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Radiological Case Report / Caso Clínico

Acute Median Arcuate Ligament Syndrome Onset: Unexpected Complication after Laparoscopic Nissen Fundoplication

Síndrome do Ligamento Arcuato Mediano Agudo: Complicação Inesperada após Fundo-Plicatura de Nissen Laparoscópica

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Abstract

We present the case of a female patient who acutely developed median arcuate ligament syndrome with severe hepatic cytolysis, shortly after laparoscopic Nissen fundoplication for reflux esophagitis. CT angiography proposed the diagnosis of median arcuate ligament syndrome, causing splenic and gastric ischaemia and perfusion abnormalities in the liver parenchyma. Immediate surgery confirmed the diagnosis and division of the ligament was successfully accomplished, with subsequent organic reperfusion and no major complication on follow-up. To the best of our knowledge, the potential association between median arcuate ligament syndrome and Nissen fundoplication has not previously been documented in the literature.

Keywords

Iatrogenic median arcuate ligament syndrome; Nissen fundoplication; Acute abdominal ischemia.

Resumo

Os autores apresentam o caso de uma doente do sexo feminino que desenvolveu de forma aguda o síndrome do ligamento arcuato mediano, com um quadro de acentuada citólise hepática, no pós-operatório de fundoplicatura de Nissen laparoscópica por queixas de refluxo esofágico. O diagnóstico de síndrome do ligamento arcuato mediano foi proposto com base nos achados da angio-TC, que demonstrava sinais de isquémia esplénica e gástrica e alterações da perfusão do parênquima hepático A doente foi de imediato submetida a cirurgia, a qual confirmou o diagnóstico; a divisão do ligamento foi conseguida com sucesso e, subsequentemente, verificou-se reperfusão orgânica sem complicações major no seguimento. De acordo com o nosso conhecimento, a potencial associação entre síndrome do ligamento arcuato mediano e fundoplicatura de Nissen não foi previamente documentada na literatura.

Palavras-chave

Síndrome do ligamento arcuato mediano iatrogénico; Fundoplicatura de Nissen; Isquémica abdominal aguda.

Introduction

The median arcuate ligament (MAL) is a fibrous arch that connects the diaphragmatic crura, forming the anterior margin of the aortic hiatus.[1,2] The ligament usually passes superior to the origin of the celiac axis, but in some individuals it may cross over the proximal segment of the artery, causing a characteristic indentation. In a small group of these individuals, the MAL can compress the celiac axis enough to be hemodynamically significant, causing symptoms.[3] The median arcuate ligament syndrome (MALS) was first described in 1963 by Harjola and, two years later, Dunbar reported the first surgically treated patients.[4,5] The syndrome is also known as Dunbar syndrome and celiac artery axis (or trunk) compression syndrome.

Clinical Case

We report the case of a 53-year-old woman, who presented abdominal pain, nausea and vomiting. She had performed a laparoscopic Nissen fundoplication surgery for hiatal hernia and reflux esophagitis five days before and was discharged home earlier that day. No other relevant previous history. At physical examination, she was prostrated but reactive to painful stimulus, had mild tachycardia with normal blood pressure, was apyretic, had pale and dehydrated skin and mucosa, and the abdomen was mildly distended with tenderness at the peri-umbilical region, without peritoneal reaction. Blood test showed hemoglobin of 8,6 g/dL (basal value of 9,4 g/dL), leukopenia of $3{,}30 \times 10^9$/L ($4\text{-}11 \times 10^9$/L), C-reactive protein of 1,5 mg/dL (< 0.5mg/dL), ALT of 5078 U/L, AST of 4083 U/L, total bilirubin of 4,3 mg/dL (< 1.2mg/dL), lactate dehydrogenase 6385 U/L (140- 280-U/L), gamma glutamyl transferase of 88 (<30U/L). A computed tomography (CT) angiography (Fig. 1) was performed and showed a significant stenosis at the emergency of the celiac axis with post-stenotic dilatation and a hook-like morphology in sagittal plane, as well as signs of liver perfusion abnormalities and gastric and splenic ischaemia. The diagnosis of MALS causing severe abdominal ischaemia was proposed.

