case study

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Ultrasound Findings in Median Arcuate Ligament Syndrome

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About the Author

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Abstract

Median arcuate ligament syndrome, which is often associated with abdominal pain, vomiting, and weight loss is an unusual disease caused by extrinsic compression of the celiac artery (CA) by the median arcuate ligament. Such tension on the CA increases the chance of geometric distortion.

This is a case of a 69-year-old female with abdominal pains. Ultrasound revealed cyclical CA geometric and Doppler changes. At deep expiration, CA bends and then gradually became tortuous with a 130° trunk deflection. At this phase, Peak systolic velocity (PSV) and celiac/aortic ratio measure 287cm/s and 4:1 respectively. There is also post-stenotic flow turbulence. However, during full inspiration, the CA deflection straightens out. As a result, the PSV decreases (167 cm/s) while the flow turbulence disappears. A clearer mechanism of CA compression with the resultant hemodynamic effect is demonstrated in this case.

Keywords: Median arcuate ligament syndrome, Doppler ultrasound, celiac artery compression.

Introduction

A fibrous arch formed by the diaphragmatic crura, the median arcuate ligament can be found in an unusual anatomical location such that it compresses the CA. The compression can cause distortion in the arterial geometry thus inducing hemodynamic wall shear stress.[1,2] median arcuate ligament syndrome (MALS) is a rare disease characterized by postprandial pain, nausea, and weight loss. It is 4 times more common in females compared to males, often affecting those between the ages of 30 and 60 years.[3]

Many authors have described ultrasound findings in MALS. In literature which described ultrasound exam demonstrates elevated PSV greater than 200 cm/s, deflection angle > 50° and a retrograde common hepatic artery flow were some of the identified criteria.[3] A different study of about 364 patients with suspected MALS suggested a PSV greater than 350 cm/s in combination with a deflection angle > 50°.[4]

The published PSV criteria vary widely probably due to measurement difficulty in a tortuous vessel together with Doppler angle dependency. At the same time, adequate evaluation of dynamic arterial

distortion in the presence of artifacts and increase penetration depth is challenging. As such optimization of ultrasound technique is crucial.

Case Presentation

A 69-year-old female outpatient was sent for abdominal visceral arterial ultrasound. She had a complaint of vague epigastric pain. In a 6-hour fasting state, she was scanned supine using a Philips iU22 C5-1 [1–5 MHz] broadband convex transducer with 160 elements and 96° field of view.

In sagittal grey scale (2-D), the CA was identified just below the xiphoid process as it branches from the abdominal aorta. The transducer orientation was adjusted to demonstrate the entire length of the vessel. Time gain compensation and overall 2-D gain were then adjusted to compensate for attenuation. The image depth and focus were used to position the CA in the center of the image frame, bringing it into a focus of maximum intensity. In order to improve contrast resolution, dynamic compression (DR) was adjusted to 55 decibels (dB). Afterward, tissue harmonic imaging (THI) was used to improve spatial resolution and eliminate artifacts. At this point, spatial, contrast and temporal resolutions were optimal. Images were then obtained in orthogonal planes. The deflection angle and CA caliber were measured and documented.

After 2-D optimization and measurements, colour Doppler was used to interrogate the CA, colour scale or pulse repetition frequency was adjusted to between 2.4 and 2.7 KHz and the gain optimized to reduce artifact. Following colour optimization, there was considerable aliasing within the CA at full expiration versus inspiration. Regions of aliasing signify high flow velocities with turbulence and hence potential sites for careful spectral interrogation. Spectral Doppler interrogation was done using a 60° Doppler angle. PSV was obtained at full inspiration and expiration. The origin, mid and distal segments of the CA were sampled with special attention to aliasing regions. Cine loop was used to obtain dynamic images demonstrating the changes in the geometry of the vessel during respiration both in 2-D and in Doppler modes.

The ultrasound study revealed an elevated CA PSV measuring 287 cm/s during deep expiration with a 4:1 celiac/aortic PSV ratio. There was also colour Doppler aliasing and post-stenotic turbulence demonstrated by the spectral broadening of the waveform (Figure 1). In contrast, during full inspiration, PSV measured 197cm/s (Table 1) with no post-stenotic turbulence.

