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Simultaneous Compression of the Celiac Trunk, Superior Mesenteric Artery, and Renal Arteries by the Median Arcuate Ligament: About One Case

Siddick Oumar, Kenza Fathallah, Hassan Ahmed, Aicha Merzem, Hasnaa Belgadir, Omar Amriss, Nadia Moussali, Naima El Benna

Department of Radiology, University Hospital Center Ibn Rochd, Casablanca, Morocco
Email: koyofil@yahoo.fr, Kenza.fathallah@gmail.com, Hassan.ahmed20aout@gmail.com, aichamerzem@gmail.com, hasnaabelgadir2023@gmail.com, omaramriss3@gmail.com, nmoussali@gmail.com, elbennan@gmail.com

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Abstract

Median arcuate ligament syndrome (MALS), is a rare abdominal vascular compression syndrome caused by the compression of the proximal celiac trunk by the median arcuate ligament. According to many authors, a low insertion of the diaphragmatic crura or an abnormally high origin of the celiac trunk from the aorta can cause compression of the celiac artery. Usually, patients with MALS are asymptomatic. Computed tomography (CT) angiography of the abdomen is the main imaging modality to confirm the diagnosis. The coexistence of celiac trunk and superior mesenteric artery compression by the median arcuate ligament is rarely described in the literature. To our knowledge, until now, a simultaneous combination of three abdominal vascular compressions by the median arcuate ligament has never been described. From this case, we report a simultaneous compression of the celiac trunk, superior mesenteric artery, and renal arteries by the median arcuate ligament.

Keywords

Median Arcuate Ligament Syndrome (MALS), Celiac Trunk Compression, Superior Mesenteric Artery and Renal Arteries Compression, Computed Tomography Angiography

1. Introduction

Median arcuate ligament syndrome (MALS), also known as Dunbar syndrome

and celiac compression syndrome, is a rare and controversial vascular compression syndrome caused by the compression of the celiac trunk by the median arcuate ligament [1]. The median arcuate ligament (MAL) is a fibrous arch connecting the right and left crura of the diaphragm and is typically found at the level of either the twelfth thoracic or the first lumbar vertebrae, usually passing superior to the celiac trunk artery [2].

Its pathophysiology remains controversial. For many authors, the syndrome is thought to be caused by vascular compression exerted by the median arcuate ligament in patients with an abnormally low insertion of the diaphragmatic crura or an abnormally high origin of the celiac trunk from the aorta [3].

MALS is typically a diagnosis of exclusion. Computed tomography (CT) angiography is the main imaging modality to confirm the diagnosis [4].

The coexistence of celiac trunk and superior mesenteric artery compression by the median arcuate ligament is rarely described in the literature [5]. To our knowledge, until now, simultaneous compression of the celiac trunk, superior mesenteric artery, and renal arteries by the median arcuate ligament has never been described.

In this case, we discuss the characteristic finding of an atypical narrowing of the proximal celiac trunk, superior mesenteric artery, and renal arteries with post-stenotic dilation by the median arcuate ligament on computed tomography angiography.

2. Clinical Observation

A 60-year-old patient with well-controlled hypertension was consulted for muscle-like pain in both lower limbs, more pronounced on the left, which worsened with walking and was relieved by rest. The patient did not report any other associated symptoms. Given this presentation of intermittent claudication, the patient was referred to the Radiology department for a Doppler ultrasound of the lower limbs. The ultrasound was inconclusive, showing demodulated waveforms. A CT angiography of the abdominal aorta and lower limbs was subsequently performed.

CT angiography showed a focal narrowing of the proximal celiac trunk forming a hooked or "J" appearance (on sagittal reconstruction) with post-stenotic dilatation (**Figure 1(a)** and **Figure 1(b)**). Additionally, there was a stenosis of the proximal superior mesenteric artery with post-stenotic dilatation (**Figure 1(a)** and **Figure 1(b)**), accompanied by significant collateral formation.

The imaging also revealed a focal narrowing of the proximal renal arteries with post-stenotic dilatation (**Figure 2(a)** and **Figure 2(b)**). CT angiography showed a dilatation of the inferior mesenteric artery without stenosis. There was significant collateral formation, especially the Riolon's arcade (**Figure 3**).

