrome.

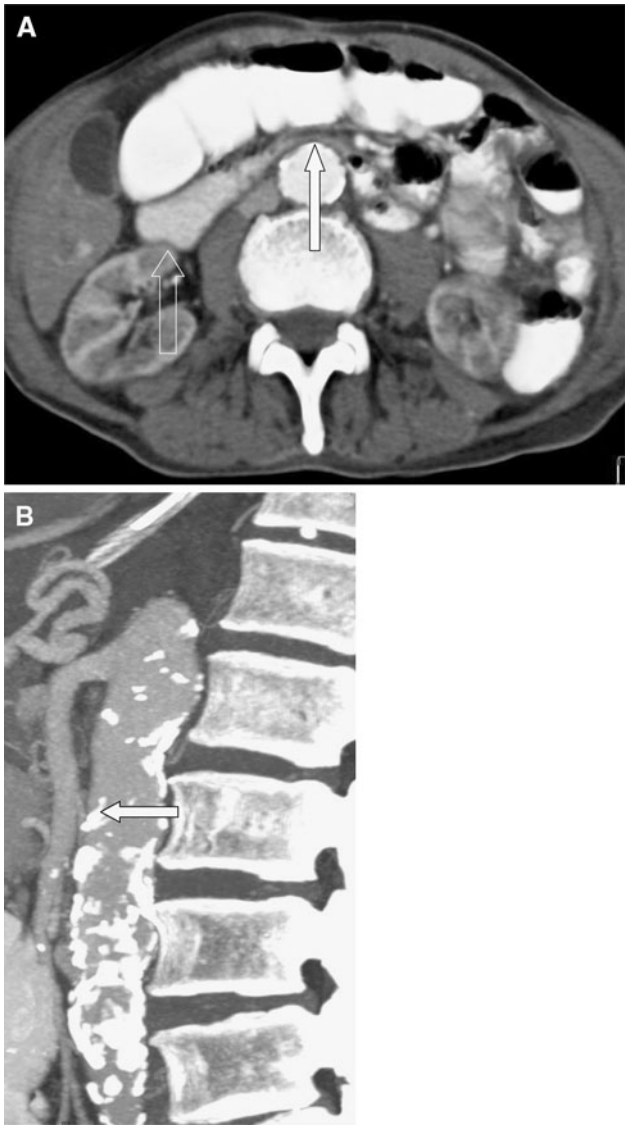

Fig. 5. A 76-year-old male with chronic abdominal pain. Axial CT image (**A**) demonstrates a dilated duodenum (*open arrow*) proximal to the SMA, with abrupt narrowing of the duodenum (*closed arrow*) as it passes under the SMA. Sagittal CT MIP image (**B**) demonstrates that although the aortomesenteric angle is normal, significant atherosclerosis and tortuosity of the SMA and aorta result in an area of narrowing between the aorta and SMA (*arrow*).

