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CASE REPORT

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Coeliac artery compression syndrome as a rare cause of recurrent abdominal pain

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Abstract

Coeliac artery compression syndrome is a rare cause of recurrent abdominal pain. This article describes two cases of coeliac artery compression syndrome that initially diagnosed on ultrasound. Follow by the subsequence CT or MRI angiography that confirmed the diagnosis. This article also discussed the ultrasound diagnosis criteria, and treatment options of coeliac artery compression syndrome. With additional knowledge of the clinical symptoms and ultrasound presentations of coeliac artery compression syndrome can help sonographers to diagnose this condition in the clinical settings.

KEYWORDS
general, vascular

1 | INTRODUCTION

Coeliac artery compression syndrome (CACS), also known as median arcuate ligament syndrome, Dunbar syndrome, or Harjola-Marable syndrome, is a rare condition that results from the coeliac trunk being compressed by the median arcuate ligament, leading to coeliac artery stenosis. It has a reported incidence of 2 per 100,000 population. It is commonly seen in females with a female to male ratio of 4:1, and commonly seen in young patients with most symptomatic cases occurring between the ages of 30 and 50 years.[1] Patients with this condition can present with chronic, recurrent abdominal pain, nausea, and vomiting.[2] CACS is commonly diagnosed with computed tomography angiography (CTA) or magnetic resonance angiography (MRA) but is often overlooked on initial abdominal ultrasound scans.

In this article, we present two cases of patients with ongoing chronic epigastric abdominal pain, with initial diagnosis of CACS by routine general abdominal ultrasound examinations, then later confirmed by CTA or MRA. CACS is often overlooked during ultrasound examination. Additional knowledge of the clinical symptoms and ultrasound presentations of CACS can help sonographers to diagnose this condition in the clinical settings.

2 | ANATOMY

The anatomical compression of the coeliac artery was first reported by Lipshutz in 1917. CACS was then first described by Harjola in 1963[3] and Dunbar described the first clinical study on CACS in 1965.[4]

The median arcuate ligament is a fibrous arch that unites the diaphragmatic crura forming the anterior arc of the aortic hiatus, which the aorta, thoracic duct, and azygos vein pass through. The coeliac trunk is a major branch of the abdominal aorta, originating anteriorly near the level of the diaphragm and usually in close proximity to the median arcuate ligament, as shown in Figure 1.[5,6] However, due to variation in positioning of both the coeliac trunk and the diaphragm, up to 24% of the population have the ligament positioned more inferiorly relative to the coeliac artery. These results in various levels of compression, and approximately 1% of these individuals can experience severe levels of compression. The extent of compression typically varies with respiration, with highest degrees of compression occurring at the end of expiration. Then relief of compression can occur during inspiration or in upright position, as the coeliac artery descends in the abdominal cavity, moving away from the median arcuate ligament.[7]

The aetiology of abdominal pain from CACS is unclear. There is a theory that CACS leads to ischaemia of the gastro-intestinal system due to

