

CASE REPORT

Superior Mesenteric Artery Syndrome as Rare Cause of Abdominal Pain: An Ultrasound Case Study

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ABSTRACT

Superior mesenteric artery (SMA) syndrome is a rare disease that is often overlooked on routine abdominal ultrasound. This article describes two cases of SMA syndrome that were initially diagnosed on ultrasound, followed by the subsequent CT angiography that confirmed the diagnosis. This article also discussed the ultrasound diagnosis criteria and treatment options of SMA syndrome. With additional knowledge of the clinical symptoms and ultrasound presentations of SMA syndrome, sonographers can help diagnose this condition in clinical settings.

1 | Introduction

Superior mesenteric artery (SMA) syndrome is a rare disease that is often overlooked on routine abdominal ultrasound. It is defined as compression of the third, or transverse, portion of the duodenum between the abdominal aorta and the SMA [1]. Other names for SMA syndrome have included chronic duodenal ileus, Wilkie syndrome, arterio-mesenteric duodenal compression syndrome, and cast syndrome [2]. In this article, we present two cases of patients with abdominal pain, with initial diagnosis of SMA syndrome by routine general abdominal ultrasound examinations, then later confirmed by CT angiogram. Additional knowledge of the clinical symptoms, ultrasound presentations, and criteria for diagnosis for SMA syndrome can help sonographers to diagnose this condition in the clinical setting.

2 | History

The disease was first reported as a case report in 1842 by Czech-born Austrian physician Baron Carl Von Rokitsansky, and American physician Walter B. Lafferalso vaguely described this symptom in 1908. English surgeon David Wilkie first named it

chronic duodenal obstruction in 1921. He further detailed the pathophysiology and diagnostic findings of the disease in 1927. To honor Wilkie's accomplishment, Canadian physician F.W. Grauer first used the eponym Wilkie's (Wilke's) syndrome or duodenal ileus arterio-mesenteric ileus in 1948. Cast syndrome was used by American physician Marvin H. Dorph in 1950 to describe signs and symptoms caused by compression of the abdomen by a hip spica cast or full body cast. SMA syndrome was used finally by Kaiser et al. in 1960 [2].

3 | Clinical Presentation

SMA syndrome is a rare condition characterized by extraluminal compression of the third part of the duodenum by the SMA and the abdominal aorta, attributed to the loss of mesenteric fat pad [1]. By the year 2022, more than 730 articles with approximately 2400 cases of SMA syndrome had been reported. The incidence of SMA syndrome in the general population has been estimated at 0.013%–0.78% based on medical imaging studies, which has a female predominance over males with a ratio of 3:2 [3]. Patients commonly have symptoms of postprandial abdominal pain, nausea, anorexia, weight loss, and vomiting [4].

4 | Anatomy and Pathophysiology

The SMA is the second major branch of the abdominal aorta, providing blood flow to the third portion of the duodenum, the jejunum, the ileum, the cecum, the ascending colon, and the proximal aspect of the transverse colon [5]. It originates on the anterior aspect of the aorta at the level of the L1 vertebra, then branches inferiorly at an acute downward angle, a consequence of the erect posture of humans. The third part of the duodenum passes between the SMA and the aorta, which is vulnerable to becoming compressed between the SMA anteriorly and the aorta and vertebral column posteriorly due to the acute downward angle between the SMA and aorta [6].

Various conditions can result in the narrowing of the angle or shortening of the distance between the SMA and the aorta, which predisposes patients to SMA syndrome. The duodenum is suspended in the angle by the suspensory ligament of Treitz, and the site of attachment and structure can be different for each individual person. Commonly, the duodenum crosses the vertebral column at the level of the third lumbar vertebra, but the duodenum can locate more superiorly into the vascular angle between the SMA and the aorta due to a short suspensory ligament. The duodenum can also locate more inferiorly, and the distance between the SMA and aorta at that level can also be reduced due to the anterior curvature of the spine at the L4 level [6, 7]. In addition, surgeries for spinal deformities or body casts for scoliosis can result in the narrowing of the gap between the SMA and the aorta [8, 9].

