

Case Report

Median Arcuate Ligament Syndrome (Dunbar Syndrome): The Importance of the Pre-Operative Study in Surgical Planning

Síndrome do Ligamento Arqueado Mediano: A Importância do Estudo Pré-Operatório no Planeamento Cirúrgico

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ABSTRACT

The median arcuate ligament syndrome, also known as Dunbar syndrome, is caused by stenosis of the celiac trunk due to compression by diaphragmatic fibres. The incidence of this condition remains unclear and its clinical presentation is multifarious. Diagnosis is established by angio-computed tomography to determine the extent of stenosis and the existence or not of collateral circulation. Surgical intervention remains the favoured treatment. We present the case of a 40-year-old man diagnosed with a cephalopancreatic neuroendocrine tumour proposed for a pancreatoduodenectomy. Preoperative imaging studies revealed significant celiac-mesenteric collateral circulation, raising doubts about whether it was caused by a celiac trunk agenesis versus a

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median arcuate ligament syndrome. This finding would significantly influence the choice of surgical strategy, particularly the need for vascular anastomoses or not. We hereby present its intraoperative findings and the surgical solution, enhancing the importance of the preoperative imaging studies.

Keywords: Celiac Artery/surgery; Median Arcuate Ligament Syndrome/diagnostic imaging; Median Arcuate Ligament Syndrome/surgery

RESUMO

A síndrome do ligamento arqueado mediano, também conhecido como síndrome de Dunbar, é causada por estenose do tronco celíaco devido à compressão por fibras diafragmáticas. A incidência desta patologia permanece indefinida e a sua apresentação clínica é variada. O diagnóstico é estabelecido por angio-tomografia computadorizada, com o objetivo de determinar o grau de estenose e a presença ou não de circulação colateral. A intervenção cirúrgica continua a ser o tratamento de eleição. Apresentamos o caso de um homem de 40 anos diagnosticado com um tumor neuroendócrino cefalopancreático, proposto para duodenopancreatectomia. Os exames imagiológicos pré-operatórios revelaram circulação colateral celíaco-mesentérica significativa, levantando dúvidas quanto à possibilidade de se tratar de uma agenesia do tronco celíaco versus um síndrome do ligamento arqueado mediano. Este achado teria um impacto relevante na estratégia cirúrgica a adotar, nomeadamente quanto à necessidade ou não de anastomoses vasculares. Apresentam-se os achados intraoperatórios e a solução cirúrgica adotada, destacando a importância dos exames de imagem pré-operatórios.

Palavras-chave: Artéria Celíaca/cirurgia; Síndrome do Ligamento Arqueado Mediano/cirurgia; Síndrome do Ligamento Arqueado Mediano/diagnóstico por imagem

INTRODUCTION

The median arcuate ligament syndrome (MALS, also known as Dunbar's syndrome) is a rare condition caused by compression of diaphragmatic fibres at the aortic hiatus at the level of T12-L1 vertebrae, compromising blood flow at the celiac-mesenteric region. Its incidence is not yet well defined, although it is more frequent among female patients and in the age range between 30-50 years.^{1,2} Most of the patients are asymptomatic. More than 50% of stenosis at the celiac trunk may only cause symptoms in 12.5% of the patients.³ However, clinical complaints increase with stenosis severity, mostly related to chronic bowel ischemia (fear of eating, postprandial pain and weight loss). The gold standard for diagnosis is an angiography, but the majority of patients are diagnosed after an angiography computed tomography (angio-CT) scan performed for other purposes.⁴ It was estimated that nearly 42% of asymptomatic patients had collateral circulation seen in the celiac-mesenteric arch. Besides, these patients have a higher incidence of aneurysms of the splenic artery, celiac trunk and pancreato-duodenal arch.^{5,6} Surgery should be the preferred treatment, either through laparoscopic, robotic, endovascular or open approaches. It consists on the decompression of the diaphragmatic fibres, gradual dilation of the celiac trunk or, lastly, with a bypass reconstruction.⁷