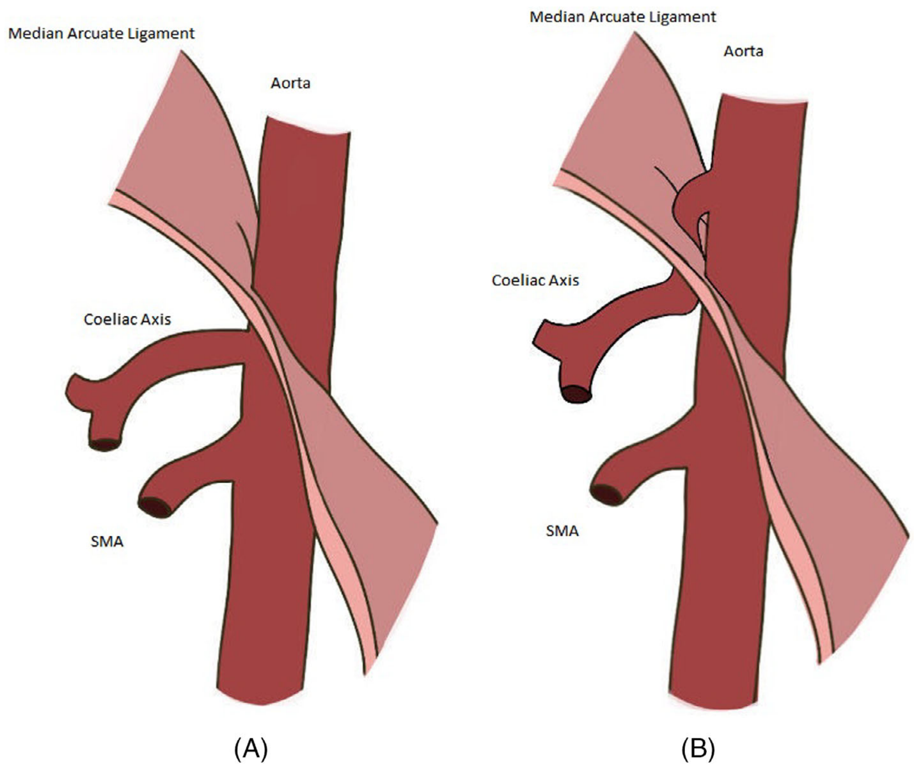


FIGURE 1 Anatomy of the median arcuate ligament in normal position (A) and inferiorly located causing compressions of coeliac axis (B).

impaired flow secondary to compression, with an alternative theory of the compressive effect on the coeliac plexus resulting in neuropathic changes.[8]

3 | ULTRASOUND METHOD

For the evaluation of CACS, the patient is positioned in a supine position. Lower frequency (3–5 MHz) transducers with abdominal vascular imaging settings are commonly used. The probe should be placed in the mid line of abdomen, at the level of epigastric region. The long axis of the proximal abdominal aorta should be demonstrated with the coeliac axis origin and the superior mesenteric artery (SMA) origin bifurcate from aorta anteriorly. B-mode, colour Doppler and spectral Doppler evaluation of the abdominal aorta, coeliac axis, superior and inferior mesenteric arteries are included. As shown in Figure 2A–C. A sample gate of 1.5–2 mm should be used to obtain spectral Doppler traces from the related vessels, with velocity measurements obtained with angle correction of 60° or less.[9]

Spectral Doppler samplings are performed in the aorta at the level of the mesenteric arteries and at the origin and proximal segments of the coeliac artery, SMA, and inferior mesenteric artery (IMA), with the spectral sampling of the coeliac artery to be obtained during both inspiration and expiration with the patient in a supine position.[9,10] Scans following the coeliac artery should also be performed with the patient in the erect position.[10] The coeliac artery descends further into the abdominal cavity when the patient is in an erect position during inspiration, resulting in an even more vertical orientation, and as a result, the arcuate ligament compression can be relieved.[11]

3.1 | Normal coeliac Axis

Commonly used ultrasound criteria that supports the diagnosis of CACS, is elevated coeliac artery peak systolic velocities with deep expiration of greater than 200 cm/s. This criterion has a reported sensitivity and specificity of 75% and 89%, respectively, in detecting a stenosis of at least 70%.[10,12,13] Other research has shown that the combination of a maximum expiratory PSV of over 350 cm/s and a coeliac axis to aorta deflection angle greater than 50 degrees seems to be a most reliable indicator for CACS.[14]

4 | CASE 1

A 25-year-old female patient presented for a general abdominal ultrasound with a clinical suspicion of gallstones/biliary pathology. From the general abdominal ultrasound examination, no hepatobiliary pathology was identified. The patient mentioned she had chronic epigastric pain at the same region and has had multiple negative abdomen ultrasound examinations, and other negative pathology test, which prompted further evaluation. When using colour Doppler imaging to exam the region of interest, aliasing was observed at the coeliac axis origin (Figure 3), the spectral Doppler demonstrated elevated velocity at the level of coeliac axis origin, which is above 400 cm/s (Figure 4A,B), and the deflection angle increased to above 50 degrees (Figure 5). Additional abdominal Doppler examination demonstrated sonographic signs of CACS, and a CTA was performed. As shown in Figures 6 and 7A,B, CTA demonstrated proximal coeliac artery narrowing with hooked shaped configuration by the median arcuate

