ORIGINAL ARTICLE

Median arcuate ligament syndrome: vascular surgical therapy and follow-up of 18 patients

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Abstract
Introduction The median arcuate ligament syndrome
(MALS) or celiac artery compression syndrome is a rare
vascular disorder caused by an extrinsic compression of the
celiac artery from the median arcuate ligament, prominent
fibrous bands, and ganglionic periaortic tissue. Clinical
symptoms are postprandial abdominal pain, nausea, vomit-
ing, unintentional weight loss, and sometimes, abdominal
pain during body exercise caused by an intermittent visceral
ischemia. The aim of this study was to evaluate the
operative management of patients with MALS in our
institution, especially in consideration of various vascular
reconstructive techniques.
Patients and methods Between June 2000 and January
2009, a total of 341 patients were treated in our department
for vascular pathologies of the visceral arteries (225 chronic
visceral ischaemia, 84 acute visceral ischaemia, and 14

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visceral artery aneurysms). In a retrospective study of 18
patients with MALS, the records, clinical symptoms, diag-
nostic evaluation, and surgical procedures were compiled.
This was completed by a reassessment for a follow-up.
Results A MALS was diagnosed in 15 female (83.3%) and
three male (16.7%) patients. The mean patient age was
46.2 years (range 20–68 years). The diagnosis of MALS
was based on a radiological analysis in all patients by a
digitally subtracted angiogram, but duplex ultrasound was
used lately more frequently to study the influence of
respiration on the stenotic degree of the celiac trunk. All
18 patients were treated with open surgery in an elective
situation. Due to the local and specific pathology of the
celiac trunk with a fixed stricture or stenosis, out of 18
cases beside decompression, 11 (primary, seven; secondary,
four patients) further procedures were performed on the
celiac artery (aorto-celiac vein interposition $n=6$, aorto-
hepatic vein interposition $n=1$, resection of the celiac artery
and end-to-end anastomosis $n=2$, patchplasty of the celiac
artery with vein $n=1$, and transaortic removal of a stent of
the celiac artery $n=1$) Follow-up was obtained in 15
patients (83.33%) with a mean duration after surgery of
about three and a half years (40.68 months, range from 2 to
102 months). Eleven of the 15 patients (73.33%) were
completely free of abdominal symptoms, and nine of them
had gained between 3 and 10 kg in weight after surgery.
The weight of two patients remained stable. Of the 11
patients with a successful outcome in the follow-up, six of
them had undergone decompression solely. In the other five
patients, vascular co-procedures on the celiac trunk had
been performed.
Conclusions The MALS is a rare vascular disorder caused
by an extrinsic compression of the celiac artery and induces
upper abdominal, mostly, postprandial pain. A definite
diagnosis of MALS can be achieved by lateral aortography

of the visceral aorta and its branches during inspiration and expiration. Open surgical therapy is a safe and reliable procedure with no mortality and low morbidity. As to the local and specific pathology of the celiac trunk after decompression with fixed stricture or stenosis, further vascular procedures are necessary. The long-time follow-up seemed adequate. The laparoscopic approach reduces the procedure of decompression only, something which seemed inadequate for most cases. Endovascular treatment with percutaneous transluminal angioplasty and insertion of a stent does not solve the underlying problem of extrinsic compression of the celiac trunk and often requires open procedures during the long-term course. Due to the low incidence of MALS, no guidelines will do justice to all the patients sufficiently, and the choice of treatment must depend on the specific clinical situation for each patient.