During deep expiration, the celiac trunk became tortuous with a "flexed-knee" appearance and demonstrated a 130° deflection angle. (This is the angular deviation between the expiratory and inspiratory orientation of the celiac trunk). Comparably, at full inspiration, the CA straightened out and the trunk deflection disappeared (Figure 2).

The geometric and hemodynamically significant changes are suggestive of MALS. The patient is now under the care of a vascular surgeon.

Discussion

The clinical implication of CA compression continues to be a hot debate and diagnostic workup may require different approaches depending on the protocol at different institutions. CT scans typically demonstrate a characteristic hook-shaped appearance as described by Horton, Talamini, and Fishman[5] (Figure 3). However, on ultrasound, PSV and deflection angle appear to be the leading criteria. Celiac trunk deflection greater than 50° was consistently and widely reported in many works of literature. PSV finding, on the other hand, varies (Table 2).

Saleem and Baril[3] suggested an elevated expiratory PSV > 200 cm/s and a CA deflection angle > 50° as significant criteria for MALS appearance. In this case, ultrasound findings revealed a 287 cm/s CA expiratory PSV while deflection angle measured 130°.

In another study, Gruber et al [4] compared 6 patients who have classical MALS symptoms and previous diagnosis with 20 other asymptomatic volunteers (control). They concluded that an expiratory PSV greater than 350 cm/s in combination with a deflection angle of more than 50° are significant criteria for a higher probability of MALS with a 78% and 100% sensitivity and specificity respectively. In this study, we found 287 cm/s PSV at full expiration and a 130° deflection angle. There is also significant Doppler aliasing and post-stenotic turbulence during the expiratory phase.

Tembey et al[6], on the other hand, described a case of MALS done supine and erect. They reported a 323 cm/s expiratory PSV with turbulence in the supine position and a 99 cm/s expiratory PSV in the erect position. This case, however, was done in a supine position only. We discovered an expiratory PSV measuring 287 cm/s and also flow turbulence and colour aliasing. The erect technique is important when

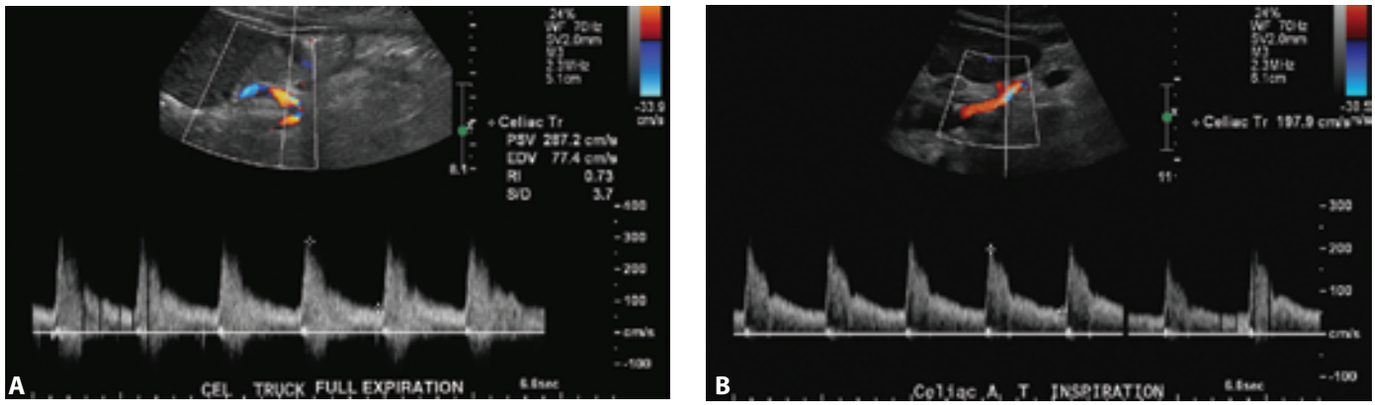

Figure 1. (A) Demonstrates elevated celiac artery PSV 287 cm/s with post-stenotic turbulence during expiration. (B) Shows a PSV of 197 cm/s during inspiration.

supine findings are ambiguous or when inspiratory and expiratory PSV are both elevated in the supine position.