In a normal person, there is an area of adipose and lymphatic tissue around the origin of the SMA, displacing the SMA anteriorly away from the aorta, increasing the space for the duodenum to pass through, which also provides protection to the duodenum against compression [6, 10]. There are numerous medical and psychiatric conditions that can contribute to early rapid weight loss. As a result, these conditions can lead to diminished intraabdominal adipose tissue and narrowing of the distance between the SMA and the aorta. Consequently, the duodenum can be compressed in between the SMA and the aorta, resulting in functional obstruction of the duodenum (Figure 1) [10, 11].

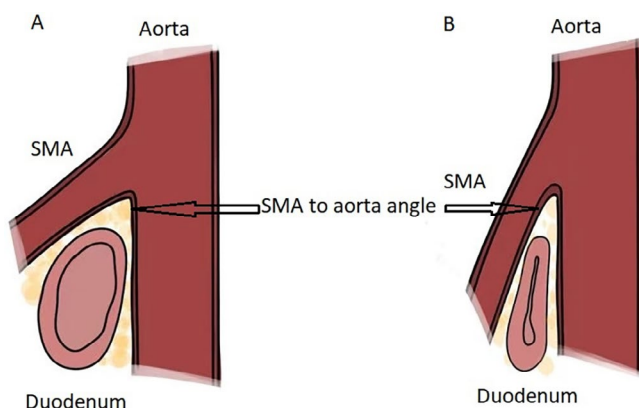


FIGURE 1 | (A) Normal SMA to aorta angle and fat pad. (B) Loss of fat pad, reduced SMA to aorta angle causes compression of duodenum.

5 | Ultrasound Method

For the evaluation of SMA syndrome, the patient is positioned in a supine position. Low frequency (3–5 MHz) curved transducers with abdominal vascular imaging settings are commonly used. The probe should be placed in the midline of the abdomen, at the level of the epigastric region.

The long axis of the proximal abdominal aorta should be demonstrated with the origin of the SMA. B-mode, color Doppler, and spectral Doppler evaluation of the abdominal aorta and SMA are included. The angle between the SMA origin and aorta should be measured, and the distance between the SMA and aorta should be measured at the level where the duodenum passes between them [12], as shown in Figures 2 and 3A,B. Ultrasound diagnosis criteria of SMA compression syndrome is a SMA to aorta angle less than 25° and SMA to aorta distance less than 8–10 mm. In the general population, the SMA to aorta angle is between 25° and 65° and SMA to aorta distance is between 10 and 28 mm [13].

5.1 | Case Studies

5.1.1 | Case 1

A 26-year-old female patient presented with abdominal pain and postprandial nausea. Routine abdomen ultrasound was performed with no cause of the pain identified. Upon further investigation, the SMA to aorta angle was reduced to 6° (Figure 4) and SMA to aorta distance was reduced to 3.4 mm on ultrasound (Figure 5). No focal stenosis was identified in the origin and proximal segment of the SMA (Figure 6). There was also the appearance of a dilated stomach and compression of the duodenum between the SMA and aorta (Figure 7), which led to the ultrasound diagnosis of SMA syndrome. Follow-up CT angiography was performed. CT findings included compression of the third part of the duodenum between the SMA and aorta (Figure 8), reduced aorta to SMA angle to 6.7° (Figure 9) and aorta to SMA distance to 3.3 mm, respectively (Figure 10); the stomach and duodenum proximal to the third part were also distended, which confirmed the diagnosis of SMA syndrome.



FIGURE 2 | The proximal abdominal aorta SMA origin in long axis.



FIGURE 3 | (A) The angle between SMA origin and aorta. (B) The distance between SMA and aorta.

