

Evaluation of Asymptomatic Patients with Median Arcuate Ligament Syndrome (Mals) Using Color Duplex Ultrasound and Computed Tomographic (Ct) Angiography

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Abstract Median arcuate ligament syndrome is considered as rare syndrome manifested by variety of symptoms. Aim of the Work: To assess the presence of asymptomatic patients showing the typical color duplex ultrasound and CT angiographic features of MAL syndrome and the impact of these findings on the pathogenesis and management of the syndrome. Material and Methods: 200 asymptomatic patients were examined with color duplex ultrasound and further CT angiography was performed to all patients with color duplex findings of arcuate ligament syndrome. Results: 8 patients showed typical color duplex findings and CT angiographic features of MALS. Conclusion: The diagnosis of MAL syndrome shouldn't depend only on the imaging criteria and should be strongly linked to the unexplained patient symptoms after meticulous exclusion of other etiologies. The management shouldn't depend only on relieving the arterial stenosis but also on dissection of peri arterial nerve plexus.

Keywords: *arcuate ligament, color duplex sonography, computed tomographic angiography, asymptomatic*

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1. Introduction

Arcuate ligament syndrome is considered as rare syndrome manifested by variety of symptoms which include post prandial pain, nausea, vomiting, weight loss, diarrhea and upper abdominal bruit heard by auscultation [1,2,3,4].

It is caused by abnormal long diaphragmatic crura and median arcuate ligament (MAL) of diaphragm which forms the anterior aspect of aortic hiatus that cause compression of celiac artery either during expiration [5] or during inspiration and expiration [6] or caused by abnormal high origin of celiac artery compressed by the median arcuate ligament. Other etiologies include large fibrosed fused celiac ganglia that compress the celiac trunk [7,8,9]. Anomalous celiacomesentric trunk compressed by abnormally long diaphragmatic crura and MAL.

Theories which explain the source of post prandial pain and other symptoms of the syndrome postulated mesenteric ischemia and or irritation of basal ganglia [10].

The diagnosis depends upon color duplex ultrasound, computed tomographic angiography and magnetic resonance angiography [10].

The treatment is by surgical division of the MAL and dissection of celiac ganglia either by open surgery or laparoscopy, endovascular application of stent was tried

with unfavorable outcome, angioplasty of celiac artery was limited to cases with vascular damage [11,12].

2. Aim of the Work

To assess the presence of asymptomatic patients showing the typical color duplex ultrasound and CT angiographic features of MAL syndrome and the impact of these findings on the pathogenesis and management of the syndrome.

3. Material and Methods

200 patients showing no abdominal pain or any gastrointestinal symptoms were examined from march 2012 to march 2013, their age range from 10 years to 45 years old and the mean age was 25 years, they were 100 males and 100 females.

All were examined at Fayoum university hospital with color duplex Sonography using GE logiq 7 ultrasound machine (Milwaukee, WI, USA) using 3.5 Mhz and 2.5 Mhz sector probes.

The patients were examined and the angle corrected peak velocity of proximal celiac artery was estimated at maximum deep inspiration, maximum deep expiration, in neutral state and in erect position.

Color duplex diagnosis of median arcuate ligament syndrome was made if peak systolic velocity at deep

expiration was 2 folds or more than its peak systolic velocity at deep inspiration or was >2m/sec and the peak velocity was normal on erect views. Fixed stenosis was diagnosed if the peak systolic flow velocity > 2m/sec in all views [13,14].

The celiac artery flow volume /min was estimated in patients with features of celiac artery stenosis.

All positive patients who showed color duplex criteria of arcuate ligament syndrome were further examined with CT angiography using a 4-slice Toshiba MDCT scanner (Asteion). and the diagnosis of arcuate ligament syndrome was made with certainty if they showed the typical hook-shape contour of the superior aspect of celiac trunk or showed focal proximal narrowing associated with post stenotic dilation in Sagittal C.T views ± the presence of prominent peri-arterial collaterals [15,16,17].

4. Results

192 patients showed normal color duplex findings with no stenosis to the celiac trunk (Figure 1).

8 patients among the 200 patients (4%) showed typical imaging findings of MALS (Figure 2, Figure 3) with increased peak systolic velocity of blood flow > 2m/sec at deep expiration. Among them 6 cases diagnosed as expiratory compression of the celiac artery with MAL showed stenosis of celiac artery at maximum deep expiration with normal flow velocities at maximum deep inspiration and on erect views and 2 cases showed fixed stenosis at all views. All 8 patients subjected to CT angiography and showed proximal narrowing of celiac artery by post stenotic dilation or hook-shaped contour of celiac artery.