Keywords Median arcuate ligament syndrome ·
Celiac artery compression syndrome · Dunbar syndrome ·
Celiac axis compression syndrome · Celiac band compression syndrome · Intestinal ischemia · French bypass

Introduction

The median arcuate ligament syndrome (MALS) or celiac artery compression syndrome (CACS) is a rare vascular disorder caused by an extrinsic compression of the celiac artery from the median arcuate ligament, prominent fibrous bands, and ganglionic periaortic tissue. The incidence is two per 100,000 patients with recurrent, nonspecific upper abdominal pain [1]. Mostly young woman with asthenic habitus are affected. Clinical symptoms include postprandial abdominal pain, nausea, vomiting, and unintentional weight loss caused by an intermittent visceral ischemia. On examination, a typical clinical sign is epigastric bruits with its maximum during expiration. The distinctive radiological diagnostic tool is a lateral angiography of the visceral part of the aorta demonstrating a short segment of stenosis by external compression of the anterior wall of the proximal celiac trunk. Duplex ultrasound is used lately more frequently to study the influence of respiration on the stenotic degree of the celiac trunk.

With a view to history, B. Lipshutz was the first who described an external compression of the celiac trunk in 1917 [2]. Later on in the year 1963, P. T. Harjola reported about the causal connection between compression of the celiac artery and epigastric abdominal pain focusing on intestinal ischemia [3]. In 1965, the radiologist J. D. Dunbar reported in his article "Compression of the celiac trunk and abdominal angina" about the first series of patients with MALS and their diagnostic and therapeutic management [4]. In appraisal to the author of the first published series, the MALS or CACS is named *Dunbar syndrome* as well.

Consequently, no guidelines are available for the appropriate treatment of this rare vascular disorder. The diagnostic approach and therapeutic management has been reported in some case reports [5–16] and small series [1, 17–32]. The largest data collection was comprised of 51 patients with MALS and was published by Reilly et al. in 1985 [33]. Besides open surgery procedures, laparoscopic [10, 15, 34] and endovascular techniques [5, 9] have gained interest within the last decade. Their significance in the spectrum of various therapeutic concepts is still unknown because the number of patients treated to date is still small and as every new concept has to be compared to well-established methods.

The aim of this study was to evaluate the operative management of patients with MALS in our institution, especially in consideration of various vascular reconstructive techniques.

Patients and methods

Between June 2000 and January 2009, a total of 341 patients were treated in our department for vascular pathologies of the visceral arteries (225 chronic visceral ischaemia, 84 acute visceral ischaemia, and 14 visceral artery aneurysms). Of this group, a MALS was diagnosed in 18 patients. In a retrospective study of the patients' records, clinical symptoms, diagnostic evaluation, and surgical procedures were compiled, and the investigation was completed by a reassessment for follow-up.

Results

A MALS was diagnosed in 15 female (83.3%) and three male (16.7%) patients. The mean patient age was 46.2 years (range 20–68 years), In terms of gender, the mean female age was 47.9 years (range 20–68 years) while the mean male age was 37.7 years (range 26–54 years). The mean body mass index (BMI) in the collective was 21.1 (range 16.7–27.5). In terms of gender, female BMI was 20.7 (16.7–27.5), male BMI was 22.7 (20.0–26.8). All patients complained of abdominal pain. Cardiovascular risk factors were found in six patients (smoker $n=5$, arterial hypertension $n=5$, hyperlipoproteinaemia $n=2$, and diabetes $n=0$). As to the social-economic background, six out of 18 patients were teachers or educators, and four were physicians. All patients complained of abdominal pain of different intensity, nine of them of postprandial abdominal angina beginning from 30 min up to 3 h after meal. Further symptoms were nausea, vomiting, and diarrhoea. The most

Fig. 1 Lateral angiogram of the visceral segment of the aorta showing the celiac trunk during inspiration (*left*) and expiration (*right*) of a 35-year-old woman: in expiration, a short segment of stenosis by external compression of the anterior wall of the proximal celiac trunk demonstrates a median arcuate ligament syndrome

extreme documented loss of weight was 5 kg in a few weeks. Medical history revealed reflux oesophagitis $n=3$, pancreatic atrophy $n=1$, and a previous major abdominal surgery in one patient (gastrectomy due to Non-Hodgkin lymphoma). All patients were admitted to our hospital in an elective situation. The relevant blood tests for intestinal ischemia were normal: mean lactate, 0.91 mmol/l (range 0.5–1.6 mmol/l); mean leukocytes, 6.41/nl (range 4.4–

11.4/nl); and mean C-reactive protein, 1.71 mg/dl (range 0.0–9.8 mg/dl). The diagnosis MALS was performed radiological in all patients by digitally subtracted angiogram $n=18$, computed tomogram scan $n=2$, magnetic resonance scan $n=3$, and duplex ultrasound scan $n=5$ (multiple choices possible).