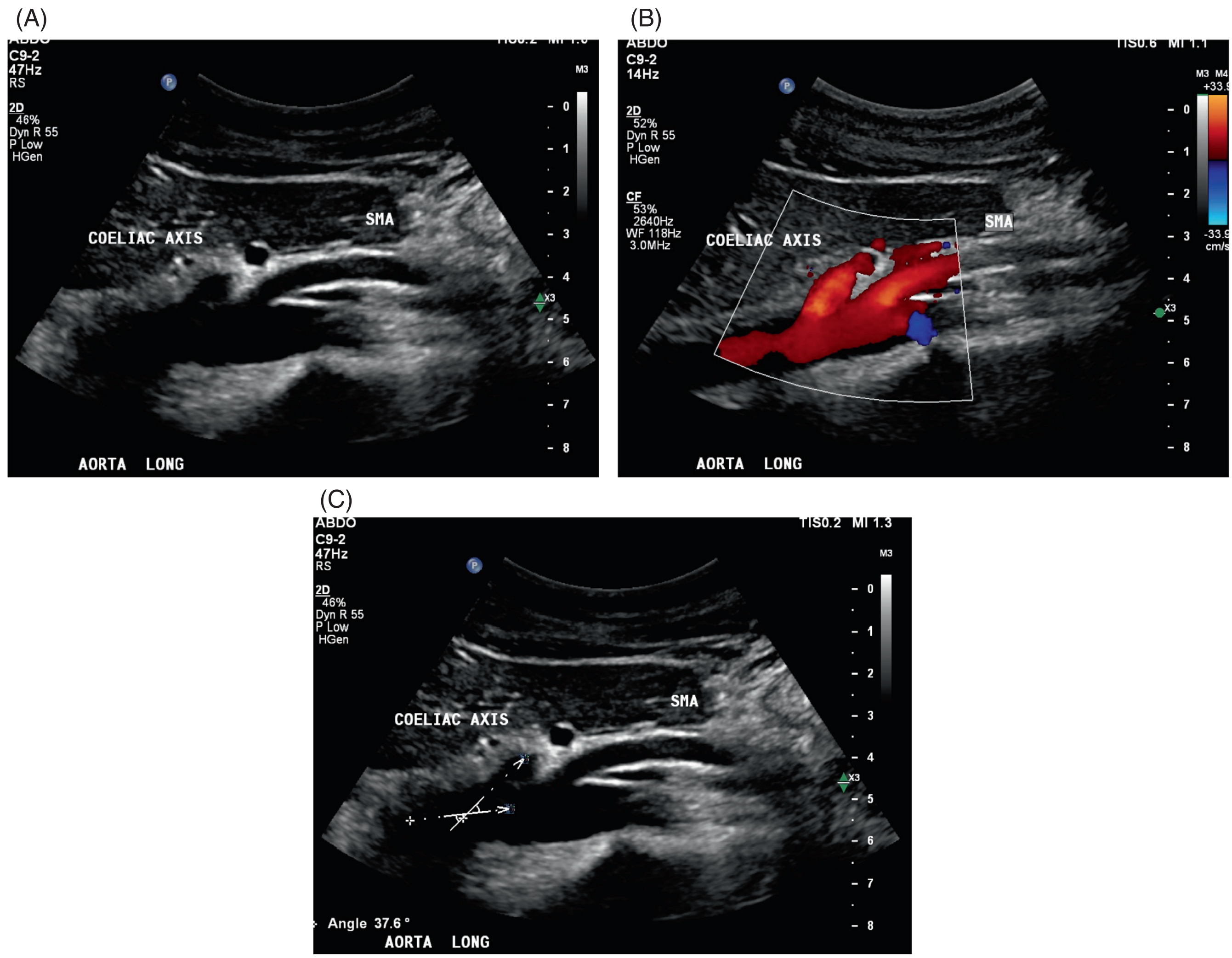
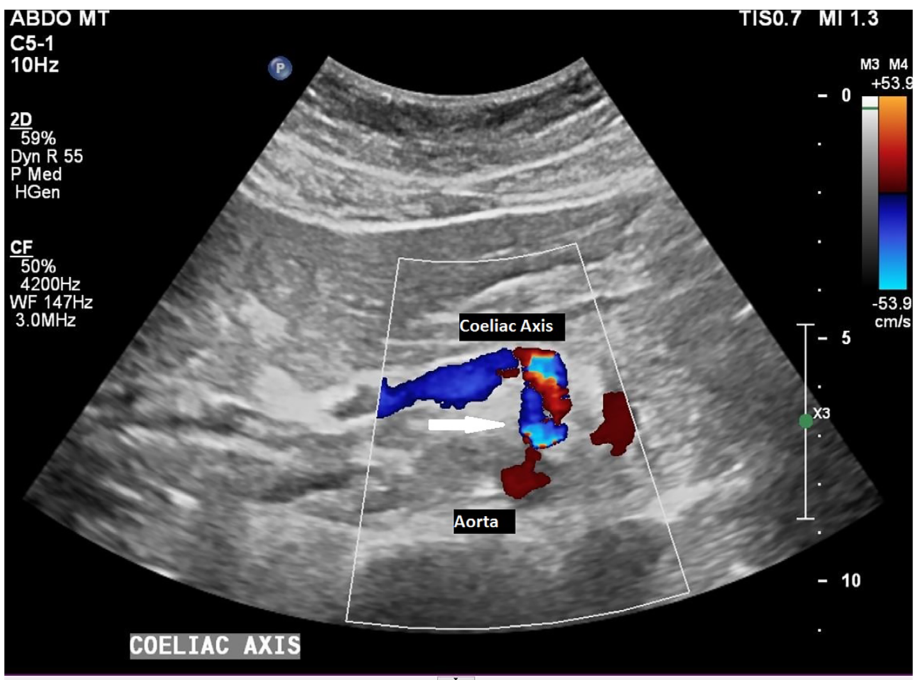


