CASE REPORT

Surgical treatment of Dunbar syndrome

Tratamento cirúrgico da síndrome de Dunbar

Luís Henrique Gil França[1], Carla Mottin[2]

Abstract

Dunbar syndrome or celiac artery compression syndrome is an infrequently described clinical condition with poorly defined diagnostic criteria and an obscure pathophysiology. It is usually associated with an extrinsic compression upon the celiac axis near its takeoff from the aorta by fibrous diaphragmatic bands or sympathetic neural fibers. The authors report the case of a 70-year-old male patient presenting with nausea, epigastric pain, and weight loss. An aortography showed a compression of the celiac trunk. A preliminary attempt at percutaneous transluminal angioplasty and stenting proved unsuccessful. The patient became asymptomatic and his clinical condition improved after surgical release of the celiac trunk by partial section of the arcuate ligament of the diaphragm and with resection of the neural, fibrotic, and lymphatic tissues surrounding the aortic and visceral vessels. The purpose of this report is to discuss the indications and the therapeutic options of this syndrome.

Keywords: arterial occlusive diseases; ischemia; celiac artery.

Resumo

A síndrome de Dunbar ou compressão do tronco celíaco é uma condição clínica infrequente, com poucos critérios para diagnóstico e com patofisiologia obscura. Está usualmente associada à compressão extrínseca do tronco celíaco por banda fibrosas do diafragma e fibras neurais simpáticas, próximo a sua emergência da aorta. Os autores relatam um caso de um paciente de 70 anos de idade com quadro de náuseas, dor epigástrica e perda de peso. Uma arteriografia mostrou compressão do tronco celíaco. Uma primeira tentativa de angioplastia com ***stent*** foi realizada em outro serviço, mas sem sucesso. Após o tratamento cirúrgico que consistiu de secção parcial do ligamento arqueado do diafragma com ressecção dos tecidos fibróticos, neurais e linfáticos que circundavam a aorta e as artérias viscerais, o paciente obteve melhora clínica e tornou-se assintomático. O objetivo deste estudo é discutir as indicações e opções terapêuticas desta síndrome.

Palavras-chave: arteriopatias oclusivas; isquemia; artéria celíaca.

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Conflict of interest: None
Financial support: No conflicts of interest declared concerning the publication of this article.
Submitted on: 18.05.11. Accepted on: 11.07.12.

Research performed at the Surgical Department (Vascular Surgery) of Hospital Geral (Exército) of Curitiba (HGeC). Curitiba, PR, Brazil.

INTRODUCTION

Celiac artery compression syndrome (CACS) (also referred to as celiac axis syndrome, median arcuate ligament syndrome and Dunbar syndrome) is defined as abdominal pain related to compression of the celiac artery by fibers of the median arcuate ligament[1,2]. Initially described in the 1960s, it is an uncommon disorder that is characterized by the triad of postprandial abdominal pain, weight loss, and sometimes an abdominal bruit[3]. The etiology is incompletely understood and its real existence is questionable. The symptoms of this disorder are similar to those of atherosclerotic chronic intestinal ischemia. This condition is most frequent in women aged 40 to 60 years old and its natural history may involve weight loss and severe malnutrition[2,4]. Lateral aortography is the primary modality for diagnosing ligamentous compression of the celiac artery. Abdominal computed tomography (CT) can show abnormalities in diaphragm and mesenteric vessels[3-6]. Duplex Doppler sonography performed during deep expiration can demonstrate a marked increase in flow velocities at the compressed region of the celiac artery, suggesting the diagnosis of celiac arterial constriction due to the diaphragmatic ligament[7]. In this report we discuss the indications and the therapeutic options of this syndrome as well as a review of the literature is being given.