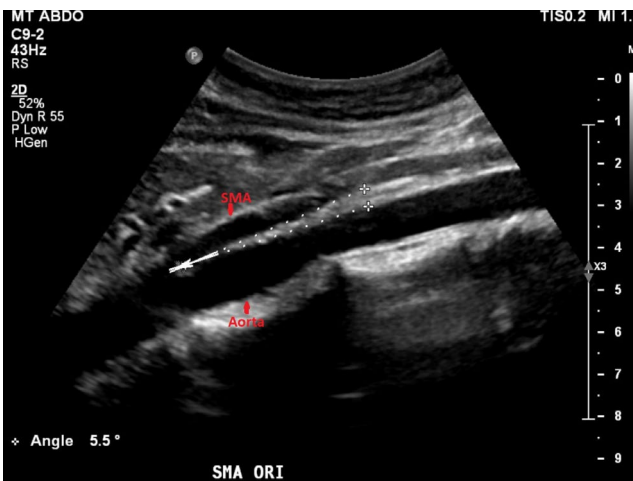


FIGURE 4 | Case 1 SMA to aorta angle.

5.1.2 | Case 2

A 34-year-old female patient presented with early satiety, abdominal pain, and postprandial nausea. Routine abdominal ultrasound was performed with no cause of the pain identified. Upon further investigation, the SMA to aorta angle was reduced

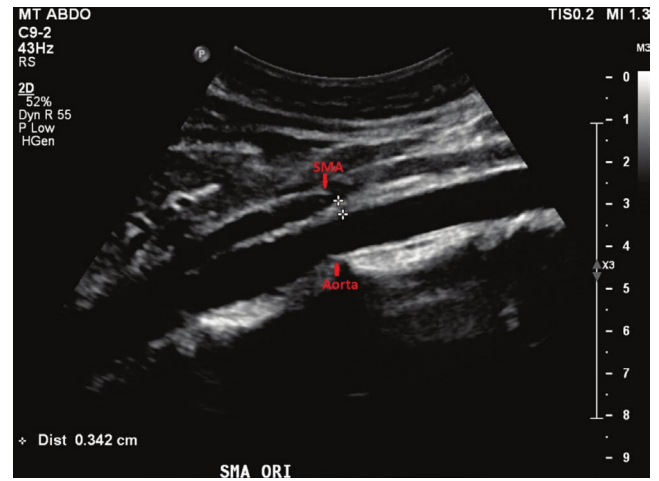


FIGURE 5 | Case 1 SMA to aorta distance.

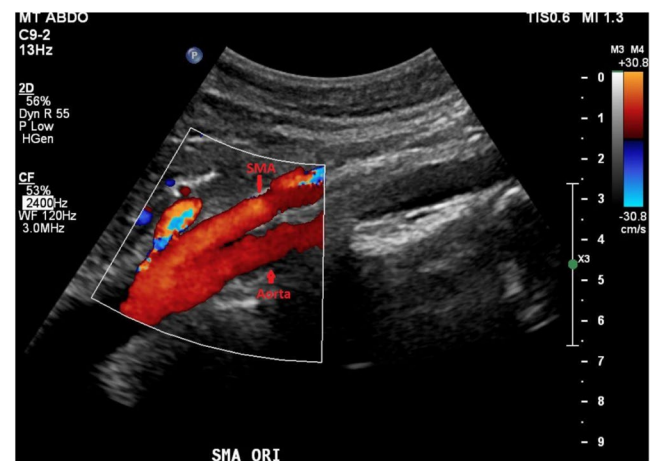


FIGURE 6 | Case 1 Color Doppler demonstrated no focal stenosis at SMA origin.