FIGURE 2 (A) Long axis of aorta with coeliac axis origin and superior mesenteric artery (SMA) origin bifurcate from the aorta anteriorly. (B) Normal long axis of aorta with coeliac axis origin and SMA origin in colour Doppler. (C) Normal coeliac axis to aorta angle.

FIGURE 3 Colour Doppler of the Coeliac Axis Origin. The aliasing area is seen at the coeliac axis origin (white arrow).



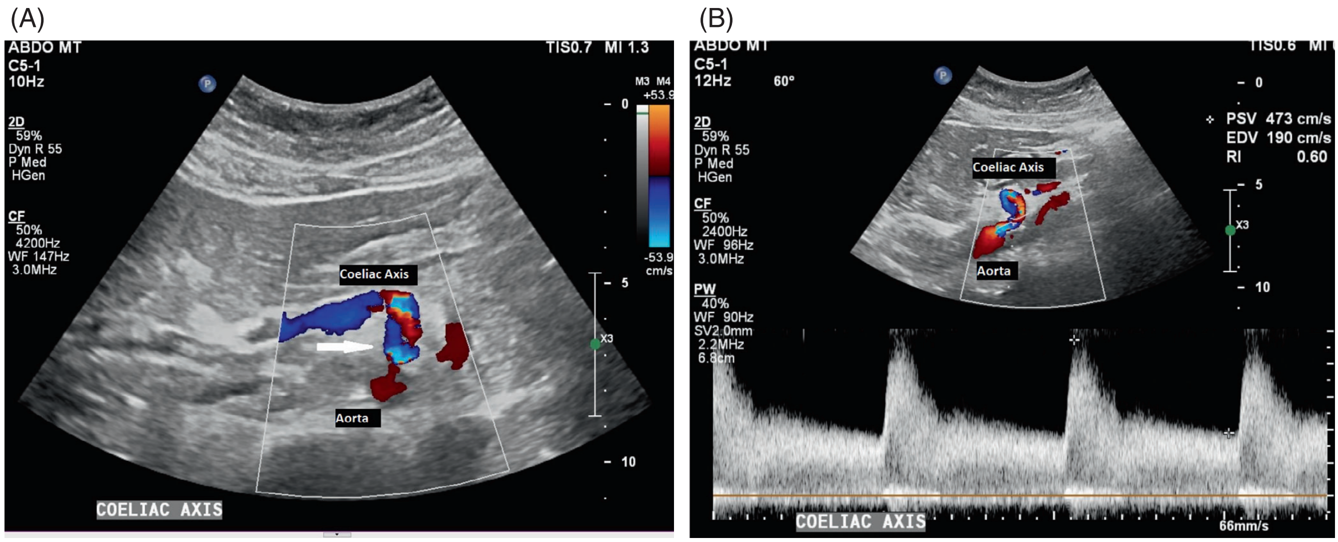


FIGURE 4 (A) Colour Doppler of the Coeliac Axis Origin. (B) Spectral Doppler trace of the Coeliac Axis Origin.