All 18 patients were treated surgically. The diagnosis MALS was confirmed intraoperatively after exposure of the celiac trunk with extrinsic compression from the median arcuate ligament, prominent fibrous bands, and ganglionic periaortic tissue. These tissues were dissected and partially resected in a way that the aorta and the celiac trunk were not constricted anymore. Due to the local and specific pathology of the celiac trunk with irreversible fixed stricture or stenosis, in seven out of 18 cases (38.9%), further procedures on the celiac artery were performed. Four patients had obtained an aorto-celiac vein interposition with greater saphenous vein and one patient an aorto-hepatic vein interposition after resection of a concomitant post-stenotic celiac aneurysm. One patient had a transaortic removal of a celiac stent with in-stent stenosis, and in one patient, a patchplasty with vein was performed to widen the lumen of the celiac artery (Figs. 1 and 2). In one case, the stenotic segment of the celiac trunk was resected and the continuity reconstructed by an end-to-end anastomosis. Moreover, a transaortic thromboendarterectomy of the renal arteries due to stenosis causing renovascular hypertension and an aorto-mesenteric loop-bypass ("French bypass" [35]) to treat a chronic intestinal ischemia were performed. The superior mesenteric artery (SMA) was involved in surgery in two further cases: in the first case, a misplaced stent was removed out of the SMA, and in the second case, a patchplasty with vein was performed to widen the lumen of the SMA in combination with a patchplasty of the left renal artery due to stenosis. One patient had a patchplasty

Fig. 2 Postoperative computed tomogram in volume rendering technique in expiration of the same patient after decompression and patchplasty with vein of the celiac trunk (left anterior-posterior, right oblique) showing a patent reconstruction without external compression of the celiac trunk
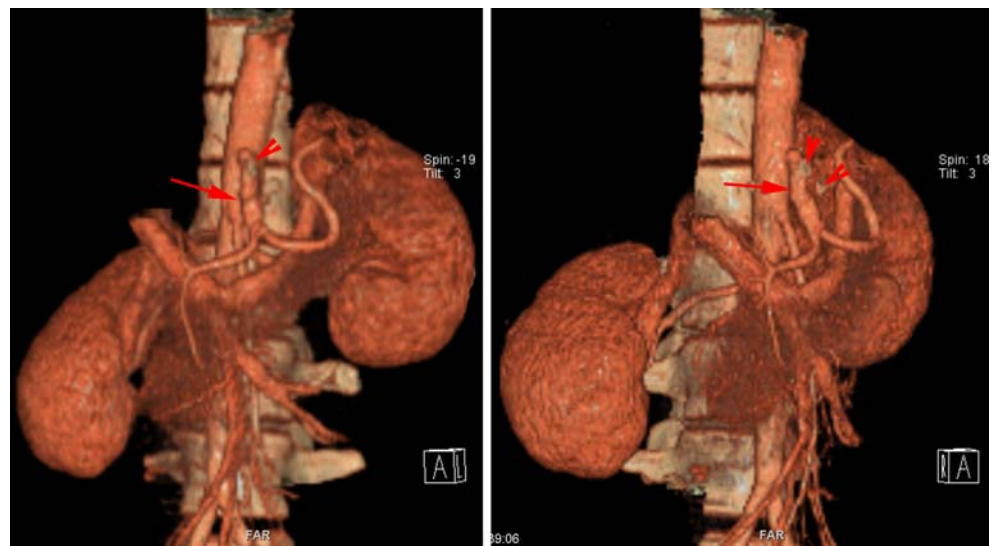
of both renal arteries due to stenosis in combination with the celiac decompression (Table 1).