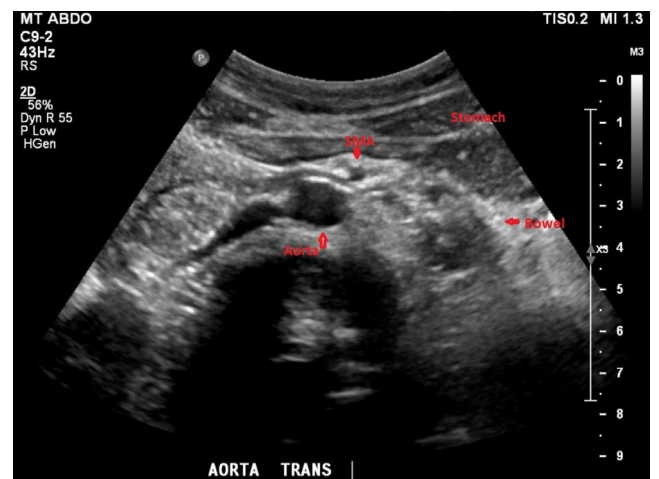


FIGURE 7 | Case 1 Compression of duodenum between SMA and aorta.

to 15.8° (Figure 11), and the SMA to aorta distance was reduced to 3.8mm on ultrasound (Figure 12). No focal stenosis was identified in the origin and proximal segment of the SMA

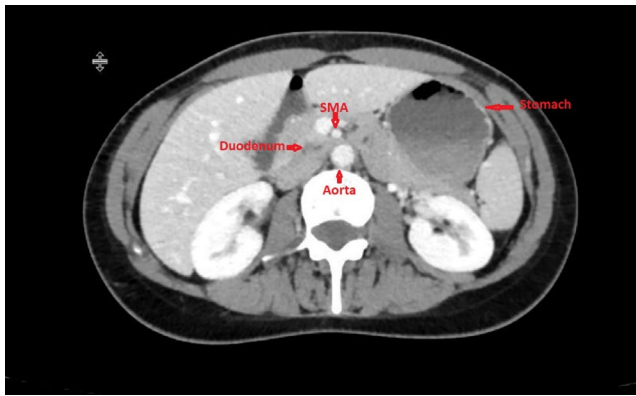


FIGURE 8 | Case 1 Compression of the duodenum between SMA and aorta.

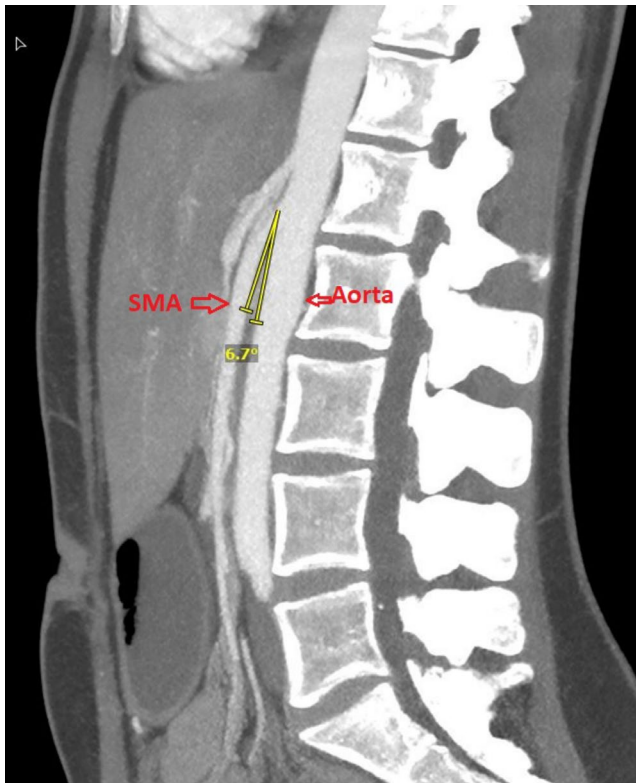


FIGURE 9 | Case 1 Aorta to SMA angle reduced to 6.7°.