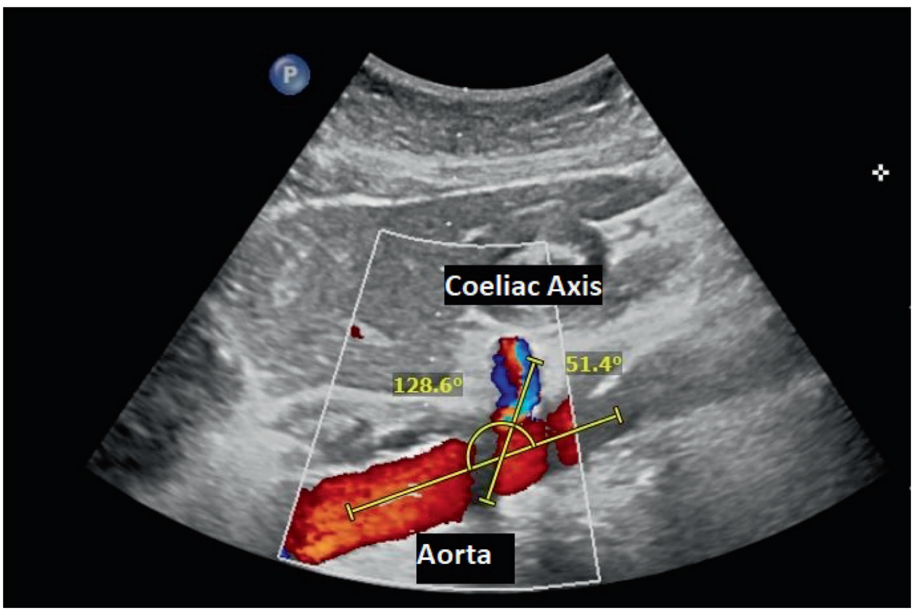


FIGURE 5 Coeliac axis to aorta deflection angle.

ligament, which in combination with patient's clinical symptoms, confirmed the diagnosis of CACS.[15]

5 | CASE 2

A 30-year-old female patient presented for abdominal ultrasound with clinical history of chronic abdominal pain, nausea and vomiting. Aliasing artefact was seen at the coeliac axis origin (Figure 8), which promoted further elevation with spectral Doppler. Increased PSV at the coeliac axis origin was observed during expiration (Figure 9). The coeliac axis to aorta angle was measured, for which there was an increased coeliac axis to aorta angle of 52.4 degrees (Figure 10). The PSV of coeliac axis returned to normal when patient moved to an erect position. (Figure 11). These ultrasound findings indicated CACS, and a MRA was performed, which confirmed the finding.

As shown in Figures 12–15, MRA images demonstrated proximal coeliac artery narrowing with hooked shaped configuration by the median arcuate ligament, combined with associated clinical syndrome, confirming the diagnosis.

6 | DISCUSSION

In these cases, both patients had chronic abdominal pain, with previous negative abdomen ultrasounds with no cause of pain identified.

CACS is often overlooked by physicians and sonographers, due to the lack of awareness of this disease and lack of specific symptoms, which is often misdiagnosed as psychogenic abdominal pain.[16] However, research has shown that 13%–50% of the population have asymptomatic coeliac artery compression and there are many causes for abdominal pain.[17] CACS is considered to be a diagnosis of exclusion, and it is important that patients are evaluated for all possible etiologies of abdominal pain prior to being diagnosed with CACS.[1,17]

During ultrasound examination, it is important to look for features of CACS for patients with chronic abdominal pain and other associated symptoms. As shown in the images of the two cases, aliasing artefact at the level of the coeliac axis origin, increase of coeliac axis to aorta angle, or hooked appearance of the coeliac axis promotes further investigation with coeliac axis spectral Doppler study.

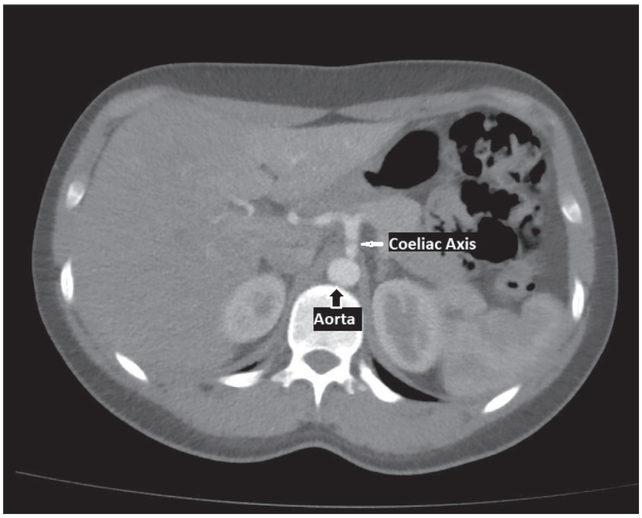


FIGURE 6 Computed tomography angiography of abdomen, axial plane at the level of the coeliac axis origin.