CASE REPORT

We present the case of a 40-year-old man, smoker and no other relevant past medical history, who presented with progressively involuntary weight loss over the course of 3 months. He performed an upper endoscopy, which revealed superficial gastritis. A toraco-abdomino-pelvic CT scan showed a hypervascular cephalo-pancreatic mass measuring 2.7x3 cm. No dilation of the biliary tract was seen, nor distant metastasis. An endoscopic ultrasound was performed, revealing a nodular hypoechogenic lesion with well-defined borders located in the uncinate process of the pancreas. Biopsies were made and a neuroendocrine tumour was diagnosed. Therefore, the patient was proposed for a pancreatoduodenectomy (PD) and a pre-operative study was performed. The CT scan previously performed showed an increase in the gastroduodenal artery diameter and an important collateral circulation from the superior mesenteric artery. Vascular reconstruction images showed no emergence of the celiac trunk from the aorta, suggesting an agenesis of the celiac trunk (Fig. 1). Arteriograms of the splenic artery, superior mesenteric artery and arc of Bühler were also performed (Figs. 2 a-c). Taking this into account, we could not confirm whether the patient had MALS or a celiac trunk agenesis. Therefore, the surgery could either occur as a conventional

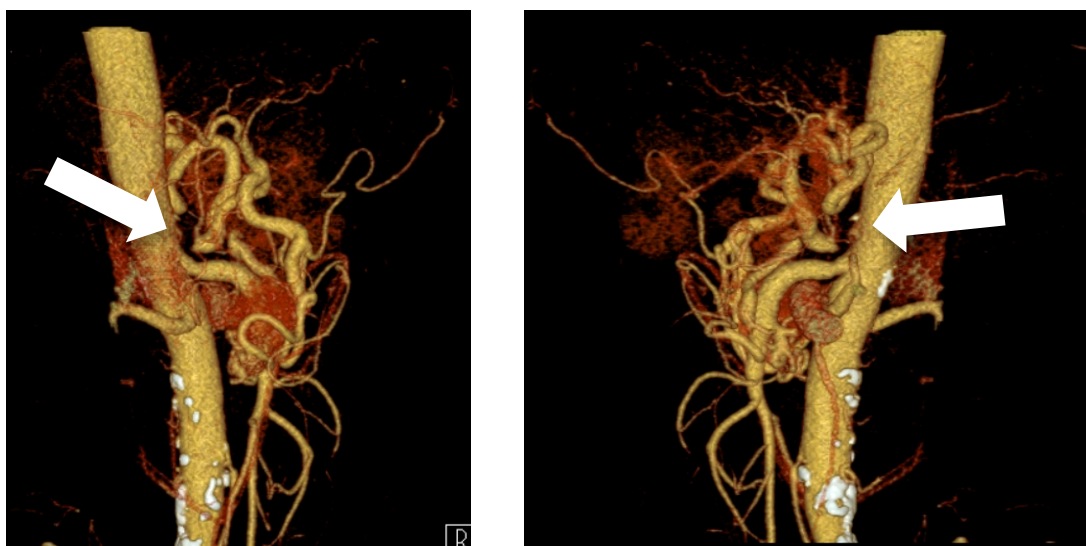


Figure 1. Vascular reconstruction images suggesting agenesis of the celiac trunk, right above the emergence of the superior mesenteric artery (arrows).