The patient was immediately taken to surgery. Exploratory laparotomy was performed, confirming extrinsic compression of the celiac trunk by the median arcuate ligament. Surgical division of the ligament was accomplished,

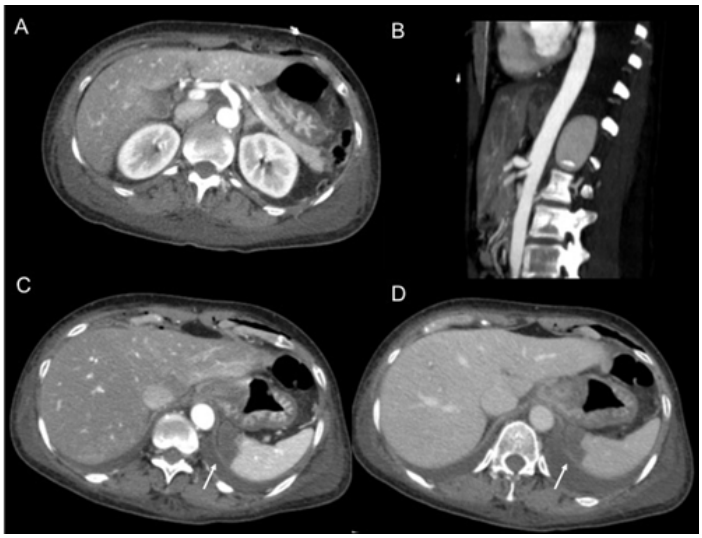

Figure 1 – Contrast-enhanced CT angiography, at emergency department. Axial view (A) and sagittal reconstruction (B) of the upper abdomen showing the characteristic hook appearance on the proximal celiac axis when it is compressed by the MAL (in sagittal view). Axial views at an upper level, in the arterial phase (C) and in the portal phase (D) showing heterogeneous opacification of the liver parenchyma in the arterial phase that becomes homogeneous in the portal phase, suggesting perfusion abnormalities in the liver parenchyma. Also, hyperenhancing gastric mucosa and an hypoattenuating area at the periphery of the spleen (arrow), suggesting ischaemic alterations.

with subsequent dramatic resumption of celiac axis and common hepatic artery pulsation and organic perfusion. The Nissen fundoplication was unremarkable and there was no need for any surgical correction. Intraoperative trial clamping of the celiac axis was performed afterwards and demonstrated breakdown of the circulation as in MALS (Fig. 2).

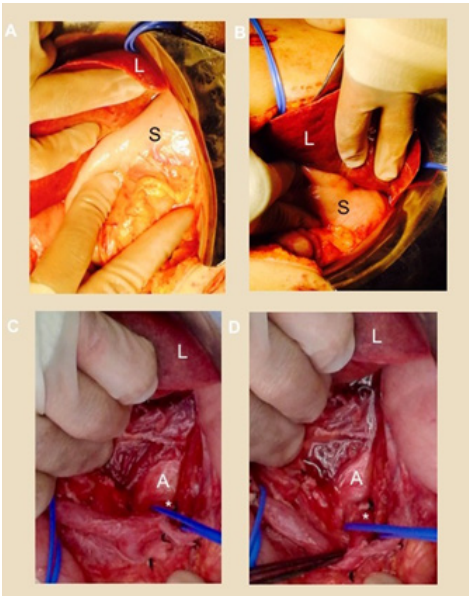

Figure 2 – Intraoperative images. Before (A) and after (B) division of the MAL, showing subsequent resumption of celiac trunk circulation and normal organic perfusion (notice the change in the colour of upper abdominal organs). Simulation of compression of the MAL (A) and simulation of decompression of the MAL (B). Intraoperative trial clamping of the celiac trunk was performed after division of the MAL demonstrating breakdown of the circulation as in MALS.

Subsequently, blood tests gradually normalized. A follow-up CT angiography (Fig. 3), taken one month later, revealed normal calibre celiac axis and preserved abdominal organic perfusion, with a residual splenic infarct scarring. The patient did not experience any major complications.

Discussion

The definition of the MALS relies on a combination of both clinical and radiographic features. It typically occurs in young patients (20–40 years of age) and is more common in thin women, who may present epigastric pain, usually intermittent, and weight loss;[6,2] additionally, abdominal pain may be postprandial, but not always.[7]

The pathophysiology of MALS is poorly understood. Loukas et al.[8,9] observed some variability in terms of anatomy, morphology and composition (muscular vs. tendinous) of the diaphragmatic crura. They hypothesized that these variations of the diaphragmatic crura, especially in their contribution to the aortic hiatus and oesophageal, could be the physiopathological basis of MALS, hiatal hernia and gastroesophageal reflux.

