Evaluation of the median arcuate ligament syndrome and the celiac axis with dynamic CT angiography in asymptomatic and symptomatic individuals

Poster No.: C-3048
Congress: ECR 2010
Type: Educational Exhibit
Topic: Vascular
Authors: I. Vivas, J. I. Bilbao, M. Arraiza, B. Zudaire, D. Cano, J. Alvarez-Cienfuegos; Pamplona/ES
Keywords: celiac trunk stenosis, median arcuate ligament, CT angiography
DOI: 10.1594/ecr2010/C-3048
Learning objectives

To review the significance of median arcuate ligament syndrome (MALS) associated with celiac axis compression

To illustrate the anatomy of the celiac trunk by CT angiography in asymptomatic patients, in patients with abdominal pain, and in patients with abdominal angina

To review the indications, techniques, results and complications of surgical therapy for MALS

Background

The median arcuate ligament syndrome (MALS) also known as celiac axis compression (CACS), was first described by Harjola in 1963. In 1965 Durban reported the first cases serie with surgery. They described a clinical syndrome of postprandial abdominal pain and malabssorption attributed to compression of the celiac trunk by the unyielding median arcuate ligament passing anterior to the artery and tightly compressing it against the aorta. The effect is accentuated by expiration, when the retroperitoneal contents (includend the aorta and its branches) move in a cephalas direction with respect to the diaphragmatic ligament which is relatively fixed in position at the aortic hiatus (Figures 1,2).

The syndrome is clinically characterized by the triade: postpandrial abdominal pain, presence of an epigastric bruit which increases in expiration and the assessment of extrinsic compression (>50%) of the celiac axis by the arcuate ligament by vascular imaging (angiography, CT-angiography, etc.).

Since the first description, the syndrome has been submitted to several controversies based on the variabilities in the results and also different physiopathological interpretations. Some authors have considered the MALS as the most frequent cause of chronic mesenteric ischemia, meanwhile its existence has been challenged by other authors.
In fact, compression of the celiac axis by the median arcuate ligament is an anatomic variant seen to variable degree in from 10 to 24% of the patients (in our series 11%) (Figure 3).

Anatomic bases

The arcuate ligament is formed by tendinous fibers between the two diaphragmatic cruras, which form the ventral arch of the aortic hiatus of which four variants have been reported. An abnormal fibrous thickening of MAL obstructing from 70% to 100% of the celiac trunk lumen has been previously reported in necropsy findings and surgical descriptions as well.

Another cause of the syndrome has been related with the celiac trunk embryological origin in the aorta. The origin of the celiac trunk undergoes variable caudal migration during embryogenesis, and may vary in location from the level of the 11 thoracic to the first lumbar vertebra. The mentioned origin can be located craneally within the thorax, causing a compression by the MAL's fibres. A craneally origin of the celiac trunk has been described in patients with astenic habitus. Some of the patients describe a pain relief with the knee-chest position, due to the relaxing of the vessels impingement by the MAL.

Charette, Balaban and other authors, related the symptoms with a thickening of the celiac plexus sustaining an extrinsic compression of the CT with flow impairment.

The syndrome is more frequent in young females with a thin body habitus, who many of them have previous history of weight loss as consequence of the intense postpandrial pain. As matter of fact, one of the characteristics of good response to surgical release was the mentioned above according to a seminal paper of Reilly, published in 1985, who reported the long-term follow-up (median 9 years, range 1-18) of 51 patients surgically treated.

A simultaneous stenosis of the celiac trunk with superior mesenteric artery and renal arteries compression originating chronic intestinal ischemia and renovascular hypertension have described.

Phisiopathology

Several hypothesis have been proposed regarding the origin of the syndrome, although is a topic of academic debate. The most common theory relates the symptoms with the visceral ischemia originated by the extrinsic compression of the CT. In support of
this there are the radiologic findings (CT-angiography, MR-angiography), the surgical descriptions and the symptoms release (close to 80%) after surgical division of the MAL and release of the CT. However, it was not entirely clear whether the surgical benefit is due to release of compression of the artery or disruption of the neural pathways.

By contrast, another fact questions this mechanism. The most mentioned is the experimental and clinical evidence that the complete occlusion of two of three main major abdominal arteries are required to provoke abdominal angina, as have been shown in animals or in oncologic and thoracoabdominal aneurism cases.