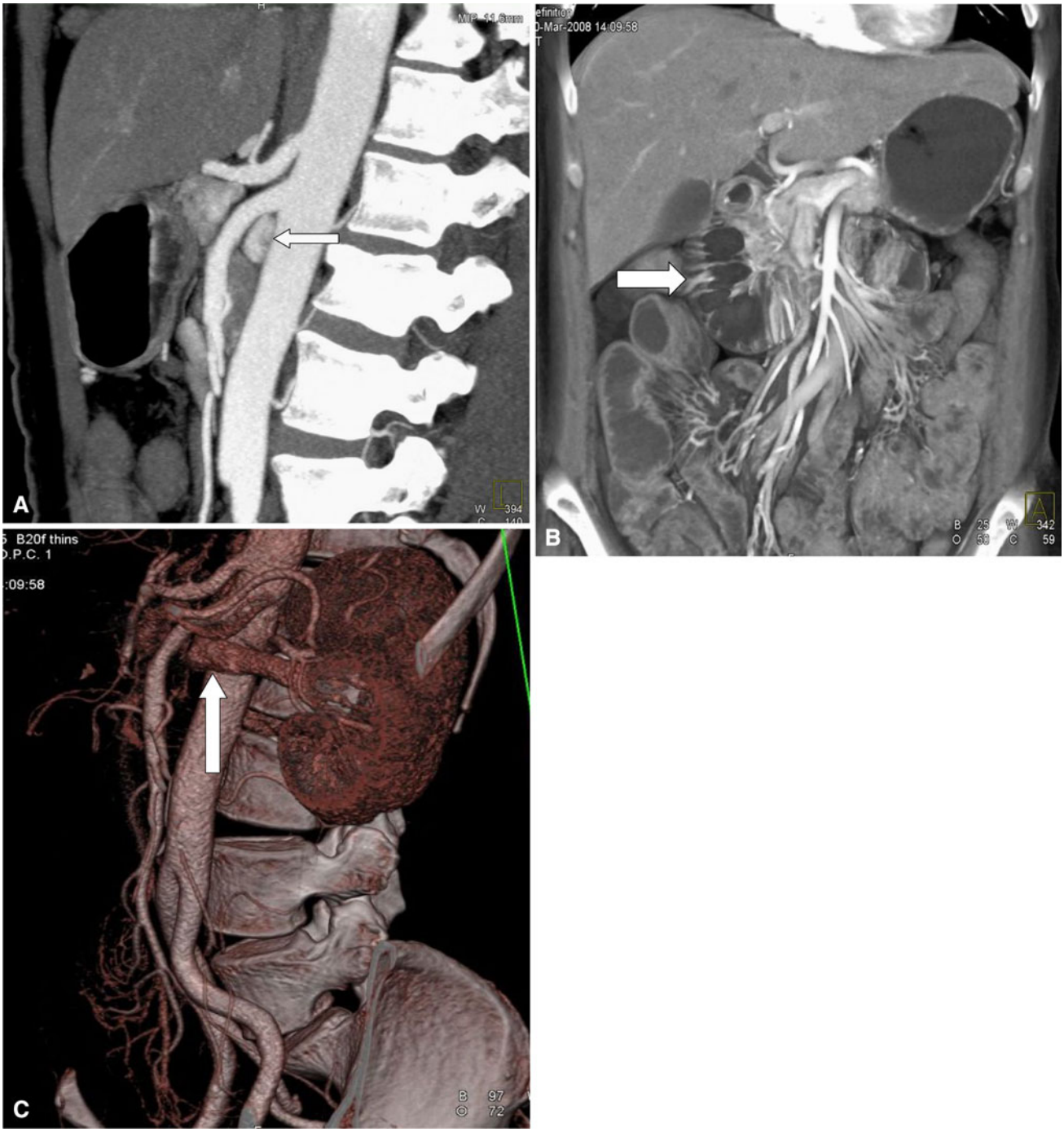

Fig. 6. A 51-year-old female with chronic abdominal pain. Sagittal MIP CT image (**A**) demonstrates narrowing of the aortomesenteric angle and distance, with compression of the duodenum (*arrow*). Coronal volume-rendered image (**B**) demonstrates distension of the stomach and proximal duodenum (*arrow*). Volume-rendered image (**C**) demonstrates pinching of the left renal vein (*arrow*) between the aorta and SMA.