(Figure 13). There was also the appearance of a dilated stomach and compression of the duodenum between the SMA and aorta (Figure 14), which led to the ultrasound diagnosis of SMA syndrome. Follow-up CT angiography was performed. CT findings included compression of the third part of the duodenum between the SMA and aorta (Figure 15), reduced aorta to SMA angle to 12.7° (Figure 16) and aorta to SMA distance to 4.3 mm, respectively (Figure 17). The stomach and duodenum proximal to the third part were also distended, which confirmed the diagnosis of SMA syndrome.

6 | Discussion

Examination of SMA syndrome is often not part of routine abdominal scanning due to the rarity of the disease. Diagnosis

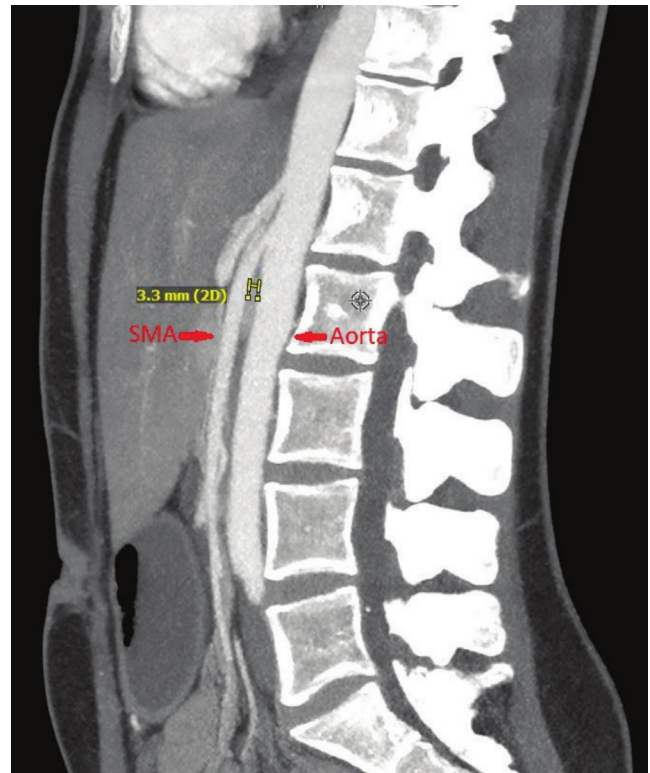


FIGURE 10 | Aorta to SMA distance reduced to 3.3 mm.



FIGURE 11 | Case 2 SMA to aorta angle.

of this condition is frequently delayed, resulting in ineffective symptomatic therapies and inappropriate investigations [10]. For patients who experience recurrent or refractory upper digestive symptoms, particularly in patients with low body mass index and who had negative routine abdominal ultrasound examinations, SMA syndrome should be considered as a potential diagnosis [14]. The SMA, aorta, and possible adjacent bowel dilation should be examined for signs of compression. The sonographer should also obtain the patient's clinical history and symptoms in detail, which is also crucial in the diagnosis of SMA syndrome.

Once the reduced SMA to aorta angle is confirmed, in addition to diagnosis of SMA syndrome, the left renal vein should also be

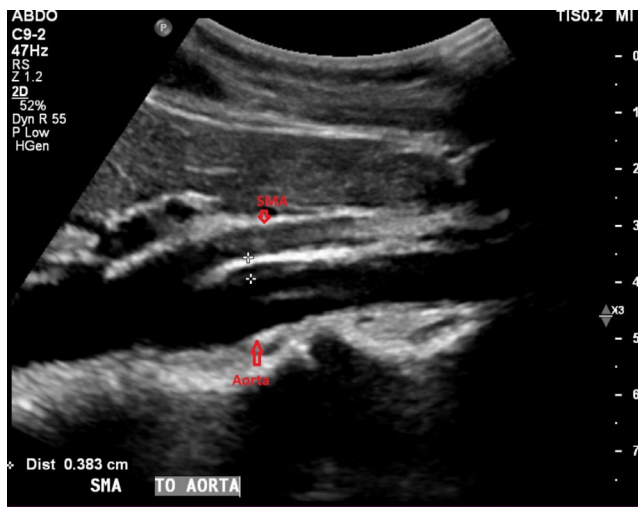


FIGURE 12 | Case 2 SMA to aorta distance.