No atherosclerosis was noted, and there were no abnormalities in the lower limb arteries in this exploration.

A referral to the vascular surgery department was made. Given that the

Figure 1. (a) contrast-enhanced CT of the abdomen, sagittal image shows a focal stenosis of the proximal celiac trunk with a "J" appearance and a stenosis of the proximal SMA with a post-stenotic dilatation. Note the absence of atherosclerosis; (b) Volume Rendering CT showing a focal stenosis of the proximal of celiac trunk and SMA with post-stenotic dilatation.

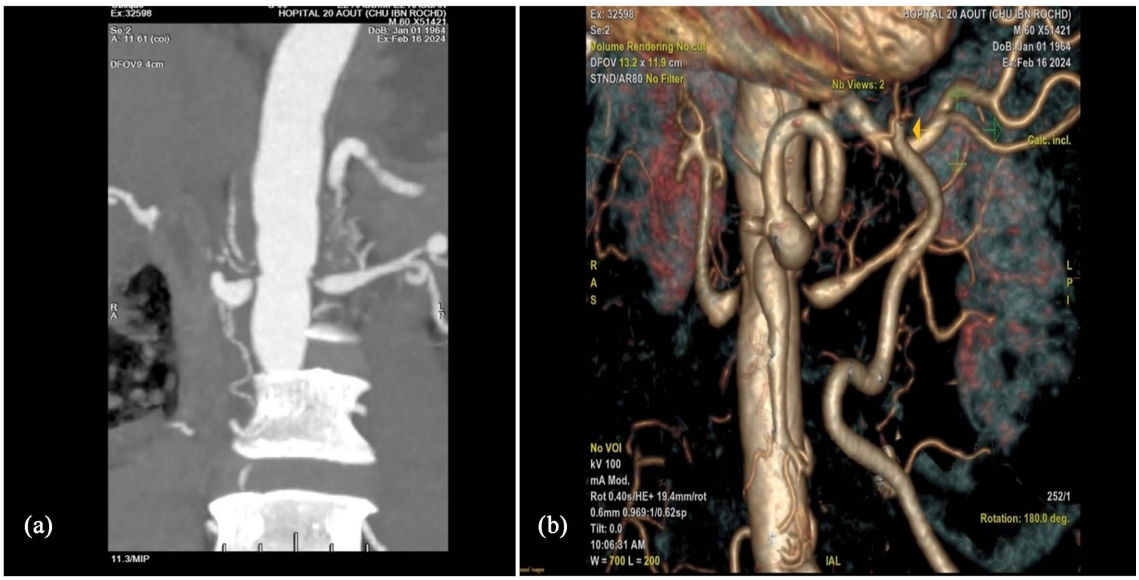

Figure 2. (a) contrast-enhanced CT of the abdomen, coronal image, shows a focal narrowing of the proximal renal arteries with a post-stenotic dilatation; (b) Volume Rendering CT showing a focal stenosis of the proximal renal arteries with a post-stenotic dilatation and importation collateral formation.

the patient did not present any abdominal symptoms and considering his advanced age, he was managed conservatively by the medical staff of the vascular surgery department. Clinical and radiological follow-up was recommended.

3. Discussion

Median arcuate ligament syndrome (MALS), also known as Dunbar syndrome and celiac artery compression syndrome, is one of the abdominal vascular compression