AbuRahma et al[7] used receiver operated curve to correlate ultrasound accuracy with angiography. They reported that 240 cm/s PSV and celiac/aortic ratio 3.8:1 are thresholds for predicting 50–69% stenosis in the CA with 87% and 83% sensitivity and specificity respectively. A 320 cm/s threshold was also reported for predicting ≥70% stenosis. Their study was done using 150 patients. In this case, expiratory PSV was 287 cm/s while celiac/aortic ratio measured 4:1. Our findings are suggestive of 50-69% stenosis (using AbuRahma et al criteria).

Doppler angle variation could be responsible for the different PSV reported by the various authors.[8,9] When an artery is tortuous, it becomes difficult to use consistent Doppler angle. Table 2 demonstrates PSV findings from different authors. While the PSV parameter varies from one author to the next, a 240 cm/s PSV seems to be a reliable benchmark while a greater than 350 cm/s PSV carry a higher risk of disease. It is thus important to consider secondary Doppler indicators of hemodynamically significant arterial disease in the form of post-stenotic turbulence and colour aliasing. When Doppler scale, gain, and image depth are optimized, colour aliasing can be confined to regions of high flow velocities where Doppler shift exceeds Nyqvist limit.[10] Hence Doppler interrogation can be better directed.

Table 1. Doppler and Geometric Findings During Expiration Compared to Inspiration

	Expiration	Inspiration
Peak Systolic Velocity (cm/s)	287	197
Celiac/Aortic Ratio	4:1	–
Spectral Broadening	Yes	No
Colour Aliasing	Yes	No
Deflection Angle	130°	–
Tortuous	Yes	No

Table 2. Comparing Celiac Artery PSV and Deflection Angles in Different Studies

	This Case	Gruber et al	Saleem and Baril	AbuRahm et al
Expiratory Peak Systolic Velocity (cm/s)	287	>350	>200	> 240 (50–69%)
Deflection Angle	>50°	>50°	>50°	–