CASE REPORT

We report a case of a 70-year-old man who presented with a three-month history of postprandial abdominal pain, diarrhea, weight loss and a bruit in the upper mid-epigastrium. This patient underwent psychiatric evaluation, which did not reveal a psychiatric disorder. CACS by median arcuate ligament was suspected after abdominal CT (Figure 1), which revealed a significant stenosis of the celiac artery with slight poststenotic dilatation and hypertrophy of the arcuate ligament. Lateral aortography confirmed the presence of stenosis in the celiac trunk (Figure 2). Transabdominal Doppler ultrasound (US) scanning revealed an increased peak systolic velocity to 4 m/sec at the origin of the celiac trunk with a low distal monophasic flow increasing during deep expiration. The patient underwent angioplasty and stenting in other hospital without complications, but his clinical status did not improve. There were no records of pressure measurements during angiography in the previous medical data of the patient. A new transabdominal Doppler US showed patency of the stent with little improvement in distal flow. After this, the patient was referred to the Vascular Section of our hospital - Hospital Geral (Exército) de Curitiba - and, after evaluating his medical data, we proposed surgical treatment for him. Surgery was performed with the patient under general anesthesia. An epigastric fan retractor was used to elevate the left lobe of the liver and the stomach fell down by gravity. The gastrohepatic ligament was divided, and beating movements identified the celiac trunk. In this way, the left gastric, common hepatic, and lienal arteries were identified; with a combined blunt and cutting dissection by shears, arteries were exposed at their origins from the aorta. Intraoperatively, the clear extrinsic nature of compression of the celiac trunk by the diaphragmatic structures was well assessed visually and compressing pathologic muscular fibers were divided; the ligament was excised with resection of the neural, fibrotic, and lymphatic tissues surrounding the aortic and visceral vessels (Figures 3, 4 and 5). Nissen fundoplication was performed to treat gastroesophageal reflux disease and hernia

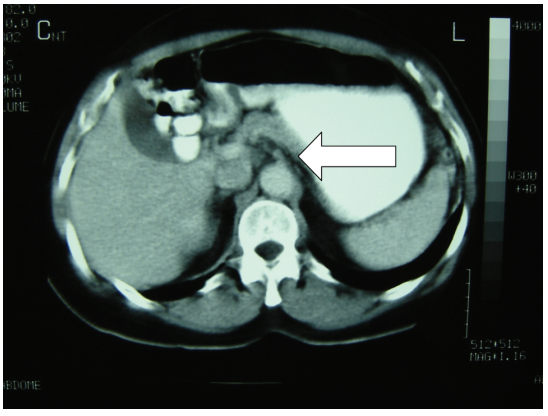

Figure 1. Computed tomography showing stenosis of the celiac trunk (white arrow).

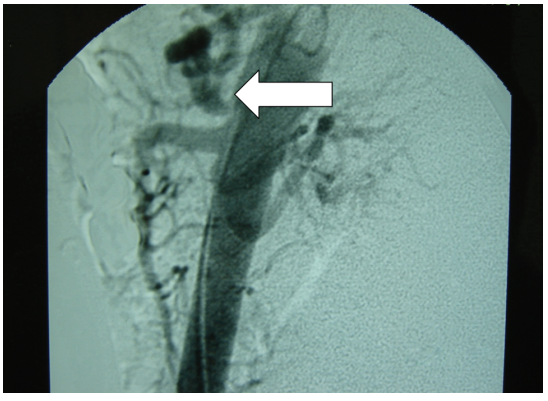

Figure 2. Angiography in a lateral view showing moderate stenosis of the celiac trunk (white arrow).

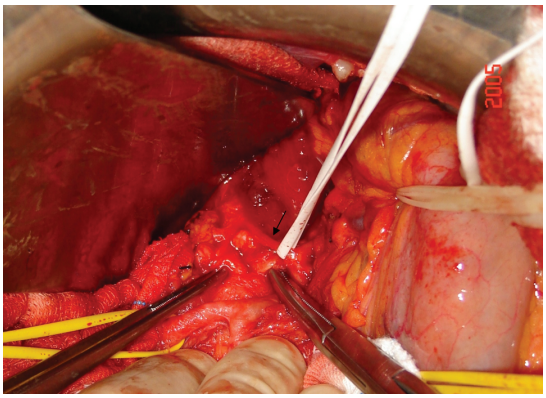

Figure 3. Dissection of the celiac trunk (black arrow).

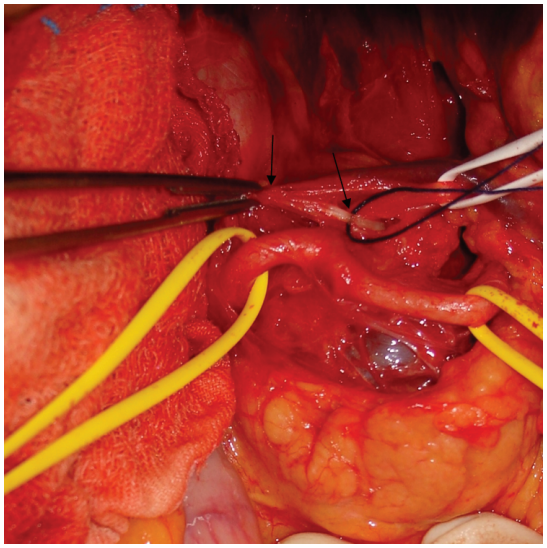

Figure 4. Dissection of the fibers of the arcuate ligament and the neural, fibrotic, and lymphatic tissues surrounding the aortic and visceral vessels (black arrow).