When the MAL passes anterior to the celiac axis, instead of above, a characteristic superior indentation is noted usually about 5 mm from the artery origin. Any compression caused by the MAL typically is less apparent during inspiration, when the celiac axis assumes a more caudal position; conversely, it increases during expiration.[1] Surprisingly, up to 13%–50% of healthy patients may exhibit the angiographic feature of compression to a variable

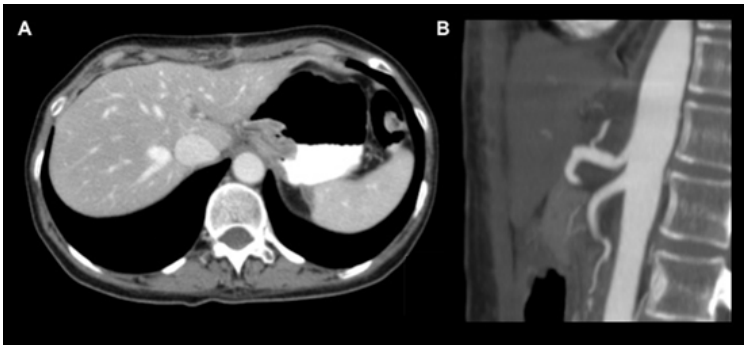

Figure 3 – Contrast-enhanced follow-up CT angiography, taken one month after surgery. Axial view (A) and sagittal reconstruction (B) of the upper abdomen, showing respectively a residual small area of splenic infarct scarring (A) and a normal calibre celiac axis (B) after MAL division.

degree.[10,11] In the vast majority, this is an isolated finding and it is not clinically significant.[1] Severe compression occurs in approximately 1% of patients and persists during inspiration.[11,12] Therefore, studies for diagnosing MALS should ideally be performed in the end-inspiratory phase.[2] Severe stenosis will result in poststenotic dilatation and, in some cases, collateral vessels, develop from the superior mesenteric artery branches.[1] These associated findings may suggest hemodynamic significance and are important for surgical planning.

Although the diagnosis of celiac artery compression was formerly made by conventional angiography, computed tomography (CT) angiography is nowadays the preferred technique. CT can detect significant arterial stenosis with 96% sensitivity and determine the aetiology of celiac axis stenosis with 92% accuracy.[13] Multiplanar reconstructions should be performed, as the sagittal plane shows focal narrowing with a characteristic hooked appearance caused by the inferior displacement of the proximal celiac axis by the MAL. Additionally, the inexistence of calcified parietal plaques helps to exclude atherosclerotic disease.[1] Doppler ultrasound is also a useful imaging modality and may show an increased peak systolic velocity of greater than 200 cm/s, which has a reported sensitivity of 75% and specificity of 89% in detecting a stenosis of at least 70%.[14] Additionally, flow turbulence which is accentuated during the expiratory phase, can be assessed.[14]

To the best of our knowledge, the potential association between MALS and Nissen fundoplication surgery has not previously been documented in the literature. We theorize that surgical manipulation around the celiac trunk might have induced the stenosis. However, we don't know whether the patient had a predisposing anatomical constitution, since no pre-surgical imaging work-up is available. Interestingly, according to two reports, it seems that MALS can develop acutely after pancreaticoduodenectomy even when all preoperative and intraoperative evaluations were normal.[15] Since the appearance of iatrogenic induced MALS after surgery is very rare, the appropriate treatment has to be determined according to the patient's general status.[15] We decided on exploratory laparotomy since our patient was extremely prostrated, experiencing severe hepatic cytolysis and CT showed widespread liver, gastric and splenic ischaemia. The median arcuate ligament surgical division is a safe and fast procedure that allows trunk decompression and resolution of ischaemic disorders.[13]

In conclusion, hepatic cytolysis after Nissen Fundoplication surgery should alert clinicians to the possibility of MALS.[15] CT is extremely useful in finding the characteristic hooked stenotic appearance of the proximal celiac axis, as well as in showing indirect signs of ischaemia in this arterial territory. Patients who acutely deteriorate despite medical treatment should undergo surgical exploration to rule out other conditions or accomplish division of the MAL and prevent any major complications if MALS is indeed present.[15]

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Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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