In 1969, Bron and cols., reported 12.5% of asymptomatic patients with "radiologic findings" of CT compression. In 1972 Colapinto found signs of compression and stenosis on the 31% of asymptomatic patients. These data and the symptoms recurrence following the surgical division of the MAL would support the caution in attributing the patients' symptoms to the arterial occlusion and the existence of the syndrome.

The second pathogenic theory is known as "steal phenomenon" by which blood from superior mesenteric artery territory is diverted through collateral vessels concurrent with a high demand of oxygen by the gut. Kalapatapu has recently reproduced the symptoms inducing a selective splachnic vasodilatation in the superior mesenteric artery. In the experience of this author, the induction of symptoms is a clear sign for selecting surgical candidates, obtaining a 75% of success with MAL release.

In favour of this theory would be the reported consequences in young athletes after intense exercise in liver transplantation and following pancreatoduodenectomies in patients who had an unnoticed celiac trunk compression.

**Clinical features and diagnosis**

The intense postprandial epigastric pain (mean duration 20-30 minutes), nausea and vomiting are the most common symptoms. The postprandial pain induces a "food fear" and avoid eating, being cause of weight loss (5-10 kg). The pain may be relieved in the knee-chest position, and is more frequent in young females (3/1) about 40-50 years; although it has been reported in children and adults. The mean duration of symptoms range between three months and more than 10 years.

Because of the difficulty in diagnosis of the syndrome, many patients have been previously diagnosed of functional disorders and had undergone an extensive gastrointestinal evaluation including endoscopy, motility studies, abdominal CT or surgical
intervention as exploratory laparoscopy/laparotomy, without establishing a diagnosis. In some series, 10-15% of the patients had psychiatric antecedents.

On physical examination, patients usually have a thin and leptosomatic habitus and an epigastric bruit - which increases in deep expiration - is present in 83%; although abdominal bruits occur in approximately 30% of healthy adults.

**Treatment**

Several endovascular procedures as angioplasty with or without stenting have been carried out with poor results. The sustained ant tight compression of the diaphragmatic fibers led to permanent changes on the vessel wall, fragmenting the stents and making this procedure unsuccessful (Figure 4).

The other surgical techniques are the simple surgical division of the MAL and celiac trunk release with blood flow restoration assessing by Doppler-US and other complex procedures including vascular reconstruction with patch angioplasty of the celiac artery, aortoceliac bypass with saphenous vein or Dacron graft and reimplantation of the celiac trunk in the aorta.

The most common procedure is the surgical division of the constrictive fibers of the celiac plexus and release of the celiac trunk up its origin the aorta (table 1). Since the introduction of the laparoscopic approach in 2000, excellent results - with the benefits of this procedure, less hospital stay, faster recovering, less postoperative pain, etc. have been reported. The technique is depicted in (Figure 5). The cases reported in these series were well selectionated: young females with intense pain and weight loss. In our experience of 7 cases underwent surgery by laparoscopic technique, similar results were obtained (table 2).

As mentioned before, the results have been variable. Perhaps the main reason of that is the short-term follow-up and the diversity of abdominal pain medical profiles diagnosed as MALS.

The seminal paper published by Reilly in 1985 describing the long-term follow-up of 51 patients treated surgically was crucial for explaining these divergences. In this report, 80% of young patients (40-60 years) with postpandrial pain, weight loss and poststenotic dilatation in the celiac angiogram, releaved the symptoms. Similar results were published by other authors and the same prognostic factors were found in regression studies: epigastric pain related with ingesta as more relevant parameter.
The accumulated experience points out the importance of a detailed anamnesis and the selective imaging techniques as Doppler-US, CT-angiography; avoiding unnecessary explorations.

The laparoscopic approach and minimally invasive techniques: MAL division, with ulterior endovascular techniques when necessary -angioplasty, Stent-, open a new "era" in the diagnosis and treatment of the syndrome described in 1963.

Tables

Table 1

Results obtained by the simple division of the MAL in the most numerous series

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th># cases</th>
<th>Release MAL (*)</th>
<th>Improvement %</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dunbar</td>
<td>1965</td>
<td>13</td>
<td>13</td>
<td>69%</td>
<td>9 -15 months</td>
</tr>
<tr>
<td>Szilagy (***)</td>
<td>1972</td>
<td>165</td>
<td>129</td>
<td>82%</td>
<td>1 months - 4 years</td>
</tr>
<tr>
<td>Evans</td>
<td>1974</td>
<td>47</td>
<td>47</td>
<td>83%</td>
<td>6 months - 11 years</td>
</tr>
<tr>
<td>Watson</td>
<td>1977</td>
<td>20</td>
<td>20</td>
<td>80%</td>
<td>6 months - 4 years</td>
</tr>
<tr>
<td>Reilly</td>
<td>1985</td>
<td>51</td>
<td>33</td>
<td>65%</td>
<td>1 year - 18 years</td>
</tr>
</tbody>
</table>

(*) MAL. Median arcuate ligament.