7 | TREATMENT

Patients with CACS that show ongoing abdomen pain and symptoms have treatment options of open surgery and laparoscopic surgery. The open surgical treatment involves open coeliac artery reconstruction. The operation involves accessing the coeliac artery through the lesser

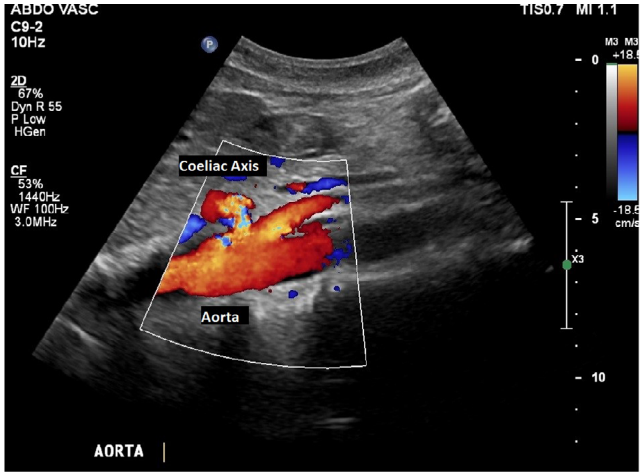


FIGURE 8 Colour Doppler of the Coeliac Axis Origin.

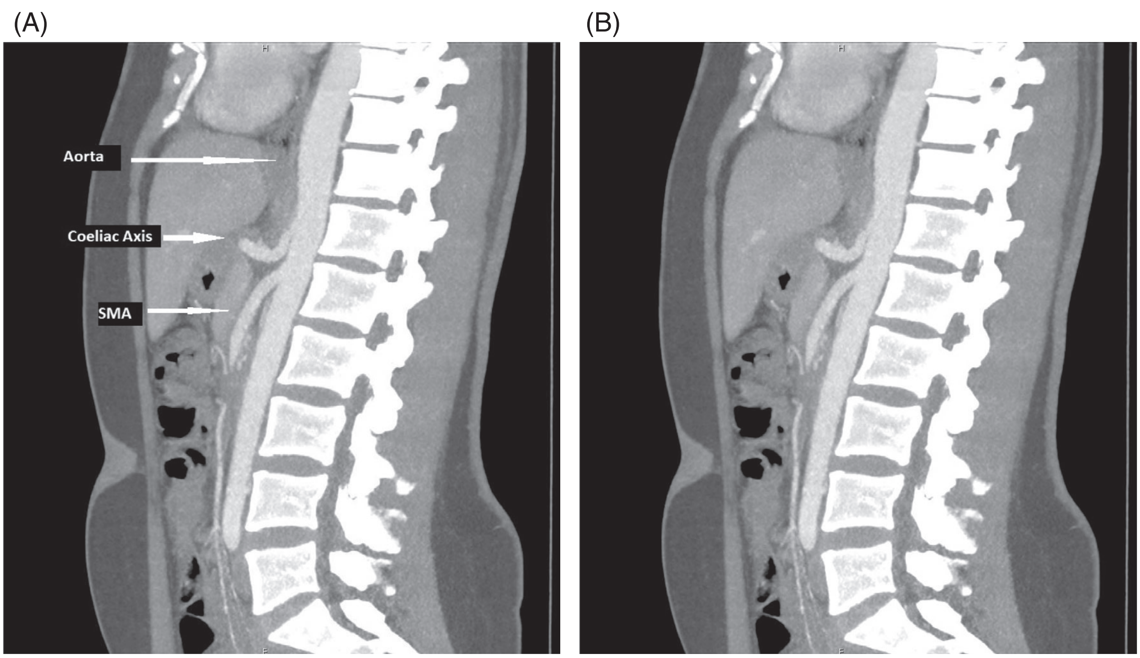


FIGURE 7 (A) Computed tomography angiography (CTA) of abdomen MIP, sagittal plane of the aorta at the level of the coeliac axis origin. (B) CTA of abdomen MIP, sagittal plane of the aorta at the level of the coeliac axis origin.

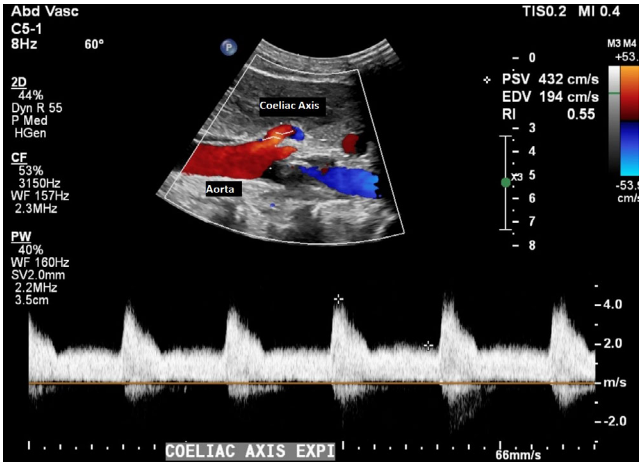


FIGURE 9 Spectral Doppler trace of the Coeliac Axis Origin during expiration.