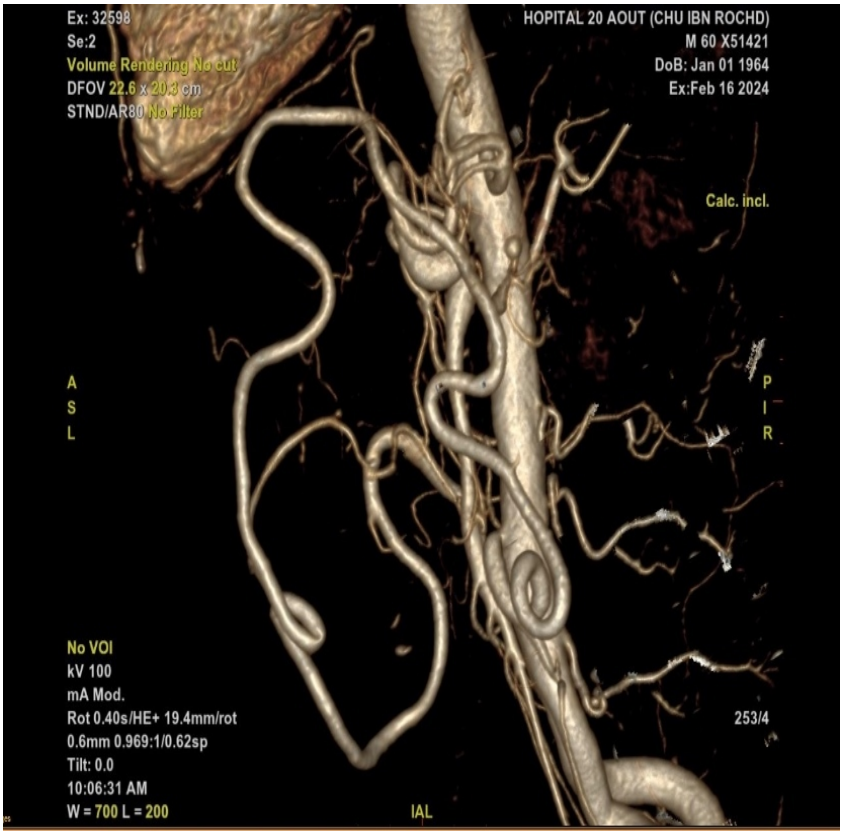

Figure 3. Volume Rendering CT of the abdomen, shows a dilatation of the inferior mesenteric artery with important collateral formation especially of the Riolon's arcade.

syndromes were first diagnosed by Harjola in 1963 [1]. MALS is an uncommon condition resulting from the compression of the celiac trunk by the median arcuate ligament [6]. The median arcuate ligament (MAL) is a fibrous arch connecting the right and left crura of the diaphragm, typically found at the level of either the twelfth thoracic or the first lumbar vertebrae, and usually passes superior to the celiac trunk artery [2].

For many authors, the syndrome is due to the compression of the proximal celiac artery by the median arcuate ligament in patients with an abnormally low insertion of the diaphragmatic crura or an abnormally high origin of the celiac trunk from the aorta [3]. While the reason for this anatomical malformation is unknown, it is believed that both hereditary and environmental factors likely play a role in the development of MALS. Due to this anatomical malformation, the celiac trunk and the celiac plexus are compressed by the MAL [7].

Most commonly, patients with MALS experience compression of the celiac artery alone. However, a few cases of patients with coexistence of celiac trunk and superior mesenteric artery compression by the median arcuate ligament have been described in the literature. Curl *et al.* reported one of the first cases of hemodynamically significant compression of both the SMA and the celiac artery by the median arcuate ligament [5]. Compression of the celiac artery and SMA by the median arcuate ligament has been infrequently reported, with the series

by Reilly *et al.* demonstrating that only 4 of the 51 patients with MALS had both celiac artery and SMA compressions [8]. Jeffrey J *et al.* reported a case of external compression of the superior mesenteric artery by the median arcuate ligament [9]. A rare case of simultaneous combination of three abdominal vascular compression syndromes: median arcuate ligament syndrome, superior mesenteric artery syndrome and Nutcracker syndrome has been reported by Renato Farina *et al.* [10].

Usually, patients with median arcuate ligament syndrome (MALS) are asymptomatic. However, a smaller subset will develop varied symptoms due to celiac compression, such as nausea, vomiting, postprandial pain, and weight loss [11]. These symptoms are typically more severe upon expiration due to the diaphragm moving caudally.

MALS is typically a diagnosis of exclusion combined with imaging findings of vessel compression by the MAL. Imaging plays a crucial role in the diagnosis of median arcuate ligament syndrome. There are multiple modalities to aid in the diagnosis, though some have proven to be more useful than others. Computed tomography (CT) angiography, ultrasound Doppler, and magnetic resonance angiography (MRI) are commonly used [4].