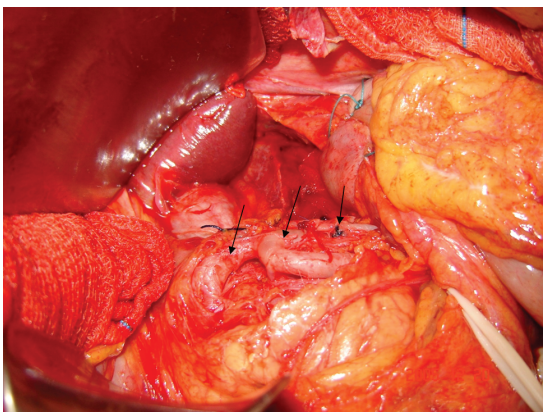

Figure 5. Aspect of the celiac trunk and branches (black arrows) after resection of the arcuate ligament and the neural, fibrotic, and lymphatic tissues surrounding the aortic and visceral vessels.

as an additional procedure. Control angiographies reveal no signs of compression of the celiac axis and patency of the stent (Figures 6 and 7). The patient remains well and free of symptoms two and a half years since operation.

DISCUSSION

In 1963, Harjola[3] described a case of chronic abdominal pain in a young woman that he attributed to mesenteric ischemia caused by extrinsic compression of the celiac artery. Since then, the topic has been the focus of numerous controversies regarding its pathophysiology, definitive diagnosis, and optimal treatment[4]. There is still considerable doubt about the real existence of the CACS. Asymptomatic compression or stenosis of the celiac artery is common, and compression of this artery by fibers of the median arcuate ligament has been

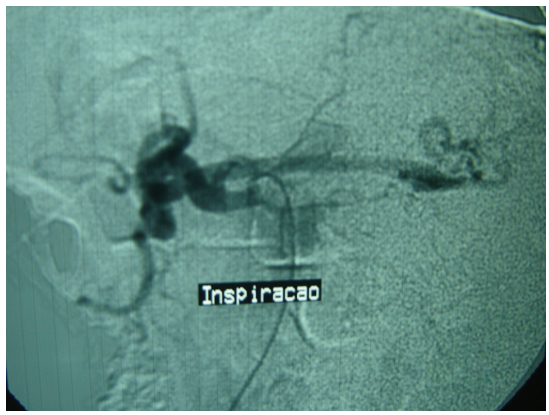

Figure 6. Post-procedure angiography.

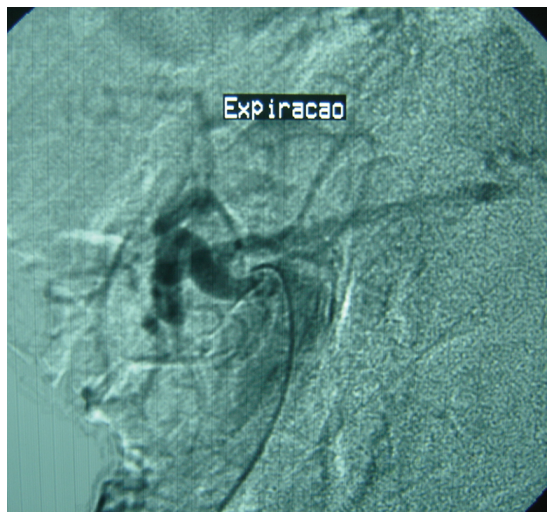

Figure 7. Post-procedure angiography.

demonstrated on arteriography in asymptomatic patients[1]. Similarly, autopsy studies have shown that the celiac artery is compressed by the median arcuate ligament in up to one-third of individuals[5]. The difficulty in interpreting reports of CACS is that not all studies used the same definition for the disorder. Many studies included patients with a range of abdominal complaints and those with a variety of medical (and often psychiatric) problems[4-6]. Thus, it is not surprising that the results of surgical treatment have also been variable. Nevertheless, most authors agree that carefully selected patients, in whom other causes of abdominal pain have been thoroughly exhausted, will benefit from operative treatment[8-10].