In the early postoperative course, four patients underwent revision procedures. In two patients, a decompression of the celiac trunk alone was not sufficient, and the patient's complaints remained. An aorto-celiac vein interposition with great saphenous vein was performed on both patients on days 8 and 14, respectively, after the operation. Due to suspicion of an acute intestinal ischemia, a re-laparotomy was performed in one patient on the second day after the primary procedure but revealed no visceral malperfusion. The postoperative angiogram of the aorto-mesenteric loop-bypass ("French bypass") showed a stenosis of the distal anastomosis and was dealt successfully with by percutaneous angioplasty without insertion of a stent. One 20-year-old female student required further revisions after a transaortic removal of the celiac artery stent with in-stent stenosis. After an uneventful recovery without abdominal complaints, she developed postprandial abdominal symptoms 3 months later. The angiogram showed a stenosis of the celiac artery as a "reaction to injury" on stent removal. A transposition of the splenic artery in the supravisceral

aorta to prevent continuing visceral ischemia was performed. Eighteen months after the primary procedure, an aorto-celiac interposition with great saphenous vein was supplemented due to ongoing complaints. A recurrent stenosis of the anastomosis of the aorto-celiac graft was treated by a percutaneous angioplasty and insertion of a stent 43 months after the primary surgery. Seventeen months later (5 years from the first procedure), a hemodynamic significant stenosis around the stent led to a further revision with interposition of a vein graft in "Rheinländer" technique in an aorto-celiac position. Meanwhile, the patient is free of abdominal symptoms and gained some weight (Table 1).

Four patients had complications in their postoperative course without the need of re-do surgery. Two patients had remarkable neurological disorders, one patient had a distinctive delirium over some days, and the other had diplopic images and impaired vision on the left eye; the magnetic resonance imaging (MRI) scan did not reveal any pathological findings, and both patients were discharged fit and well on days 19 and 15, respectively, after the operation. One patient with sanguinous diarrhoea had a

Table 1 Surgical procedures to treat a median arcuate ligament syndrome MALS, simultaneous co-procedures, and revisions in 18 patients

Decompression of the CA	$n=18$ (n)	100%
Celiac artery	9	50%
Aorto-celiac vein interposition	4	
Aorto-hepatic vein interposition	1	
Resection of the CA and end-to-end anastomosis	2	
Due to stenosis	1	
Due to aneurysm	1	
Transaortic removal of a stent of the CA	1	
Patchplasty of the CA with vein	1	
Superior mesenteric artery	3	16.7%
Transaortic removal of a stent in the SMA	1	
Patchplasty of the SMA with vein	1	
Aorto-mesenteric loop-bypass ("French bypass")	1	
Renal artery	2	11.1%
Transaortic thromboendarterectomy of the RA	2	
Patchplasty of the RA with vein (multiple occurrences possible)	1	
Revisions	5	27.8%
Aorto-celiac vein interposition (eight respectively, 14 days postoperatively)	2	
Second look laparotomy due to suspected intestinal ischemia (2 days postoperatively)	1	
Angioplasty of the "French bypass" due to anastomosis stenosis (7 days postoperatively)	1	
Transposition of the splenic artery in the supravisceral aorta (3 months postoperatively) Aorto-celiac interposition with great saphenous vein (18 months after the primary procedure) Percutaneous angioplasty and stent anastomosis of the aorto-celiac graft (43 months after the primary procedure) Aorto-celiac interposition of a vein graft in "Rheinländer" technique (60 months after the primary procedure)	1	

CA celiac artery, SMA superior mesenteric artery, RA renal artery, TEA thromboendarterectomy

colonoscopy performed, but no spot of bleeding or ischemic segments of the colon was found. In one patient, the course of recovery was affected by cholestasis due to pancreatitis: he had an endoscopic retrograde cholangiopancreaticography with insertion of a stent in the pancreatic duct.