Table 2

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th># cases</th>
<th>Surgery</th>
<th>Hospital stay</th>
<th>Follow-up</th>
<th>Improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Roayaie</td>
<td>2002</td>
<td>1</td>
<td>135 min.</td>
<td>55 cc.</td>
<td>15 hours</td>
<td>3 months</td>
</tr>
<tr>
<td>Name</td>
<td>Year</td>
<td>Cases</td>
<td>Duration</td>
<td>Fluids</td>
<td>Recovery</td>
<td>Complications</td>
</tr>
<tr>
<td>-------------</td>
<td>------</td>
<td>-------</td>
<td>----------</td>
<td>--------</td>
<td>----------</td>
<td>---------------</td>
</tr>
<tr>
<td>Dordoni</td>
<td>2002</td>
<td>1</td>
<td>125 min.</td>
<td>65 cc.</td>
<td>48 hours</td>
<td>6 months</td>
</tr>
<tr>
<td>Carbonell</td>
<td>2005</td>
<td>1</td>
<td></td>
<td></td>
<td>72 hours</td>
<td>7 months</td>
</tr>
<tr>
<td>Baldassarre</td>
<td>2007</td>
<td>1</td>
<td>130 min.</td>
<td>-</td>
<td>No consta</td>
<td>3 months</td>
</tr>
<tr>
<td>Jaik</td>
<td>2008</td>
<td>1</td>
<td>160 min.</td>
<td>50 cc.</td>
<td>48 hours</td>
<td>6 months</td>
</tr>
<tr>
<td>Jarry</td>
<td>2008</td>
<td>1</td>
<td>168 min.</td>
<td>-</td>
<td>24 hours</td>
<td>1,5 months</td>
</tr>
<tr>
<td>Duffy</td>
<td>2008</td>
<td>1</td>
<td>218 min.</td>
<td>-</td>
<td>24 hours</td>
<td>10 months</td>
</tr>
<tr>
<td>Vaziri</td>
<td>2009</td>
<td>3</td>
<td>151 min.</td>
<td>&lt; 50 cc.</td>
<td>24 hours</td>
<td>6 months</td>
</tr>
<tr>
<td>Rotellar</td>
<td>2009</td>
<td>7</td>
<td>111 min.</td>
<td>&lt; 50 cc.</td>
<td>65 hours</td>
<td>6 months - 8 years</td>
</tr>
</tbody>
</table>

(*) Authors' results in 2009.

(**) Surgery using Da Vinci Surgical System (Intuitive Surgical, Sunnyvale, California).

Experience with laparoscopic approach in the treatment of MALS

Images for this section:
FIGURE 1: Sagittal 3D volume-rendered images of a patient with epigastric pain demonstrate acute angulation and narrowing (arrow) of the proximal celiac axis. The hooked appearance of the celiac trunk is characteristic of MALS.

Celiac artery compression decreases with inspiration as the abdominal viscera descend, causing the caudal orientation of the celiac artery. It increases with expiration, and in the worst cases the celiac artery occludes.

Fig. 1
FIGURE 2:
The compression of the celiac trunk is thought to be caused by the converging connective tissues at the margins of the aortic hiatus arising from the arching fibers of the right and left diaphragmatic crura, known as the median arcuate ligament. A soft-tissue band extending across the anterior aspect of the celiac trunk may be observed on axial and sagittal projections (arrow). Poststenotic dilatation of the celiac trunk anterior to the soft-tissue band is also seen.