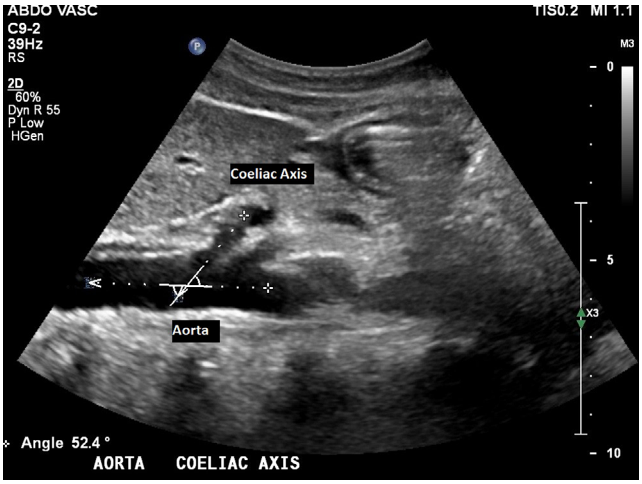


FIGURE 10 The Coeliac axis to aorta deflection angle.

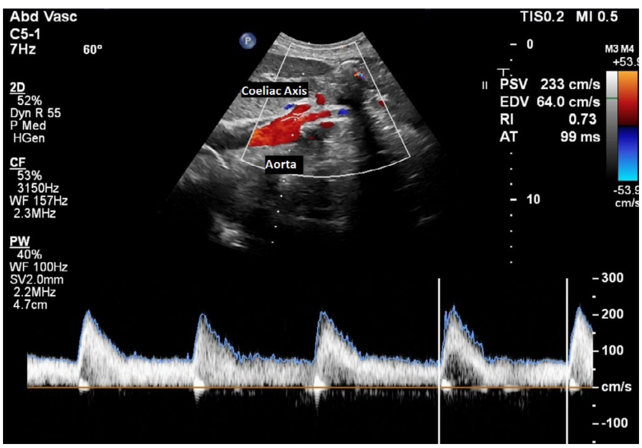


FIGURE 11 Spectral Doppler of the coeliac axis origin when patient was at the erect position, PSV return to normal.

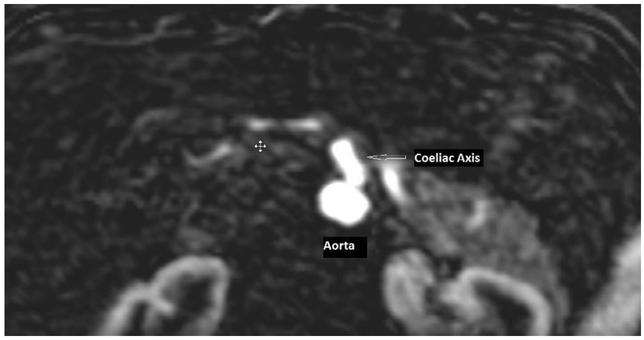


FIGURE 12 Magnetic resonance angiography of the coeliac axis in axial plane.

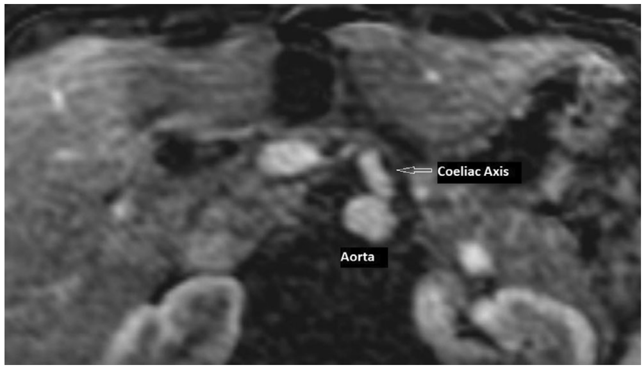


FIGURE 13 Magnetic resonance angiography of coeliac axis in axial plane.