No one showed occlusion or prominent peri arterial collaterals.

Among the positive patients 2 were prepubertal, 5 patients were females and 3 were males.

Positive patients showed estimated blood flow volume per minute =690 ml/min ±30 ml at the post stenotic segment of celiac artery.

5. Figures

Figure 1 A, B, C: Normal B mode, estimated Doppler flow volume and CT angiography in patient with no imaging features of median arcuate ligament syndrome.



Figure 1A. Normal B mode ultrasound of celiac artery during inspiration and expiration with no stenosis observed. The artery descends with more vertical orientation during expiration

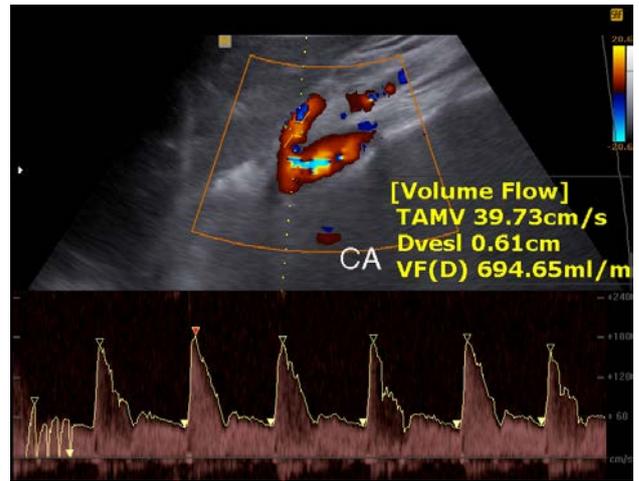


Figure 1B. Estimated flow volume of celiac of celiac artery in the same patient 694ml/min



Figure 1C. Normal CT angiography of celiac artery (white arrow) with no proximal stenosis, arterial kink or post stenotic dilation

Figure 2 A, B, C, D, E, F: Moderate degree celiac artery stenosis noted during expiration in asymptomatic patient with imaging features of median arcuate ligament syndrome.

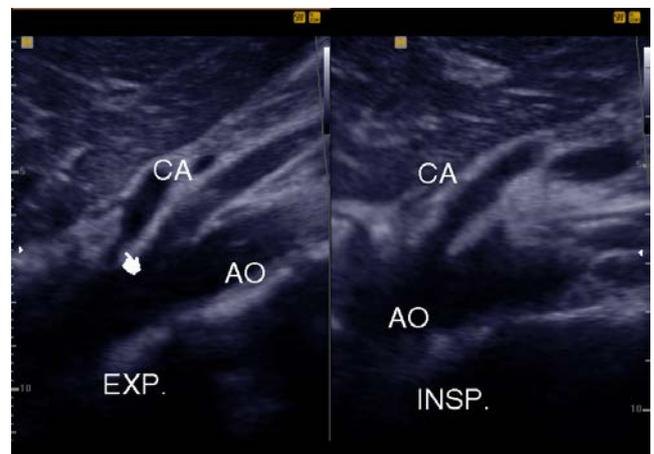


Figure 2A. B mode ultrasound showing expiratory stenosis of celiac artery

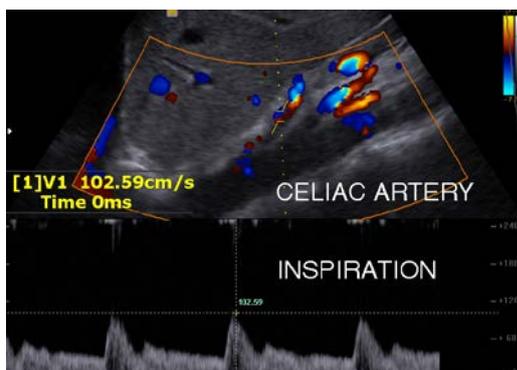


Figure 2B. Normal peak arterial velocity of celiac artery during inspiration by color duplex ultrasound