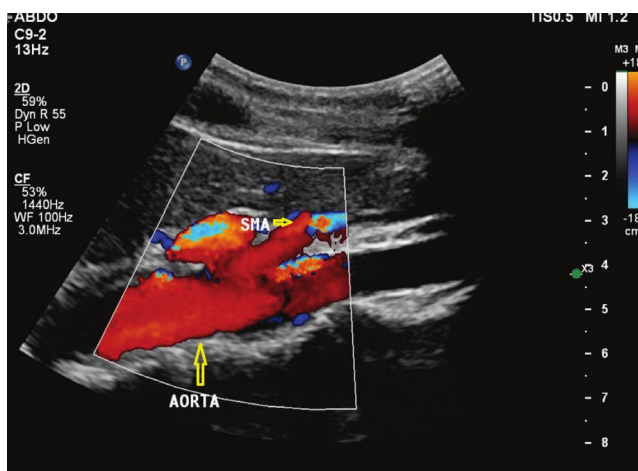


FIGURE 13 | Case 2 Color Doppler demonstrated no focal stenosis at SMA origin.



FIGURE 14 | Case 2 Compression of duodenum between SMA and aorta.

checked for potential co-existence of SMA and nutcracker syndrome, as the etiology of anterior nutcracker syndrome is similar to SMA syndrome. Commonly, the left renal vein passes in between the SMA and aorta; anterior nutcracker syndrome can also

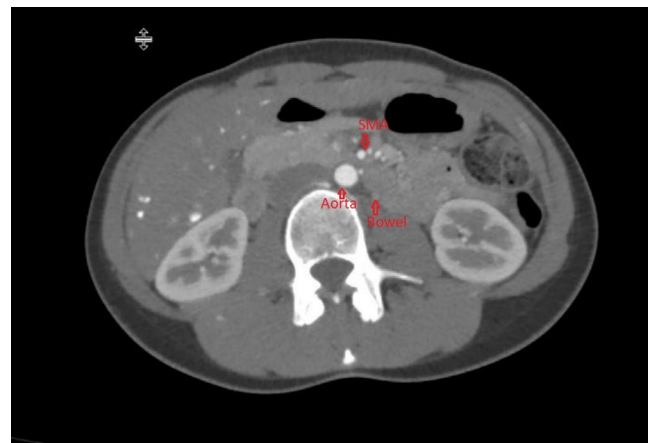


FIGURE 15 | Case 2 Compression of the duodenum between SMA and aorta.

