

CASE REPORT

Spontaneous bleeding of the inferior pancreatic-duodenal artery in median arcuate ligament syndrome: do not miss the diagnosis

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Abstract

Median arcuate ligament syndrome (MALS) is a rare condition in which the median arcuate ligament (MAL) causes compression of the coeliac trunk. The chronic compression leads to coeliac trunk luminal narrowing and reduced blood supply to the abdominal splanchnic organs with possible local complications such as pseudo-aneurysms and spontaneous bleeding. Its incidence is probably underestimated due to the poor availability of color Doppler ultrasonography (CD-US), especially in an Emergency Department (ED) setting.

A 44-year old woman presented to Pisa University Hospital ED with acute abdominal pain. An abdominal ultrasonography scan (US scan) was performed showing the presence of free liquid in the Douglas pouch. The abdominal computed tomography scan (CT scan) highlighted the presence of a large mesenteric hematoma. A CD-US revealed a significant stenosis of the coeliac artery. A selective angiography confirmed the diagnosis of MALS with a pseudo-aneurysm of the inferior pancreatic-duodenal artery, which was successfully embolized.

INTRODUCTION

The coeliac artery arises in close proximity with the diaphragm and hence has an important anatomical and pathophysiological relationship with it. Median arcuate ligament (MAL) is a fibrous structure at the base of the diaphragm that bridges anteriorly the right and left diaphragmatic crura. Some MAL anatomic variants can occur in MAL syndrome (MALS) resulting in the compression of the coeliac axis between the MAL and the diaphragmatic crura [1].

The classical clinical presentation of MALS is postprandial epigastric pain and unexplained weight loss accompanied by an epigastric bruit. However, some patients may be completely

asymptomatic, and diagnosis can be reached only after a complication or even by chance [2]. These different clinical presentations can make the diagnosis challenging resulting in the real figures being underestimated.

While angiography is the gold standard for the diagnosis of MALS, color Doppler ultrasonography (CD-US) is a non-invasive method to diagnose MALS that is more widely available. Moneta [3] first described the cut-offs of peak systolic velocities (PSVs) in CD-US, in patients with angiographic evidence of MALS. PSVs of ≥ 200 cm/sec in the coeliac artery could predict a $\geq 70\%$ stenosis with a sensitivity of 87% and a specificity of 80%. Recently, Gruber and colleagues [4] compared patients with angiographic evidence of MALS and healthy volunteers in order to reassess

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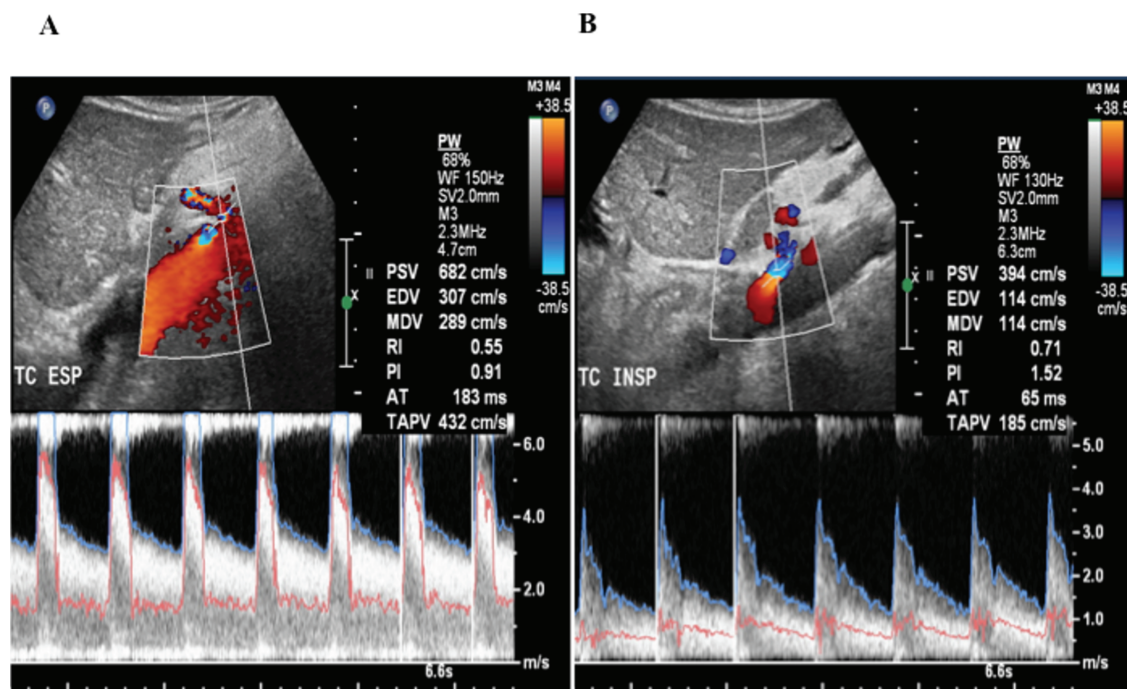


Figure 1: CD-US indicating the PSVs in the coeliac artery (A) during maximum expiration and (B) during maximum inspiration.

the velocimetric ultrasound cut-offs in MALS. They reported that a coeliac artery end diastolic velocity ≥ 350 cm/s, and a coeliac artery deflection angle of 50° , predicts MALS with high sensitivity (83%) and specificity (100%) [4].