Fig. 2
FIGURE 3: A high prevalence of significant stenosis of the celiac artery has also been reported in asymptomatic patients (11% in our series). Sagittal reconstructions of contrast-enhanced CT show mild to severe celiac artery narrowing in a 44-year-old woman with pelvic pain. The celiac trunk stenosis was a fortuitous finding not related with her clinical case.
FIGURE 4: 34-year old man with MALS treated with stent. The lateral aortogram before treatment showing a U-shaped configuration of the celiac trunk with compression along the superior aspect (A). Arteriography after the treatment with the placement of two contiguous stents (B). Two years after, in a knee X-ray an stent was discovered in the popliteal artery. 3D volume rendered ((D, E, ) and sagittal MIP (F) images show one stent in the celiac trunk (C) and another stent impacted in the left popliteal artery, that showed good flow across

Fig. 4
Fig. 5
We reviewed by CT angiography (MDCT 64-row) anatomical variations of the celiac axis at inspiration and expiration in 100 patients with abdominal pain, some of them with mesenteric angina (45 men and 55 women, ranging in age from 28 to 72 years (average age, 47.85 years), and we have compared the results with 100 patients who were submitted to an abdominal CT, for unrelated reasons (metastatic disease (54 cases), weight loss (22), neoplasm depiction (14), and miscellaneous indications (10). There were 52 men and 48 women, ranging in age from 22 to 90 years (average age, 60.41 years). The median arcuate syndrome was not clinically suspected in any of these 100 patients. All CT examinations (study and control groups) were performed after the intravenous administration of 120-150 cc of contrast material (Omnipaque 300 mg I/ml-General Electric) by using a MDCT 64-row (Definition or Sensation 64 Siemens,Germany).

We also retrospectively review the abdominal CT findings in fifteen cases of severe celiac axis stenosis encountered in a 5-year period to determine whether CT findings about the celiac axis in these patients were specific for the diagnosis of significant median arcuate ligament compression. There were 6 men and 9 women, ranging in age from 26 to 86 years (average age, 45.46 years). The diagnostic was established by characteristic appearance on CT angiography and was confirmed surgically in seven cases.

Results

Review of CT findings about the celiac axis in the control patients showed normal appearances of the vessels in the majority (89%) of subjects. The celiac trunk was well opacified, and its origin from the abdominal aorta was clearly seen. In 11 patients a celiac trunk narrowing with minimal poststenotic dilatation was found, but in these cases any patient presented abdominal pain related to the celiac trunk stenosis.

In this group the mean diameter of the celiac trunk was 0.68 cm (0.78 poststenotic diameter in eleven cases with celiac trunk narrowing). The mean diameter of the superior mesenteric artery (SMA) was 0.70; the mean distance between the SMA and the aorta was 2.17 cm and the estimated angle between the SMA and the aorta was 40.32 degrees.

In the abdominal pain group, the mean diameter of the celiac trunk was 0.55 cm (0.79 poststenotic diameter in thirty-six cases with celiac trunk narrowing). The mean diameter of the superior mesenteric artery (SMA) was 0.71; the mean distance between the SMA
and the aorta was 1.30 cm and the estimated angle between the SMA and the aorta was 29 degrees.

In the group with the diagnosis of MALS confirmed (15 patients, average age, 45.46 years), the mean diameter of the celiac trunk was 0.47 cm (0.75 poststenostic diameter). The mean diameter of the superior mesenteric artery (SMA) was 0.65; the mean distance between the SMA and the aorta was 1.15 cm and the estimated angle between the SMA and the aorta was 23.58 degrees.

The statistic study revealed significant differences between the control group and the group with abdominal pain (p< 0.001) in the celiac trunk diameter, and the distance (p<0.001) and the angle (p<0,01) between the SMA and the aorta. There were no differences with regard to the diameter of the SMA (Figure 6).

**Imaging findings**

The radiologic confirmation of celiac trunk compression and severe stenosis or occlusion is mandatory for the diagnosis. In the past, the gold standard of MALS was aortic arteriography with lateral aortogram until the introduction of CT-angiography, with 3D software and magnetic resonance (MR)-angiography, that have greatly improved the ability to obtain high quality images of the vessels.

In the lateral projections of aortography and selective mesenteric studies, an image of "hook" -characteristic of the MAL effect upon celiac axis- is assessed (Figure 7). Postestenotic dilatation, collateral circulation and pancreaticoduodenal artery aneurisms have been reported. The patency of superior mesenteric artery and renal arteries must be evaluated. Duplex ultrasound have shown to be useful in assesing changes in CT orientation and increase in the peak systolic velocity (>1.8 m/sec) in inspiration vs expiration.

For the proper evaluation of the MALS with CT angiography, 3D imaging is necessary. The findings characteristic of the syndrome, may not be detected on axial plane alone. Currently the thin-section multidetector CT scanners along with 3D software in sagital plane is optimal for assesing the proximal portion of the celiac trunk (Figures 8, 9), ruling out other causes of celiac artery occlusion or narrowing as atherosclerotic disease.