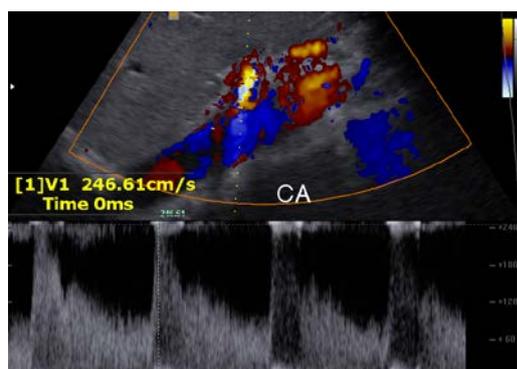


Figure 2C. Elevated peak velocity of celiac artery during expiration > 2m/sec

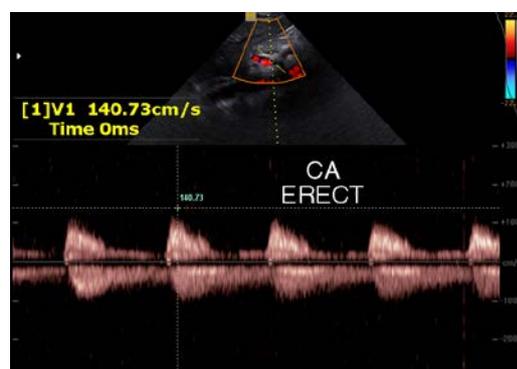


Figure 2D. normal arterial peak velocity of celiac artery in erect position due to further descent of celiac artery away from the median arcuate ligament

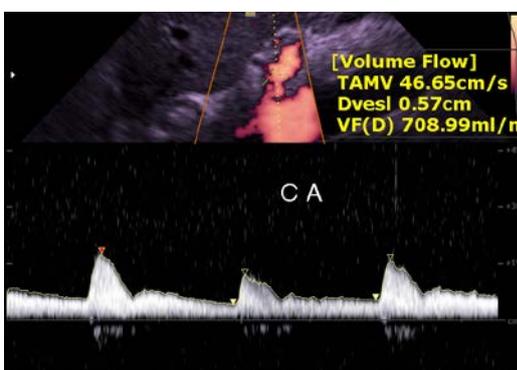


Figure 2E. Doppler estimated flow volume of celiac artery is within normal range in patient with imaging features of median arcuate ligament syndrome



Figure 2F. CT angiography revealed downward displacement and arterial kink of proximal celiac artery (white arrow)

Figure 3 A, B, C: Severe stenosis of celiac artery noted in asymptomatic patient with imaging features of median arcuate ligament syndrome.

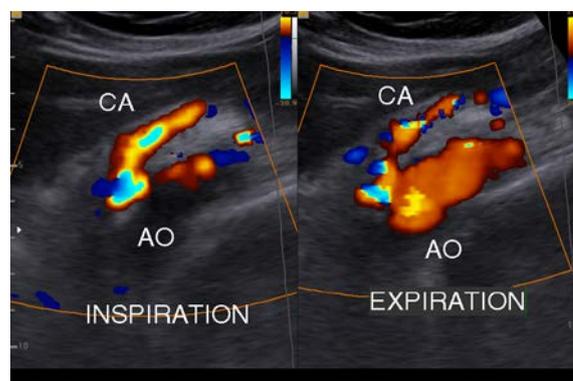


Figure 3A. Color Doppler ultrasound revealed severe stenosis of proximal celiac artery noted during expiration

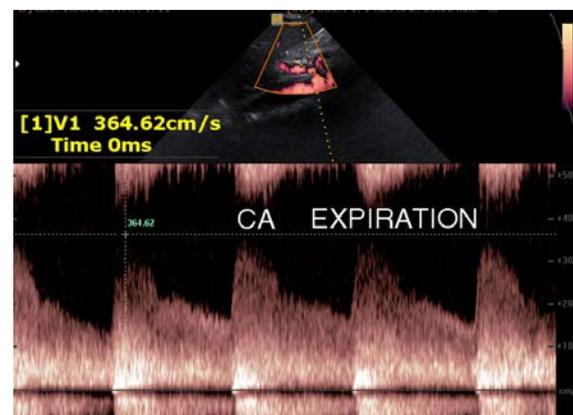


Figure 3B. Color duplex study revealed markedly elevated peak velocity of celiac artery during expiration