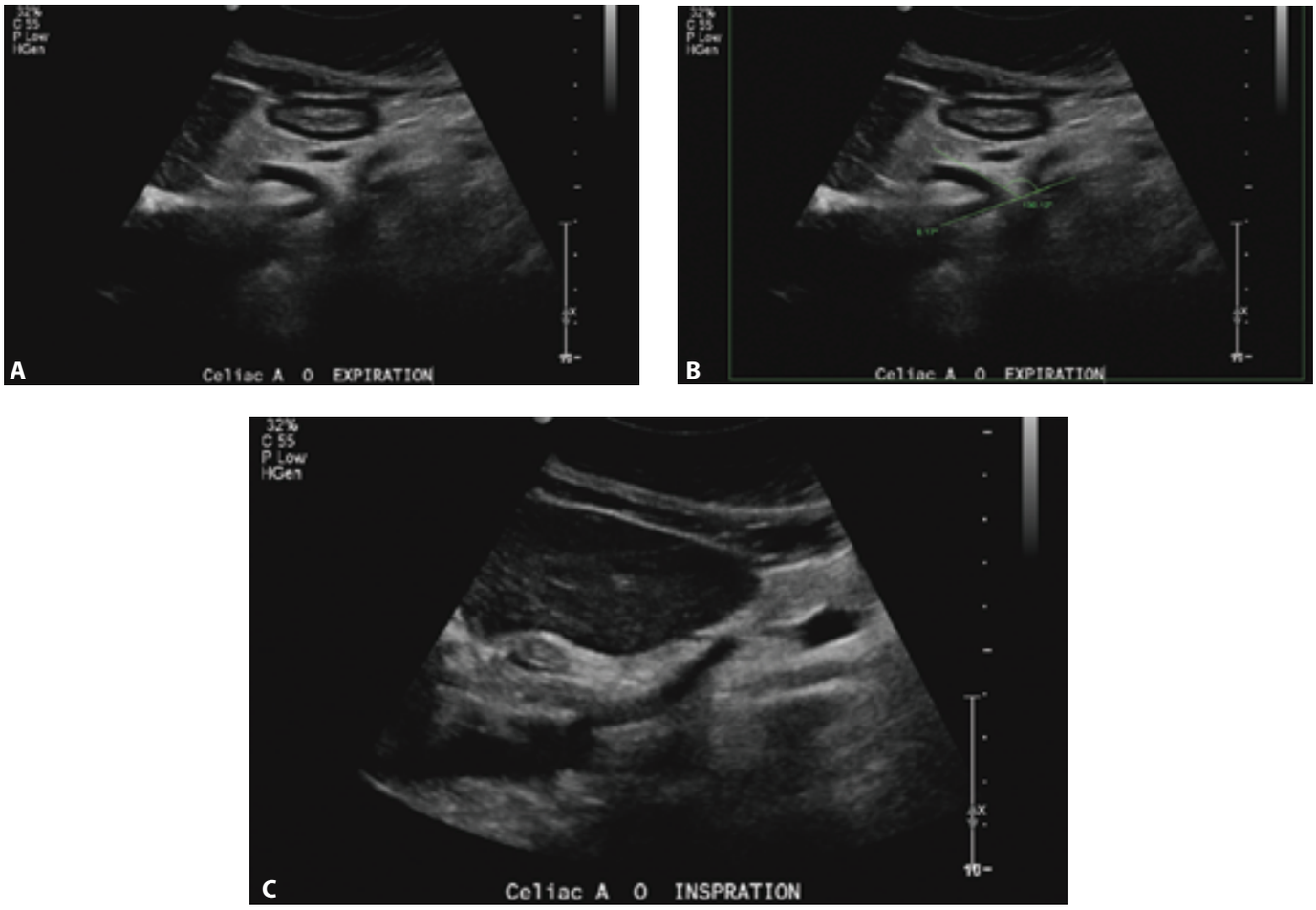

Figure 2. (A) A "flexed knee" appearance of the celiac artery at full expiration. (B) 130° deflection angle at expiration. (C) Straightened-out celiac artery at full inspiration.

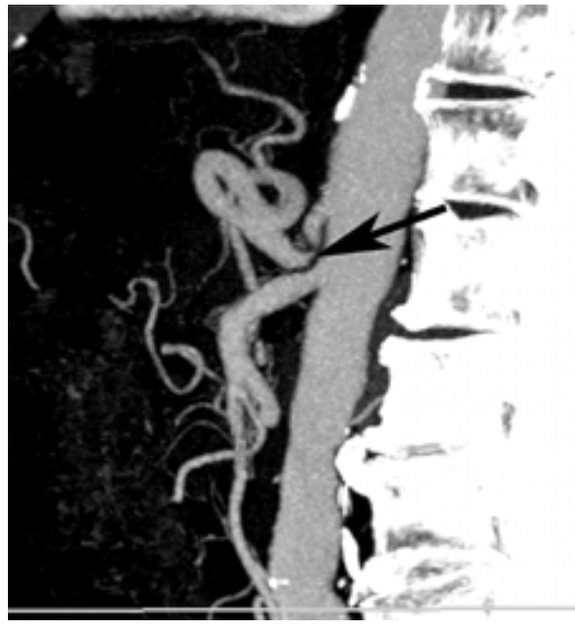

Figure 3. A computed tomography scan demonstrates filling defect in the proximal celiac artery. This is due to extrinsic compression by the arcuate ligament in a case of median arcuate ligament syndrome. The dark arrow shows the characteristic "hook-shaped" appearance.

Post-stenotic turbulence, on the other hand, encompasses chaotic, non-laminar flows that are found in regions distal to a significant narrowing. These regions usually have abnormal wall shear stress and oscillatory shear index values.[11,12] They are shown on Doppler as waveform spectral broadening.

Conclusion

Exactly how the geometric and Doppler components correspond to clinical outcome including those that will benefit from surgery is still poorly understood and the scope of such finding is beyond this case. However, ultrasound can be used as an effective tool to demonstrate hemodynamically significant CA compression. This case demonstrates a clearer mechanism of MALS, showing respiratory-induced changes. It further describes hemodynamically significant Doppler findings in the presence of geometric distortion.

Conflict of Interest.

There is no commercial interest.

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