Some authors have developed more specific studies in order to asses the celiac trunk occlusion and stablish a more precise surgical indication. Mensik y cols. studied
320 patients with symptoms of chronic intestinal ischemia, with selective angiography and gastric tonometry, yielded the CT syndrome in 43 patients (13.4%), indicating the surgical treatment exclusively in those who presented signs of gastric ischemia (gradient intraluminal gastric and arterial PCO$_2$), obtaining a 83% of symptoms relieve in these patients.

Images for this section:

Fig. 1
FIGURE 7: 29-year-old woman with abdominal pain following large meals. Correlation between the lateral aortogram (A), the MIP sagittal reconstructions (B) and the 3D volume rendered reformations (C) showing the configuration of the celiac trunk with compression along the superior aspect, that increases in expiration. This is characteristic of impingement by the median arcuate ligament and/or celiac plexus.

Fig. 2
FIGURE 8: AngioCT in a 32 year old female with severe epigastric pain, specially after meals, with postprandial nausea and vomiting. The axial CT in inspiration (A) and expiration (B) shows soft-tissue band between the aorta and the celiac trunk, that comprisses the celiac axis. (C) The sagital projection in expiration shows characteristic kinking of the proximal celiac axis, creating a hooked appearance. The liver presented a mosaic pattern that in laparoscopy the surgeon made a biopsy (violaceous colour of the liver) thats revealed hepatic stasis and hepatocites necrosis (vascular repercussion by celiac trunk stenosis?)

Fig. 3
Fig. 4

FIGURE 9: The same patient than in figure 5. Sagittal 3D volume-rendered images in colour and MIP sagital in inspiration (A) and expiration (B) shows the severe compression of the celiac axis, that in expiration is practically an occlusion (more than 90% narrowing of the proximal celiac axis). The patient was surgically treated with marked improvement and disappearance of the symptoms.
Conclusion

The diagnosis of MALS can be readily delineated by CT angiography at inspiration and expiration. However, the presence of narrowing of the celiac axis alone is not enough to intervene. Post-stenotic dilatation, collateral vessels, gastroduodenal artery dilatation are, indeed, secondary effects that indicate hemodynamic changes that help in the diagnosis and therapeutic planning in MALS. Anatomical variants may imply an important source of diagnostic pitfalls in patients with abdominal angina. Surgery should be performed only when the symptoms are serious and only after careful investigation has ruled out any other possible etiology.

Personal Information

Isabel Vivas Pérez
Department of Radiology
Clínica Universidad de Navarra
Avda Pío XII, 36
31008
Pamplona (Navarra)
SPAIN

isvivasp@unav.es

References

Am. J. Roentgenol. 1965; 95 (3): 731-744

- Brandt, L.J.; Boley, S.J. Celiac axis compression syndrome.
- Linder H.; Kemprud, E. A clinicoanatomical study of the arcuate ligament of the diaphragm.
- Rob, C. Stenosis and thrombosis of the celiac and mesenteric arteries.
• Feindt, P.; Walter, P.; Omlor, G. Das kompressionssyndrom des truncus coeliacus.
• Reilly, L.M.; Ammar, A.D.; Stoney, R.J.; Ehrenfeld, W.K. Late results following operative repair for celiac artery compression syndrome.
• Lawson, J.D.; Ochsner, J.L. Median arcuate ligament syndrome with severe two-vessel involvement.
• Lord, R.S.; Tracy, G.D. Coeliac artery compression.
• Plate, G.; Eklof, B.; Vang, J. The celiac compression syndrome: myth or reality?
• Bron, K.M.; Redman, H.C. Splanchnic artery stenosis and occlusion incidence, arteriographic and clinical manifestations.
  Radiology 1969; 92: 323-328. ...: Ein seltenes Krankheitsbild. ........ ...

  Radiology
3. Desmond, C.P.; Roberts, S.K.; Exercise-related abdominal pain as a manifestation of the median arcuate ligament syndrome.
6. Fortner J.; Watson, R. Median arcuate ligament obstruction of celiac axis and pancreatic cancer.


20. Takach, T.J.; Livesay, J.J.; Reul, G.J.; Cooley, D.A. Celiac compression syndrome: Tailored therapy based on intraoperative findings.


facilitated by laparoscopic ultrasound scanning to confirm restoration of flow.