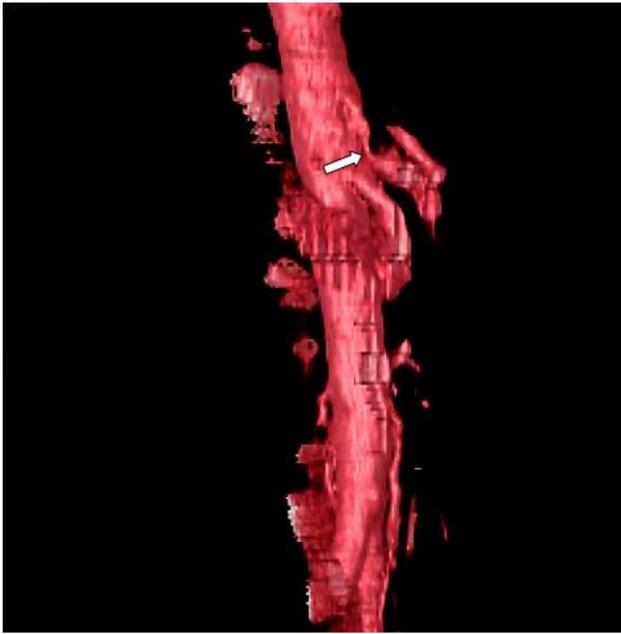


Figure 3C. 3 dimensional CT angiography revealed severe stenosis of proximal celiac artery (white arrow)

6. Discussion

In spite of 4% of our asymptomatic patients showed the typical color duplex ultrasound and CT angiographic features used to diagnose MAL syndrome, we can't deny its existence since many studies showed improvement of patient symptoms after surgical division of MAL either by open surgery or by laparoscopy [11,12,18,19,20]. The present study showed that compression of celiac trunk occurs in many asymptomatic patients to a variable degree and could be inspiratory, expiratory and even on the erect position if the median arcuate ligament was abnormally long and extends too low in position. Also showed that celiac artery stenosis occur transient mostly expiratory and don't alter significantly the blood flow volume per minute in the celiac artery as the flow volume per minute of my positive patients was $690\text{ml}/\text{min} \pm 30\text{ml}$ in comparison with $674 \pm 25\text{ml}/\text{min}$ in healthy individuals [21], which is explained by the increased diastolic flow and increased peak systolic velocity across the stenotic segment. In addition to the fact that the presence of good collateral flow between the superior mesenteric artery and celiac artery compensate for the severe stenosis or occlusion of celiac trunk and at least 2 mesenteric arteries should be occluded or severely stenotic to produce symptoms of mesenteric ischemia and The study performed by Carbornell et al [22] in 17 patients with celiac artery stenosis proved by angiography showed that most symptomatic patients were due to MAL syndrome and asymptomatic patients were due to atherosclerosis which means that not the arterial stenosis who produce symptoms as it occur in both but there should be an additional factor in MAL syndrome that cause the symptoms. Also the endovascular stent which depends only on relieving the stenosis with no dissection of sympathetic plexus is not associated with favorable outcome [23,24,25], Geelkerken et al [26] showed recurrence of patients symptoms after surgical division of MAL. All the fore mentioned reasons suggest that

compression of celiac trunk can play little or no rule in the pathogenesis of patients symptoms and raise the possibility that the recurrent compression that cause irritation and sometimes fibrosis with enlargement of celiac ganglia which is abundant around the celiac trunk [7,8,9] and carry the autonomic nerve supply to the upper gut can leads to gastroparesis, a finding which is seen to improve after surgical dissection of celiac ganglia with subsequent regularization of gastric myoelectric rhythm and appears to be responsible for the anorexia, vomiting and loss of weight [27]. The improvement in patients symptoms after surgical division of arcuate ligament can be due to the unavoidable dissection of celiac plexus and the subsequent peri-arterial sympathectomy [28]. Hargola et al showed that the concentric type of celiac trunk stenosis is due to celiac ganglia fibrosis and enlargement. [9] The presence of asymptomatic patients can be explained by the fact that celiac ganglia occur away from the celiac trunk in 32 % of patients [29] so it can't be compressed with the celiac trunk, or that the compression by the arcuate ligament is not satisfactory to cause pathology or irritation to the celiac ganglia.

7. Conclusion

The diagnosis of MAL syndrome shouldn't depend only on the imaging criteria and should be strongly linked to the unexplained patient symptoms after meticulous exclusion of other etiologies. The management shouldn't depend only on relieving the arterial stenosis but also on dissection of peri arterial nerve plexus.

Statement of Authorship

Ashraf Talaat Youssef : Conception and design of the study, collection, analysis and interpretation of data, and drafting the article with final approval of its completed form.

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