The mean in-hospital stay was 11.5 days (range from 5 to 27 days). At discharge, 17 out of 18 patients had complete relief of symptoms, while one patient complained of diarrhoea and recurrent nausea. All patients had a radiological examination of the celiac artery by digitally subtracted angiogram, computed tomogram scan, magnetic resonance scan, and/or duplex ultrasound scan to approve regular blood flow.

Follow-up

Out of this group of 18 patients, the follow-up in 15 patients (83.33%) was possible with a mean duration after surgery of about three and a half years (40.68 months, range from 2 to 102 months). Three patients were lost to follow-up. Eleven of the 15 patients (73.33%) were completely free of abdominal symptoms, and nine of them gained between 3 and 10 kg after surgery. The weight of two patients remained stable. Two of the 11 patients had undergone subsequent procedures in the mean time: one patient had an insertion of an intra-abdominal drain due to chyloperitoneum and temporarily parenteral feeding 2 months after surgery, while the other patient had a re-laparotomy for lysis of adhesions of the small bowel 18 months after the primary procedure. Of the 11 patients with a successful outcome in follow-up, six of them had decompression of the celiac trunk solely. The other five patients had vascular co-procedures on the celiac trunk ($n=1$ resection of a stenotic segment of the celiac trunk and end-to-end anastomosis, $n=3$ aorto-celiac vein interposition, $n=1$ aorto-hepatic vein interposition).

Four of the 15 patients still had abdominal symptoms of varying intensity and for various reasons. Two patients complained of intermittent epigastric pain not related to the intake of food and are under further diagnostic evaluation. Another patient with gastrectomy due to Non-Hodgkin lymphoma in her medical history had dysphagia due to a stenosis of the oesophagojejunostomy and iterates bouginage. The fourth patient is the 20-year-old female student with further surgical procedures and interventions as described above.

Discussion

In 1965, J. B. Dunbar [4] summarised his experiences on patients with MALS concerning diagnosis and treatment:

"Although no definite pathophysiologic explanation can be offered for the clinical symptomatology in the 15 cases with compression of the celiac trunk, the satisfactory and often striking response to the decompression of the artery suggests a direct correlation between the abdominal pain and the partial obstruction of the celiac artery". Out of 15 patients with MALS, 13 patients (12 females, one male, mean age 34.5 years, range 19–51 years) underwent surgery with decompression of the celiac axis. The postoperative course of all cases was uneventful, and during the follow-up, up to 15 months after surgery, all patients were free of their preoperative abdominal symptoms [4]. Since then, other experiences in the diagnostic evaluation and treatment have been reported.

Diagnostic evaluation

Besides the *golden standard* of lateral conventional angiography of the visceral aorta and its branches in function (inspiration and expiration), computed tomography (CT) angiography allows to visualise not only the stenosed vessel in the sagittal plane but also the underlying median arcuate ligament and adherent tissue using 3D imaging. Additionally, the multiplanar capabilities of 3D imaging are valuable for identifying the collateral vessels which develop as a result of MALS and give information on determining the hemodynamic significance of the narrowing [14, 36]. Lee et al. accented the worth of MRI in function of inspiration and expiration to discriminate between variations of the celiac trunk in the range of normal or pathological cause of narrowing of the proximal celiac segment [37].

Schuler et al. examined 126 patients under the age of 45 suffering from abdominal pain with colour-doppler ultrasound. Systolic peak velocities (Vmax syst) higher than 1.8 m/s and diastolic Vmax higher than 1 m/s in the celiac artery were found in 19 patients. In four patients, Vmax syst >1.8 m/s during inspiration and expiration was regarded as fixed stenosis and verified by digitally subtracted angiogram as MALS. Surgical therapy was successfully performed on these four patients [38]. Erden et al. performed duplex Doppler sonography during deep expiration and found an increase in flow velocities caused by a compressed segment of the celiac artery [39]. Walter reported a peak flow on the celiac axis of more than 200 cm/s during inspiration and expiration in addition to an angiogram in 15 patients leading to the diagnosis MALS [40]. Wolfman et al. pointed out that when evaluating the celiac trunk by colour-flow Doppler sonography, an erect view has to be included in order to prevent misdiagnosis of MALS [29].