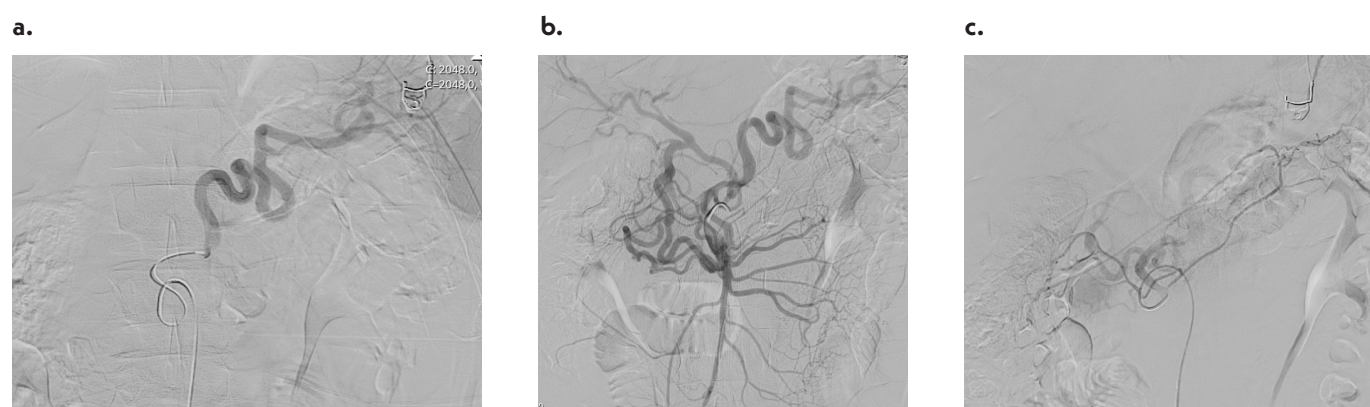


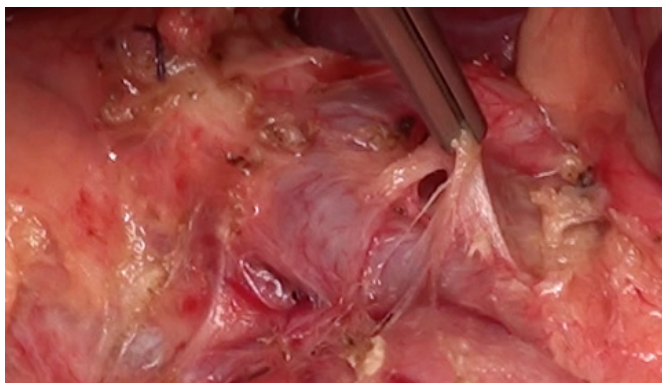
Figure 2. Arteriograms of the splenic artery (a), superior mesenteric artery (b) and arc of Buhler (c).

PD or it would force the surgeon to perform a resection with vascular reconstruction, given the anatomical variations.

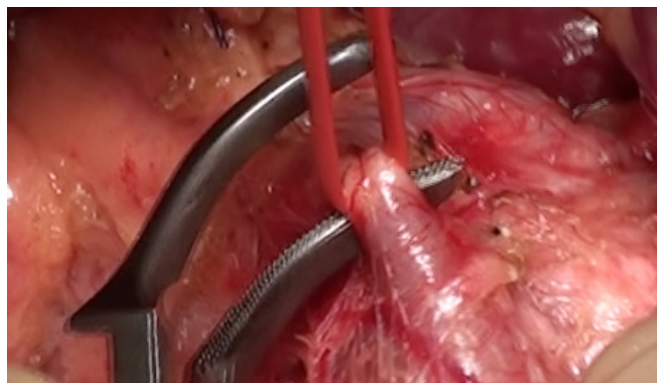
The patient is submitted to laparotomy. Intraoperatively, the gastroduodenal artery exhibited dilation and the emergence of a celiac trunk was difficult to identify (Figs. 3 a-g). Following the release of the aortic hiatus and clamping of the

gastroduodenal artery, the pulse of the splenic and common hepatic arteries persisted, thereby confirming irrigation from the celiac trunk and, therefore, the diagnosis of MALS. This finding was also confirmed with Doppler ultrasound during surgery. A conventional PD was performed uneventfully. The patient started oral diet on the fourth day after surgery with good tolerance and got discharged on day seven.

a.