There are probably two causes for CACS in symptomatic patients: vascular and neurologic disorders. In patients with vascular disorders (classic chronic mesenteric ischemia), two of the three mesenteric vessels must typically be occluded or severely stenotic for the patient to experience symptoms of abdominal pain because of the extensive collateral network for the bowel. By contrast, in CACS, the superior mesenteric artery and the inferior mesenteric artery are widely patent, thereby, in theory, providing an ample blood supply to the bowel. These results indicate that compression of the celiac axis may be merely an incidental angiographic finding, so this syndrome needs cautious evaluation[4,6].

In the group of patients with neurologic disorder, anomalous fibrous diaphragmatic bands overlying the celiac trunk and the superior mesenteric artery may be implicated. Celiac plexus, enlarged lymphatic and sympathetic neural fibers may compress the celiac trunk or the superior mesenteric artery, causing gastric and intestinal ischemia with symptoms. As a result, many have suggested that the symptoms may not be related to blood flow but rather to involvement of the splanchnic nerve plexus, which lies in the same region as the diaphragmatic fibers. In this theory, pain relief with decompression of the celiac artery results not from improvement in postprandial flow but from destruction of the splanchnic nerves during the surgical exposure of the artery[1,2].

CACS may be investigated with Doppler US, spiral CT angiography, selective catheter angiography, and magnetic resonance angiography[6]. Doppler US has been reported to have a high sensitivity for the diagnosis of CACS and was proposed to be the modality of choice, although the gold standard diagnostic method is still selective angiography, which should be performed during both inspiration and expiration[7,8]. However, the implementation of multi-slice CT has permitted the acquisition of thinner images, which not only provided increased resolution and improved lesion detection but also has permitted the production of excellent multiplanar reconstruction[9].

Angioplasty and stenting of visceral vessels have been described as being a reasonable tool in the treatment of atherosclerotic disease,[10] but their use as the sole mode of treatment in celiac artery compression appears questionable, particularly because extrinsic compression may prevent adequate dilatation of the vessel and, if there is no hemodynamic involvement and the symptoms are due to the involvement of the splanchnic nerve plexus, the role of endovascular treatment is out of question[11,12]. However, their role in combination with decompression has not been investigated and is speculative. We believe that treatment of CACS is primarily surgical, but stent insertion may have a role as a secondary procedure where there is a residual stenosis after prior release of the extrinsic compression on the celiac artery by dividing the median arcuate ligament and ganglionic tissue with open or laparoscopic surgery. Also, it is important to verify the patency of the mesenteric arteries to indicate stenting of the celiac trunk correctly. In the present case, stenting of the celiac trunk was not effective probably because of poor diagnostic evaluation of the patient.

Conventional surgical treatment with division of the median arcuate ligament and excision of celiac plexus is adequate in patients without persistent vessel deformity or pressure gradient after decompression. In 1985, Reilly et al.[13] published the largest and most comprehensive series on this disorder and clearly demonstrated that celiac reconstruction must be performed in patients who have persistent vessel deformity, persistent thrill, or persistent pressure gradient after decompression. Takach et al.[14], in their series of seven patients, demonstrated that any operation performed for treatment of celiac artery compression must include assessment of the artery after decompression for adequacy of flow. Ghosn et al[15] showed that the pain in this syndrome cannot be explained solely on hemodynamic grounds and surgical treatment should include release of the celiac axis and complete periarterial neurectomy. Actually, some authors have reported good results with laparoscopic surgery for median arcuate ligament syndrome, arguing that this might be a less invasive approach to this condition[16,17]. It has become clear from the more recently published series on CACS that simple

revascularization of the celiac axis may not be adequate for all patients with this condition and, although good results can be achieved by operative treatment in selected patients, there is no agreement on whether surgical treatment is justified[14-17]. In the present case, the patient probably had symptoms more related to neurologic disorder previously listed than significant hemodynamic vascular disease, which might be better treated with open or laparoscopic surgery.

In conclusion, open surgery for CACS is an effective and safe treatment of lesions with proper morphologic features in selected patients. We believe, as many authors do, that patients without compromised celiac trunk after decompression may benefit from division of the median arcuate ligament and resection of surrounding tissues without some adjunctive form of celiac revascularization. Although there are few cases reported in the literature, the long-term follow-up results of this therapy show good results.