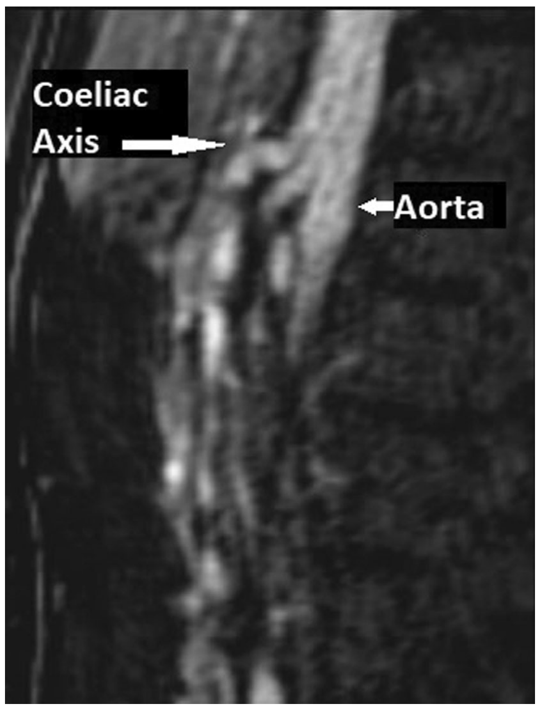


FIGURE 14 Magnetic resonance angiography of coeliac axis in sagittal plane.

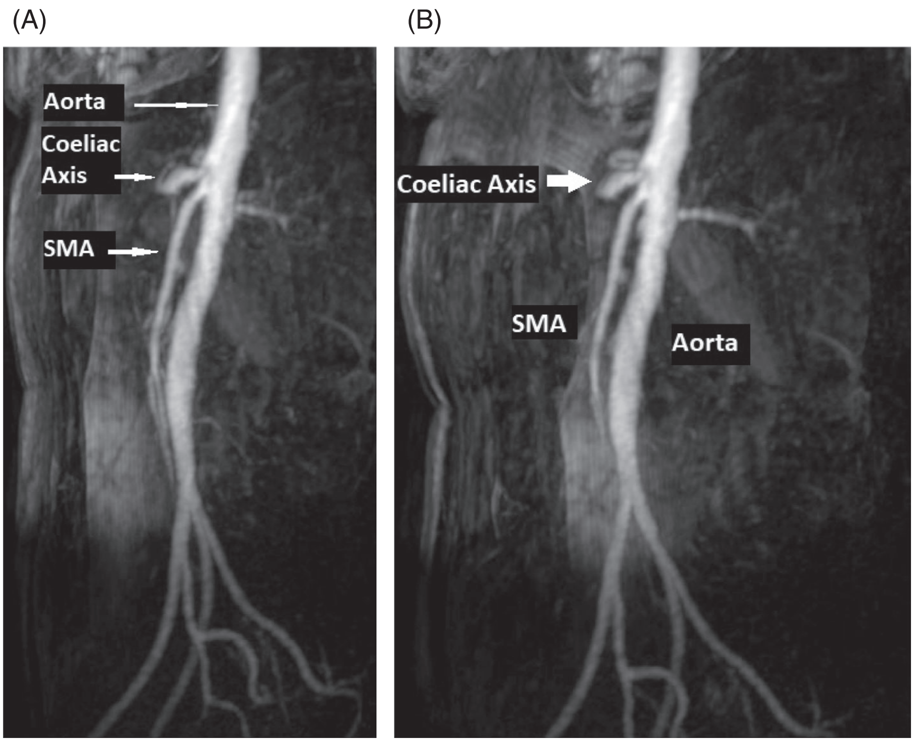


FIGURE 15 (A) Magnetic resonance angiography (MRA) 3D of coeliac axis in sagittal plane. (B) MRA 3D of coeliac axis in sagittal plane.

sac from a short upper midline laparotomy, then transecting of the median arcuate ligament and the crus of the diaphragm proximal to the coeliac artery, resulting the releasing of compression of the coeliac artery.[18] The laparoscopic treatment involves division of the right pillar of the crus to expose the aorta, then uses harmonic scalpel to divide the median arcuate ligament at the level of coeliac trunk to release the compression.[19]

Both options have shown minimal morbidity and mortality rate, and good short-term and long-term outcomes.[18–20]

8 | CONCLUSION

CACS is a rare but often overlooked cause of chronic abdominal pain. When other common causes of chronic abdominal pain are not observed, further evaluation of coeliac axis should be considered for signs of coeliac axis compression syndrome. Doppler ultrasound can be a useful and safe modality in the screening and initial diagnostic workup of patients with CACS, which can lead patients to further investigations by contrast-based CT, MR, or digital subtraction angiography.

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CONFLICT OF INTEREST STATEMENT

The author declares no conflict of interest.

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