Faries et al. reported about a patient suffering from MALS describing the application of gastric tonometry to

measure the gastric mucosal ischemia. The mucosa pH was 7.20 prior to celiac artery decompression and 7.48, post-operatively. The patient had complete relief of his abdominal symptoms and remained asymptomatic 14 months after surgery [11]. Gastric exercise tonometry as a helpful diagnostic tool in patients with MALS was published by Mensink et al. They reported about 30 patients with MALS causing ischemia of the upper intenstinum; 22 patients had a celiac trunk decompression only and further seven revascularisation. After a mean follow-up of 39 months, 83% of all treated patients were without symptoms. Repeating the gastric exercise, tonometry showed that it was normal in 100% of the pain-free-patients and in 25% in patients with persistent complaints [41].

Balaban et al. reported about a patient with documented gastropareses due to MALS focussing on gastric myoelectrical features. The surgical decompression of the celiac artery resulted in resolution of abdominal pain and in restoration of the gastric electrical rhythm to a normal 3 cycle/min [6].

In our investigation, all patients had had at least a conventional digitally subtracted angiogram of the visceral aorta and its branches to exclude further arteriosclerotic lesions of the visceral arteries.

Treatment: conservative/"wait and see"

In 1984, Gutnik reported about two patients suffering from MALS. Under conservative treatment, both patients did poorly, and the author suggested that they ought to have received surgical decompression [19].

Treatment: open surgical

The open surgical therapy of MALS has been reported in case reports [6, 8, 9, 11–13, 16] and series of up to 51 patients [1, 4, 17–23, 25–28, 33].

The largest group of 51 patients with MALS was published by Reilly et al. in 1985 [33]. Operative treatment consisted of celiac trunk decompression only in 16 patients, decompression with dilatation in 17 patients, and decompression and reconstruction by primary reanastomosis or interposition grafting in 18 patients. During follow-up in 44 patients after a mean of 9.0 years (range 1–18 years), eight of 15 patients (53%) treated by celiac compression alone remained asymptomatic in contrast to 22 of 29 patients (76%) treated by celiac decompression plus some form of celiac revascularisation. In the authors' view, these findings suggested that persistent clinical improvement in patients with symptomatic celiac artery compression can be achieved by an operative technique that ensures celiac artery patency besides decompression solely [33]. A series of 15 patients with MALS and division of the median accurate ligament alone was reported by Plate in 1981. After follow-up from 1 1/2 to 9 1/2 years, only two patients were found to be asymptomatic regarding upper abdominal pain [24]. Another series of 11 patients with MALS and decompression of the celiac trunk only was published by Geelkerken et al. [18]. All patients were free of symptoms immediately after surgery, but 3 months later, three out of 11 patients had recurrent abdominal pain. During follow-up, 15 to 23 years later, of eight patients, all had symptoms similar to those before surgery. Regarding these unsatisfactory results, the authors emphasised that operation should not be undertaken in patients with vague upper abdominal complaints and compression of the celiac artery who do not have pathological conditions which might cause their abdominal symptoms [18]. Takach et al. [25] reported on seven patients with MALS. All seven had decompression of the celiac trunk. In addition, three patients had an aorto-celiac bypass (two polytetrafluoroethylene, one vein) while one patient had an angioplasty with a polyester fibre patch and one patient had an aortic reimplantation of the celiac artery. All patients experienced immediate and long-term relief of the preoperative symptoms [25]. Kokotsakis et al. [13] described the case of a 65-year-old woman having MALS and the successful treatment with an aorto-celiac vein bypass. Two and a half years after surgery, the patient was still free of symptoms [13]. Foertsch et al. reported of the successful open surgical treatment of three adolescent patients between 15 and 16 years with MALS [1].