REFERENCES

1. Lindner HH, Kemprud E. A clinicoanatomic study of the arcuate ligament of the diaphragm. Arch Surg. 1971;103:600-5. http://dx.doi.org/10.1001/archsurg.1971.01350110102016
2. Snyder MA, Mahoney EB, Rob CG. Symptomatic celiac stenosis due to constriction by the neurofibrous tissue of the celiac ganglion. Surgery. 1967;61:372-6. PMid:6021256.
3. Harjola PT. A rare obstruction of the coeliac artery. Ann Chir Gynaecol Fenniae. 1963;52:547. PMid:14083857.
4. Bech FR. Celiac artery compression syndromes. Surg Clin North Am. 1997;77:409-24. http://dx.doi.org/10.1016/S0039-6109(05)70558-2
5. Evans WE. Long-term evaluation of the celiac band syndrome. Surgery. 1974;76:867-71. PMid:4428351.
6. Geelkerken RH, Van Bockel JH. Mesenteric vascular disease: a review of diagnostic methods and therapies. Cardiovasc Surg. 1995;3:247-60. http://dx.doi.org/10.1016/0967-2109(95)93872-M
7. Erden A, Yurdakul M, Cumbur T. Marked increase in flow velocities during deep expiration: A Doppler sign of celiac artery compression syndrome. Cardiovasc Intervent Radiol. 1999;22:331-32. PMid:10415466. http://dx.doi.org/10.1007/s002709900399
8. Gruber H, Loizides A, Peer S, Gruber I. Ultrasound of the median arcuate ligament syndrome: a new approach to diagnosis. Med Ultrason. 2012;14(1):5-9.
9. Horton KM, Talamini MA, Fishman EK. Median arcuate ligment syndrome: Evaluation with CT angiography. Radiographics. 2005;25:1177-1182. PMid:16160104. http://dx.doi.org/10.1148/rg.255055001
10. Spies JB, Bakal CW, Burke DR, et al. Standards of practice committee, Society of Cardiovascular and Interventional Radiology: guidelines for percutaneous angioplasty. J Vasc Interv Radiol. 1990;1:1-15.
11. Cina CS, Safar H. Successful treatment of recurrent celiac axis compression syndrome. A case report. Panminerva Med. 2002;44:69-72. PMid:11887094.
12. Delis KT, Gloviczki P, Altuwaijri M, McKusick MA. Median arcuate ligament syndrome: open celiac artery reconstruction and ligament division after endovascular failure. J Vasc Surg. 2007;46:799-802. PMid:17903658. http://dx.doi.org/10.1016/j.jvs.2007.05.049
13. Reilly LM, Ammar AD, Stoney RJ, Ehrenfeld WK. Late results following operative repair for celiac artery compression syndrome. J Vasc Surg. 1985;2:79-91. PMid:3965762.
14. Takach TJ, Livesay JJ, Reul GJ, Cooley DA. Celiac compression syndrome: tailored therapy based on intraoperative findings. J Am Coll Surg. 1996;183:606-10. PMid:8957463.
15. Ghosn PB, Rabbat AG, Trudel J, D'Amico P, Lecours R, Trudel J. Celiac compression syndrome. Can J Surg. 1982;25(4):377-9. PMid:7093835.
16. Nguyen T, Neale M, Lane R, Schiavone V, Samra JS, Hugh TJ. Laparoscopic management of the median arcuate ligament syndrome. ANZ J Surg. 2012;82(4):265-8. PMid:22510185. http://dx.doi.org/10.1111/j.1445-2197.2011.05966.x
17. Tsujimoto H, Hiraki S, Sakamoto N, et al. Laparoscopic treatment for median arcuate ligament syndrome: the usefulness of intraoperative Doppler ultrasound to confirm the decompression of the celiac artery. Surg Laparosc Endosc Percutan Tech. 2012;22(2):e71-5. PMid:22487643. http://dx.doi.org/10.1097/SLE.0b013e318243a3ab

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Analysis and interpretation: LHGF
Data collection: LHGF, CM
Writing the article: CM, LHGF
Critical revision of the article: LHGF
Final approval of the article*: LHGF
Statistical analysis: not applicable
Overall responsibility: LHGF

*All authors have read and approved the final version submitted to J Vasc Bras.