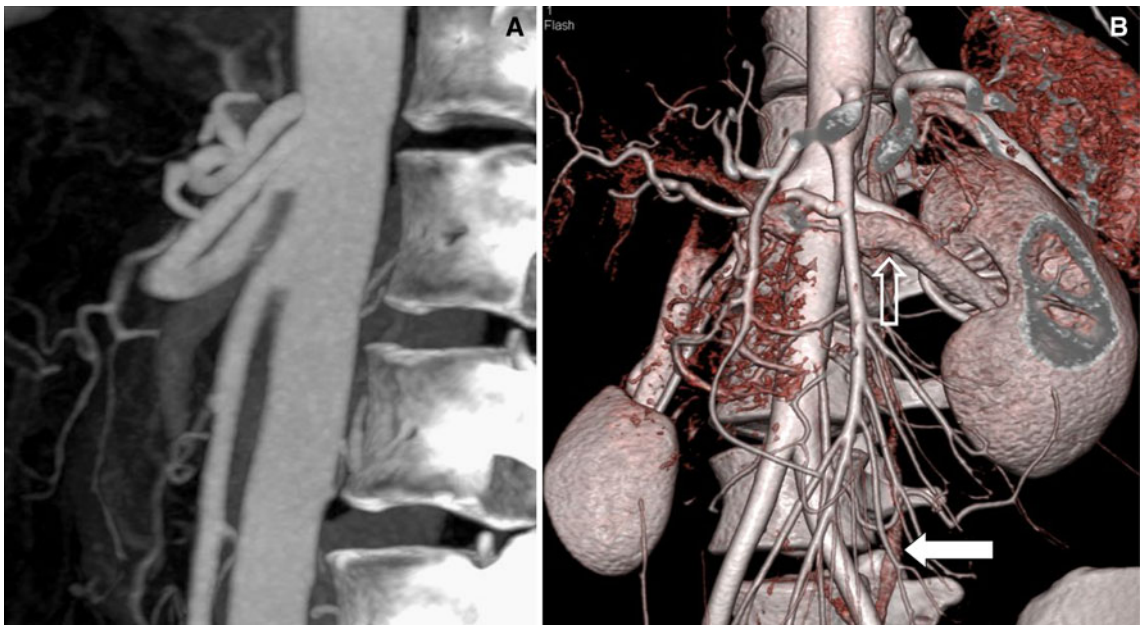

Fig. 7. A 32-year-old female with chronic abdominal pain, bloating, nausea, emesis, and weight loss. She was limited to a liquid diet as a result of her nausea and vomiting. Sagittal MIP image (**A**) demonstrates a markedly narrowed aorto-mesenteric angle. Volume-rendered image (**B**) demonstrates a dilated, compressed left renal vein (*open arrow*) with a large left lumbar collateral vein (*closed arrow*). Her symptoms largely resolved after a laparoscopic duodenojejunostomy.

Fig. 8. A 21-year-old female with a history of desmoid tumors (not shown) who presented with chronic abdominal pain and weight loss. Axial CT image demonstrates distension of the stomach and duodenum, with a narrowed distance between the aorta and SMA (*arrow*).

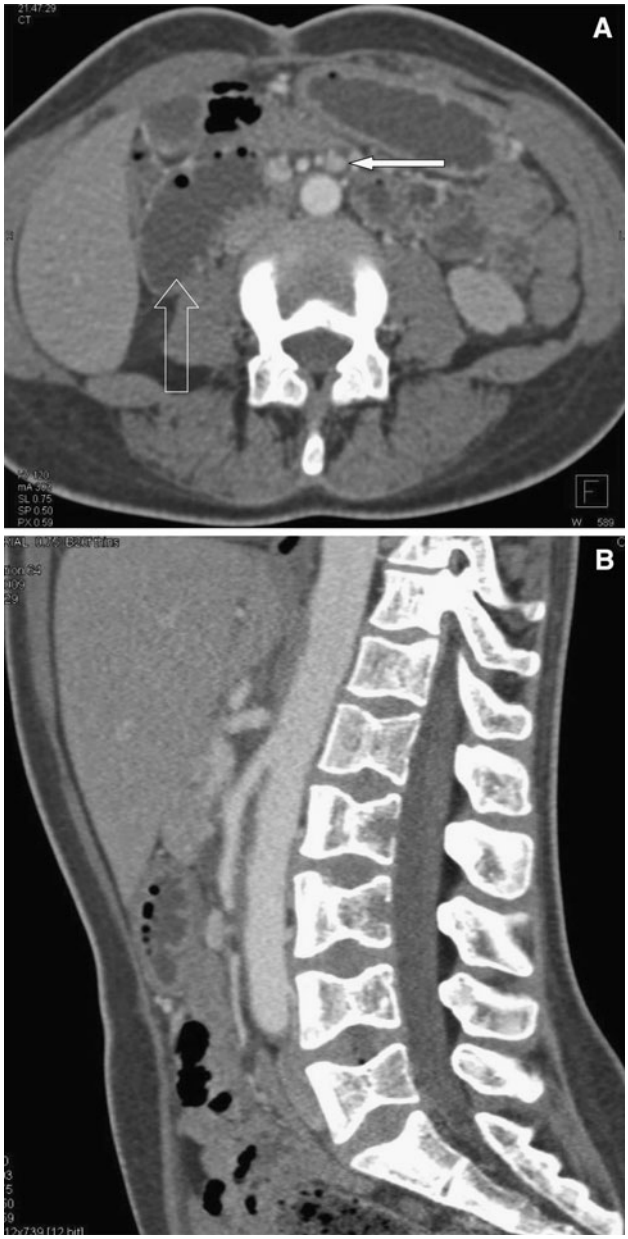

Fig. 9. A 26-year-old female with sickle cell disease and chronic abdominal pain, nausea, and vomiting. She was noted to have a dilated proximal duodenum to the level of the SMA on both endoscopy and an upper GI fluoroscopic study. Axial CT image (**A**) demonstrates a narrowed distance between the aorta and SMA (*arrow*), with dilatation of the proximal duodenum (*open arrow*). Sagittal CT image (**B**) demonstrates a narrowed aortomesenteric angle. A nuclear medicine gastric emptying study subsequently showed markedly delayed gastric emptying.