CASE REPORT

A 44-year old woman presented to Pisa University Hospital Emergency Department (ED) with atraumatic acute abdominal pain. Her medical and surgical history was unremarkable, and she was not on any regular medication.

On her arrival, the patient was distressed, tachycardic (120 beats/min) and tachypnoic (RR=24) with normal blood pressure (130/80 mmHg), normal oxygen saturation (98%) and afebrile. The abdomen was distended with widespread tenderness, guarding and positive Blumberg sign (also referred to as rebound tenderness, which is indicative of peritonitis). She was treated with intravenous Paracetamol, and an abdominal ultrasonography was performed, highlighting the presence of free liquid in the Douglas pouch. An abdominal computed tomography (CT) scan with contrast was completed, showing the presence of a large hematoma in the mesentery (maximum diameter 12 cm), without any sign of ongoing bleeding. Her blood tests were normal apart from modestly raised C-reactive protein (5.6 mg/dl) and mildly decreased Hemoglobin (11.2 g/dl). Urgent treatment either with embolization or laparotomy was excluded after a multidisciplinary evaluation (Internist, Interventional Radiologist and General Surgeon). The patient was admitted to a high dependency unit for monitoring. She was treated with intravenous painkillers and intravenous proton pump inhibitors. Total parenteral nutrition was also given in order to achieve bowel rest until the pain resolved. Total blood count was daily monitored with stable hemoglobin levels. Four days later a CT scan with contrast was repeated, showing a small reduction of the hematoma.

Finding the diagnosis of spontaneous abdominal bleeding unconvincing, a CD-US was performed revealing a significant stenosis of the coeliac artery during expiration (Fig. 1). PSV in the coeliac trunk was measured as 682 cm/s and dropped to 394 cm/s with deep inspiration (Δ PSV = 288 cm/s).

A selective angiography of the superior mesenteric artery was performed and confirmed the diagnosis of MALS with a pseudo-aneurysm of the inferior pancreatic-duodenal artery, which was successfully treated with embolization.

The patient was discharged with a surgical follow-up. Three months later a laparoscopic surgery with partial dissection of the MAL fibers was performed. At sixth month of follow-up the patient is completely asymptomatic. The follow-up CD-US showed a significant decrease of the PSV (297 cm/s in expiration), which however did not return to normal values.

DISCUSSION

In MALS blood flow to the splanchnic organs is diverted to the superior mesenteric artery and then to the pancreatic-duodenal arcades due to decreased flow in the coeliac trunk. The pancreatic-duodenal arteries cannot sustain this high flow leading to the formation of pseudo-aneurysms, which can eventually result in a spontaneous rupture.

Even though the pathophysiology of MALS seems to be established, there are challenges regarding its optimal diagnostic and therapeutic tools. Our patient has had two CT scans and neither indicated an *ab extrinsic* compression of the coeliac axis from the diaphragm. This is due to the fact that in a CT scan, if not clinically suspected, images are not acquired during both inspiration and expiration. As far as the CD-US is concerned, there is no consensus among radiologists on the velocimetric cut-offs to be applied in the diagnosis of MALS. However, in the case of our patient the very high difference in PSV between inspiration and expiration allowed us to clearly demonstrate a significant *ab*

extrinsic stenosis of the coeliac artery, which was later confirmed with selective angiography. Taken together, this case suggests that subjects who admit to the ED for atraumatic abdominal pain and their initial diagnostic work-up puts in light a hematoma with no sign of ongoing bleeding, it is of paramount importance that they should also undergo a complete abdominal CD-US as soon as possible. If such a diagnostic work-up had been undertaken in our patient, diagnosis would have been reached earlier.

Also concerning the treatment of MALS, currently there is no consensus on the gold standard approach, with some authors proposing embolization of the aneurysm alone, while others suggest relieving the celiac stenosis [5]. If the relieving approach is chosen, there is higher chance of there being no further recurrence of aneurysm. On the contrary, in case of asymptomatic subjects with incidental diagnosis of MALS, until now there seems to be no indication for treatment [5].

In conclusion, MALS is probably an underestimated clinical entity. With this case report we wish to increase the index of suspicion of practitioners, since if left undiagnosed it can lead to severe, life-threatening complications. Finally, we suggest that abdominal CD-US in inspiration and expiration should be performed in all patients presenting with atraumatic abdominal hematoma.

CONFLICTS OF INTEREST STATEMENT

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ETHICAL APPROVAL

Not applicable.

CONSENT

Obtained.

GUARANTOR

Eleni Rebelos had full access on the data and is the guarantor of this work.

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