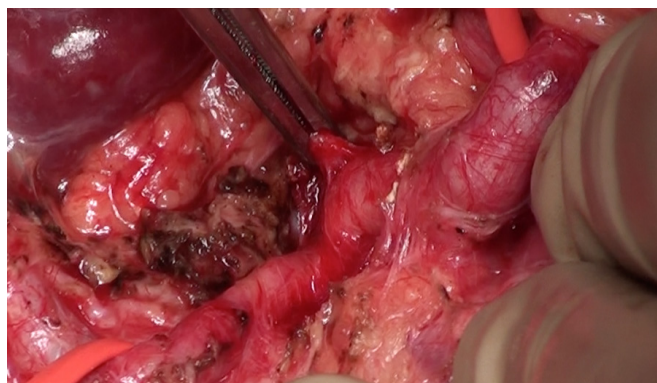
b.



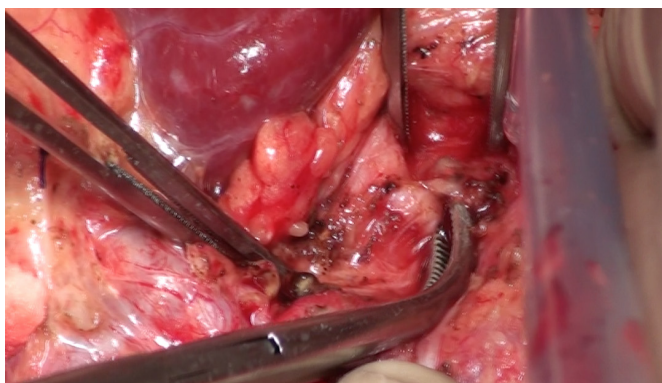
c.



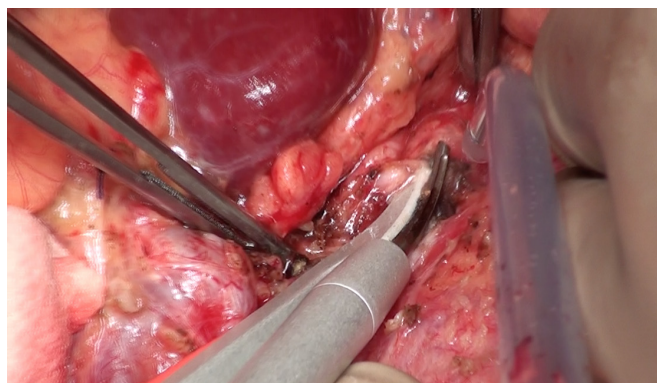
d.



e.



f.



g.

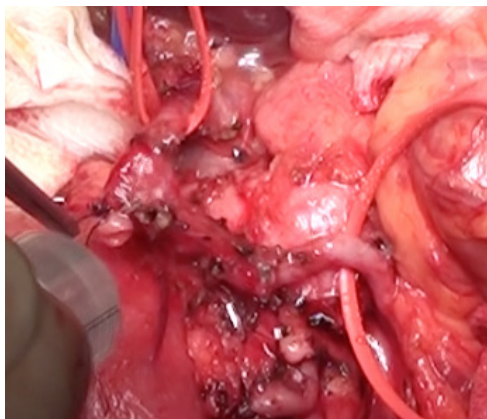


Figure 3. Intraoperative findings: dilation of the gastroduodenal artery (a); clamping of the gastroduodenal artery and confirmation of arterial pulse at the CHA and splenic artery (b); identification of the emergence of the celiac trunk and the arcuate median ligament (c); referencing of the CHA and splenic artery (d); section, decompression and liberation of the median arcuate ligament (e and f); ligation of the gastroduodenal artery and verification of the artery pulse at the celiac trunk (g).