In our series, 18 patients were treated with open surgery. Due to the local and specific pathology of the celiac trunk with fixed stricture or stenosis, primary, seven and secondary, four patients out of 18 cases had 11 further procedures on the celiac artery besides decompression performed (aorto-celiac vein interposition $n=6$, aorto-hepatic vein interposition $n=1$, resection of the celiac artery and end-to-end anastomosis $n=2$, patchplasty of the celiac artery with vein $n=1$, and transaortic removal of a stent of the celiac artery $n=1$). Out of the 11 patients with a successful outcome during follow-up, six of them had decompression of the celiac trunk only. The other five patients had vascular co-procedures on the celiac trunk.

Treatment: laparoscopic surgical

Roayaie et al. reported on a 43-year-old woman with MALS who was treated by laparoscopic division of the median arcuate ligament and resection of the overlying neural and lymphatic tissue. Intraoperatively, a laparoscopic ultrasound scanning probe documented an improved flow in the celiac artery [15]. Dordoni et al. published the

laparoscopic treatment of a patient with MALS. In this case, the ligament and the adherent tissue were dissected as well [10]. Carbonell et al. described the laparoscopic release of the median arcuate ligament in a 22-year-old male patient. The postoperative CT angiogram showed no residual stenosis in the celiac trunk [7]. The follow-up of these patients after 3, 6, and 7 months, respectively, was uneventful [7, 10, 15]. The authors accented the advantages of laparoscopic procedures such as less blood loss (intra-operative 55 ml [15] and 65 ml [10]) and early discharge of the patient (15 h after surgery [15]).

In our series, besides decompression, the need to reconstruct the blood flow in the celiac trunk was about 50%. This seemed partly to be the condition for an uneventful postoperative course and long-time success. The laparoscopic approach in those patients enables not advanced vascular procedures on the celiac trunk. This might yet abolish the advantages of the laparoscopic management in a number of cases, but allows secondary procedures in case of recurrent or unsolved stricture.

Treatment: interventional/endovascular

Cinà and Safar reported about a 62-year-old woman with postprandial abdominal pain and weight loss. The angiography revealed compression of the celiac axis, and angioplasty was successful. Afterwards, the patient underwent surgical division of the median arcuate ligament with complete relief of symptoms, which recurred 4 months later. Due to a restenosis of the celiac axis, an aorto-celiac artery bypass was performed, and a persistent relief of symptoms in a 4-year follow-up was documented [8]. Delis et al. published the case of a 21-year-old woman with MALS who was treated three times with percutaneous transluminal angioplasty (PTA) and insertions of stents over 8 months. Due to recurrence, a transection of the damaged stented segment of the celiac artery was performed, the median arcuate ligament and adjacent ganglionic tissue strangulating the celiac artery and the aorta were divided, and a reconstruction with a side-to-end aorto-celiac 8-mm Dacron prosthesis and patch angioplasty of the hepatic artery was added. The 28-month follow-up showed a patient free of abdominal symptoms [9]. In their review, Gloviczki and Duncan predicted that stenting combined with laparoscopic decompression of the celiac artery would be used with increasing frequency [42].

Conclusion

The MALS is a rare vascular disorder caused by an extrinsic compression of the celiac artery and induces upper abdominal pain, mostly, postprandial. The diagnosis of MALS can definitively be established by lateral aortography of the visceral aorta and its branches in inspiration and expiration. The open surgical therapy is a safe and reliable procedure with no mortality and low morbidity. Regarding the local and specific pathology of the celiac trunk after decompression with fixed stricture or stenosis, further vascular procedures are needed. The long-time follow-up is satisfactory. The laparoscopic approach reduces the procedure on decompression only that might not be sufficient in most cases. Endovascular treatment with PTA and insertion of a stent does not solve the underlying problem of extrinsic compression of the celiac trunk and often requires further open procedures. Due to the low incidence of MALS, no guidelines will be able to apprehend all patients sufficiently: the choice of treatment must account for the specific clinical and morphological situation of each patient.

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