Modern multidetector computed tomography (CT) scanners offer numerous advantages, making them a preferred imaging modality for diagnosing Median Arcuate Ligament Syndrome (MALS). Evaluation typically involves CT angiography or MRI angiography during different phases of breathing (the arterial phase during expiration and the venous phase during inspiration) to assess stenosis degree. These scanners are readily available, non-invasive, and provide superior resolution with faster image acquisition. Additionally, they allow for multiplanar reconstruction, enhancing the visualization of complex anatomical structures. Classical findings include focal narrowing of the proximal celiac trunk forming a hooked or "J" appearance (best appreciated on sagittal images) [12]. The typical hooked configuration helps to differentiate from atherosclerosis. Secondary indirect signs, such as post-stenotic dilatation and collateral formation, can help confirm the diagnosis. Disadvantages of using CT angiography include contrast-induced nephropathy and radiation exposure.

Doppler ultrasound, performed by an experienced operator, can serve as another imaging modality for evaluating patients suspected of having median arcuate ligament syndrome [13]. Due to its dynamic nature, Doppler ultrasound is performed in different phases of respiration. Doppler ultrasound remains highly reliable in diagnosing MALS, with reported sensitivity and specificity of 75% and 89%, respectively [14]. Moreover, Doppler ultrasound offers the advantage of being radiation- and contrast-free, enhancing its safety profile for patients.

However, Doppler ultrasound may have limitations compared to CT angiography and magnetic resonance angiography (MRA), particularly in pediatric populations and in cases requiring detailed preoperative planning. Additionally, certain variables such as the presence of air in the gut can obstruct image cap-

ture, potentially reducing the effectiveness of Doppler ultrasound in some cases. Despite these limitations, Doppler ultrasound remains a valuable imaging modality for diagnosing MALS, especially when considering its safety and reliability.

Gastric exercise tonometry has been utilized as an adjunctive method to detect gastric ischemia caused by Median Arcuate Ligament (MAL) syndrome [15]. Elevated levels of intramucosal and intraluminal PaCO2 are indicative of gastric ischemia. The procedure involves taking measurements before, during, and after 10 minutes of submaximal exercise. Pathological results are identified by a gastric arterial PaCO2 difference greater than 0.8 kPa post-exercise, an arterial lactate level below 72 mg/dL (multiply by 0.111 to convert to millimoles per liter), and an increase in gastric PaCO2 levels following exercise. Limited studies have shown this method to be effective for both diagnosing and monitoring patients with MAL syndrome.

The main treatment for symptomatic patients with median arcuate ligament syndrome is surgery [16]. Surgical approaches may include both open and minimally invasive techniques, such as laparoscopic or robot-assisted procedures [17]. The goal of surgical intervention is to completely release the overlying tissue from the diaphragmatic crura, which forms the median arcuate ligament. Additionally, neurolysis of the celiac plexus nerve fibers may be performed to alleviate associated pain.

While both open and minimally invasive surgical techniques have demonstrated comparable outcomes in treating Median Arcuate Ligament Syndrome (MALS), the laparoscopic approach offers several advantages. These include reduced postoperative pain, shorter hospital stays, faster recovery times, and improved cosmetic outcomes.

Even though surgical decompression of the median arcuate ligament is extremely effective, there are still some complications that have been reported following the procedure, such as artery bleeding, postoperative thrombosed bypass graft with stroke, gastroesophageal reflux, pancreatitis, and hemothorax [18].

In cases where stenosis persists even after decompression of the median arcuate ligament, additional procedures may be necessary to restore adequate blood flow to the celiac artery. These procedures include revascularization by stenting, celiac artery bypass surgery, and celiac ablation [19]. These additional procedures are considered when there is persistent stenosis or when symptoms persist despite initial decompression of the MAL. The choice of procedure depends on factors such as the extent of stenosis, the patient's overall health, and the preferences of the treating physician.

4. Conclusions

Median arcuate ligament syndrome is a rare and controversial vascular compression syndrome caused by the compression of the celiac trunk by the median arcuate ligament. This compression can also rarely affect other abdominal vessels.

It is a rare anatomical malformation not to be ignored and constitutes a diag-

nosis of exclusion. Imaging plays a crucial role in the diagnosis of median arcuate ligament syndrome, with CT angiography being the main imaging modality to confirm the diagnosis of MALS. The main treatment for symptomatic patients with MALS is surgery. This is usually performed laparoscopically by dividing the median arcuate ligament and the crura of the diaphragm, followed by celiac ganglion neurolysis. Revascularization of the celiac artery by either endovascular stenting or bypass can be considered a secondary procedure.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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