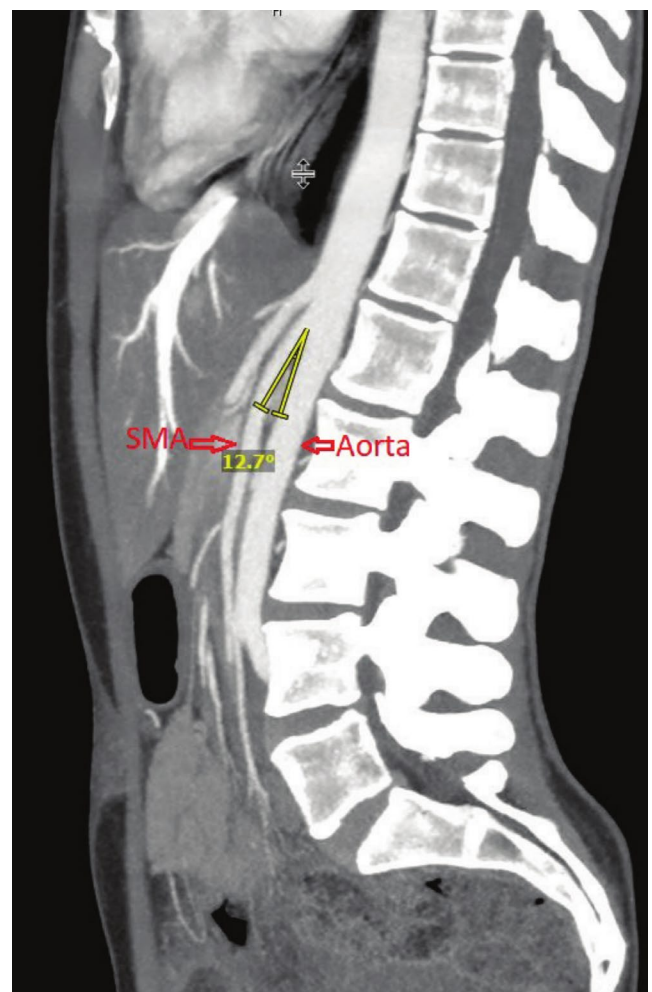


FIGURE 16 | Case 1 Aorta to SMA angle reduced to 12.7°.

be caused by the decreased angle between the SMA and the aorta, which causes external compression of the left renal vein [15].

After initial findings of SMA syndrome on ultrasound, additional imaging such as CT angiogram can be performed to confirm the diagnosis. The key findings for the diagnosis on CT include compression of the third portion of the duodenum, with



FIGURE 17 | Aorta to SMA distance reduced to 4.3 mm.

upstream severe dilatation of proximal duodenum and stomach, SMA to aorta angle less than 22° ; and SMA to aorta distance less than 8 mm. Other differential diagnoses including small bowel obstruction, annular pancreas, tumors, inflammatory lesions, aneurysms, or mesenteric ischemia can also be ruled out with contrast-enhanced CT [16]. Conventional upper gastrointestinal barium studies can also be considered in the diagnosis of SMA syndrome, although the findings are not specific when compared to CT angiography. This is due to duodenal dilatation not always being present, and slight dilatation that might be overlooked [17].

7 | Treatment

Treatment of SMA syndrome often starts with non-surgical medical management, which can include fluid equilibration, electrolyte management, increased caloric intake, parenteral nutrition, and posture therapy. For example, patients can eat small portion meals more frequently during the day and lie in the left lateral decubitus position to improve symptoms. If successfully managed, the patient should experience weight gain, and the size of the duodenal fat pad can increase, which relieves compression of the duodenum [4, 18]. However, patients can fail conservative treatment and require further surgical intervention. The surgical options include gastrojejunostomy, which connects part of the stomach to the jejunum but has risks of stomal ulceration; Strong procedure, which divides the ligament of Treitz to mobilize the duodenum but has a fail rate of 25%; trans-abdominal duodenojejunostomy; and laparoscopic duodenojejunostomy, which connects the duodenum and jejunum [19–21]. Laparoscopic duodenojejunostomy currently is the standard and most favorable operation with the best clinical outcome and success rate [21, 22].

8 | Conclusion

SMA syndrome is a rare disease that is often overlooked on routine abdominal ultrasound examination due to various reasons. Additional knowledge of SMA syndrome combined with detailed clinical history can help the sonographer to diagnose this vascular compression syndrome from ultrasound examination. Following the initial diagnosis of ultrasound, additional medical imaging, for example, CT, can help to confirm the diagnosis, which leads to a change in the clinical management of the patient.

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Ethics Statement

The study was approved by Management of Precise Radiology, Victoria, Australia.

Consent

Recorded written consents were obtained from the patients involved for the purpose of publication of this case report and any accompanying images.

Conflicts of Interest

The author declares no conflicts of interest.

Data Availability Statement

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

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