DISCUSSION

This case is an important example of how preoperative imaging studies can predict certain diagnoses and help prepare the surgical strategy. If a celiac trunk agenesis was found, the celiac circulation would be exclusively given by the superior mesenteric artery through the gastroduodenal artery. To perform an oncologic resection of the duodenum and cephalic portion of the pancreas, the surgeon is forced to divide the gastroduodenal artery, thus compromising the organs supplied by the branches ahead (CHA common hepatic artery, LGA left gastric artery and splenic artery - if no other anastomosis were found). In order to maintain this circulation, the surgical solution would be an arterial bypass connecting the superior mesenteric artery to one of the other celiac branches (CHA, LGA or splenic artery) - Fig. 4. This would require an experienced surgeon in both pancreatic surgery and arterial anastomosis or a multidisciplinary approach along with vascular or plastic surgeons.

According to the literature, the incidence of MALS is unclear. As for the agenesis of the celiac trunk,⁸⁻¹¹ its incidence varies between 0.2% to 0.7% and, similar to MALS, most of the patients are asymptomatic. Collateral circulation is maintained by the superior mesenteric artery and some branches may arise individually from the aorta. This was not the case, though.

CONCLUSION

In conclusion, the asymptomatic manifestation of MALS can be incidentally discovered through imaging studies. This

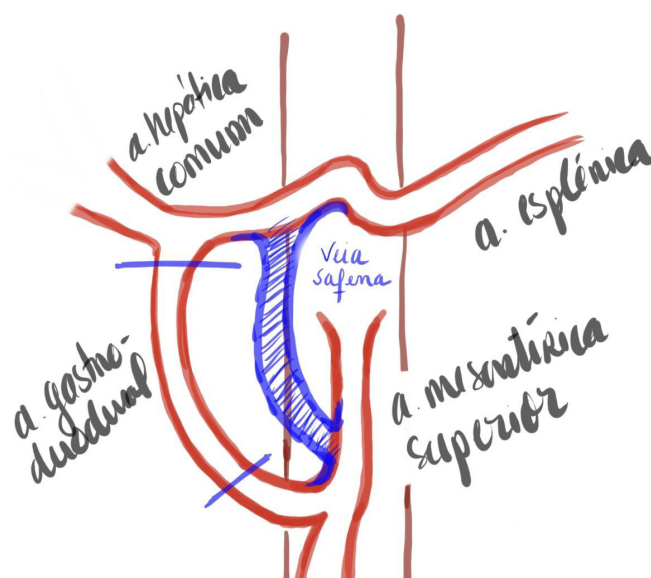


Figure 4. Arterial bypass using a venous conduct connecting the superior mesenteric artery to the other celiac branches (CHA, LGA or splenic artery) as a surgical solution in case of agenesis of the celiac trunk.
CHA – common hepatic artery; LGA – left gastric artery.

particular case serves as a prime example of the significance of preoperative assessment in the surgical planning of a patient slated for a PD, wherein the presence of MALS played a vital role in determining the surgical strategy.

ETHICAL DISCLOSURES

Conflicts of Interest: The authors have no conflicts of interest to declare.

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Patient Consent: Consent for publication was obtained.

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CONTRIBUTORSHIP STATEMENT

JPAT: Design of the study, data collection and analysis, writing of the manuscript.

HC, MA, CS and TBM: Participation in the imaging analysis, collaboration in the surgical discussion and critical revision of the content.

LG: Surgical supervision and critical revision of the article.

EB: Linguistic revision and final formatting of the article.

All authors approved the final version to be published.

DECLARAÇÃO DE CONTRIBUIÇÃO

JPAT: Conceção do trabalho, recolha e análise de dados, redação do manuscrito.

HC, MA, CS e TBM: Participação na análise imagiológica, colaboração na discussão cirúrgica e revisão crítica do conteúdo.

LG: Supervisão cirúrgica e revisão crítica do artigo.

EB: Revisão linguística e formatação final do artigo.

Todos os autores aprovaram a versão final a ser publicada.

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