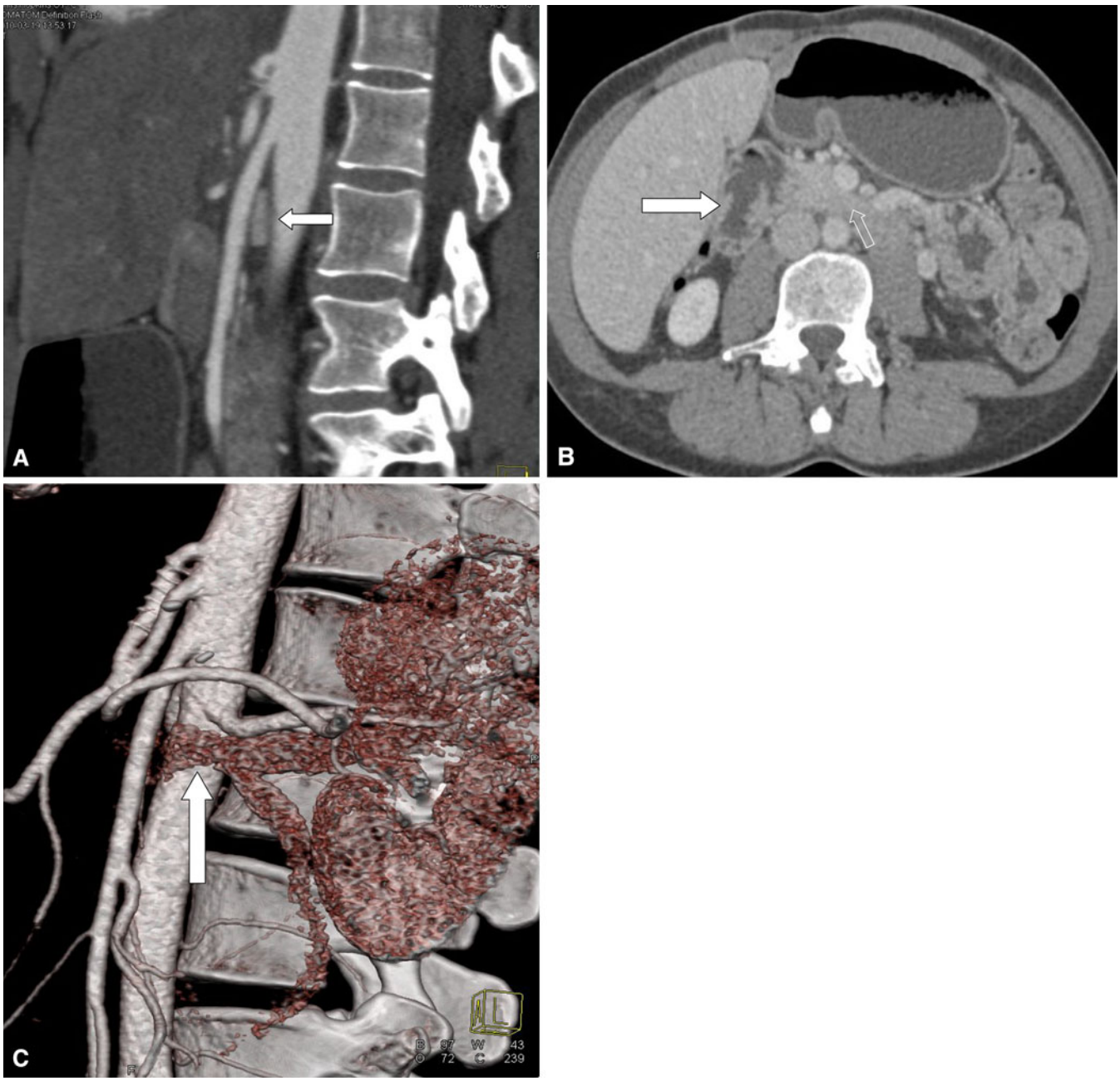

Fig. 10. A 35-year-old female who presented with gradually increasing episodes of nausea, vomiting, and abdominal pain over the course of 6 years, with multiple visits to different emergency rooms. Her symptoms were initially attributed to her gallbladder, but her symptoms did not improve after a cholecystectomy. Sagittal CT image (**A**) demonstrates a narrowed aortomesenteric angle, with compression of the duodenum (*arrow*). Axial CT image (**B**) demonstrates a dilated duodenum (*arrow*) and stomach, with narrowing of the duodenum between the aorta and SMA (*open arrow*). Volume-rendered image (**C**) demonstrates a dilated left renal vein compressed behind the SMA (*arrow*).

Fig. 11. A 27-year-old female with chronic right lower quadrant pain and bloating. Axial CT image (**A**) demonstrates a markedly dilated stomach, while sagittal CT image (**B**) demonstrates a decreased aortomesenteric angle and distance, with compression of the transverse duodenum (*arrow*).

Conclusion

SMA syndrome can present with a number of characteristic findings on CT. While none of these findings are absolutely specific, their presence in the setting of a classic clinical history can strongly suggest the diagnosis of SMA syndrome.

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