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Simultaneous treatment: Liver transplantation and median arcuate ligament syndrome

Tratamiento simultaneo: trasplante hepático y síndrome de ligamento arcuato mediano

Keywords: Complications; Orthotopic liver transplant; Median arcuate ligament

Palabras clave: Complicaciones; Trasplante hepático; Ligamento arcuato mediano

Median arcuate ligament (MALS) syndrome involves extrinsic compression of the celiac artery by the fibrous bands of the median arcuate ligament (MAL).[1] It was first described in 1917 by Lipshutz et al.,[1,2,5] but it was not until 1963 when Harjola et al. performed the first surgeries to treat MALS.[1,3] In 1972, Colapinto et al. used CT for the first time for the diagnosis of MALS, and since then it has become the best diagnostic method.[4]

MALS is usually asymptomatic because collateral circulation develops, but in 10%–25% of cases some type of functional ischemia may occur.[1] Symptomatic patients may present abdominal pain with no obvious cause, which may be postprandial or triggered by exercise, as well as weight loss, nausea and vomiting.[1,5]

MALS that is not correctly evaluated or treated can cause serious complications after supramesocolic surgeries, especially pancreaticoduodenectomy.[7] However, the number of publications on patients with MALS undergoing liver transplantation (LT) is very limited, and no internationally accepted recommendations have been published about the best therapeutic strategy to follow.[1] It has been suggested that MALS can cause postoperative dysfunction of the liver graft by decreasing the mean flow velocity of the hepatic artery, thereby hindering flow to the graft, which can lead to hepatic artery thrombosis and biliary complications.[5] We present our experience in the synchronous treatment of MALS and LT.

Using a prospective database, we have conducted a retrospective observational study of all patients who had undergone LT from September 2012 to December 2021. Patients with preoperative CT scan diagnosis of MALS were selected (Fig. 1). We performed a temporary portacaval shunt and divided the MAL after completing total hepatectomy.

Within 24 h of LT, Doppler ultrasound was used to confirm the permeability of the arterial flow. The following clinical variables were studied: preoperative (age, sex, etiology of liver disease, MELD, preoperative hemoglobin, and donor risk index [DRI]); intraoperative (cold ischemia time, blood loss, arterial and portal flow pre/post-transplantation, and operative time); and postoperative (complications defined by Clavien-Dindo classification,[8] especially vascular, hospital stay, graft and patient survival).

We have operated on 4 patients with MALS, representing an incidence in our series of 1.01% (4/394). There were no complications associated with the division of the MAL. All grafts presented good perfusion after performing the venous and arterial anastomoses, and no post-transplantation vascular or biliary complications were observed. Vascular grafts were not used in any of these patients. Patient data are

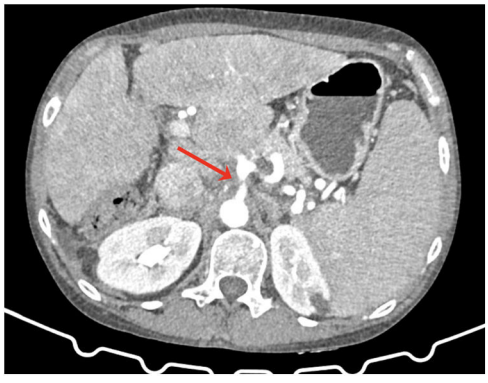

Figure 1 – CT (red arrow) of stenosis of the celiac trunk due to MALS.

Table 1A – Characteristics of the patients treated.

Data of patients with MALS and LT

Patient	Age	Cause for liver transplant	MELD	Pre-transplantation symptoms	CT scan findings	Pre-op Hb (g/dL)	DRI	Blood loss (mL)	Cold ischemia time (min)	Hepatic artery flow (mL/min)	Portal Flow (mL/min)	Immediate vascular complications	Months after LT	Complications	Resolution of complications	Clavien-Dindo	Hospital stay	IS	Mortality
1	43	Alcohol	19	–	Compression of the CA by MAL	9.1	1.26	600	259	185	2920	No	106	Melenas	Endoscopic treatment	IIIA	6	Corticosteroids Tacrolimus MMF	No
2	55	Alcohol	11	Abdominal pain	Significant stenosis of the segment proximal to the CA Aneurisms in the gastroduodenal artery and SMA	11.7	1.28	300	284	114	2479	No	107	Deteriorated renal function/kidney failure Hyperglycemia	Insulin	II	5	Corticosteroids Tacrolimus MMF	No
3	45	Alcohol	16	Occasional vomiting Pyrosis	Dilatation of common hepatic artery	11.6	2.17	620	278	122	1742	No	108	–	–	0	2	Corticosteroids Tacrolimus MMF Anti CD25	No
4	46	Alcohol	15	–	Compression of the CA without notable collateral circulation; mild stenosis of SMA	9.4	1.96	1500	219	91	1316	No	107	Kidney failure Oliguria Ascites Pleural effusion	Prism Drainage	IVA	34	Corticosteroids Tacrolimus MMF Anti CD25	No

CT: computed tomography. CA: celiac artery. MAL: median arcuate ligament. SMA: superior mesenteric artery. Hb: hemoglobin. DRI: donor risk index. cc: cubic centimeters. LT: liver transplantation.

Table 1B – Published series.

First author	Study type	Patients MALS(n)/TH(n) %	Intraoperative variables studied	Treatments	Conclusions
Fukuzawa et al. 1993	Retrospective cohorts	5/307 (1.6%)	Hepatic artery flow	• MAL division 3/5(60%) • Aorto-hepatic reconstruction 2/5(40%)	CA compression reduces hepatic artery flow in the implant.
Jurim et al. 1993	Cohorts retrospective	17/164 (10%)	Hepatic artery flow	• MAL division 17/17 (100%) • Aorto-hepatic division after unsuccessful MAL division 2/17(12%)	The identification of MALS and the division of the MAL are important to prevent potential complications after LT.
Agnes et al. 2001	Cohorts retrospective	5/140(3.6%)	Blood pressure Radial pressure Differential pressure Clinical variation of pressures in respiratory phases.	• MAL division 5/5 (100%)	Compression of the CA by the MAL should be treated with its division to avoid acute and chronic complications.
Lubrano et al. 2008	Retrospective cohorts	10/168 (6%)	Not available	• Aorto-hepatic reconstruction 4/10 (40%) • MAL division 1/10 (10%)	The presence of MAL does not hinder classic arterial reconstruction. MAL can be divided if necessary.
Czigany et al. 2019	Retrospective cohorts	34/286 (12%)	Cold ischemia time Warm ischemia time Blood transfusion	• MAL division 26/34 (77%) • Reconstruction 4/34 (12%)	MAL division is safe and feasible and can be considered in patients with relevant stenosis of the celiac artery observed on preoperative CT scan.
Gialamas E et al. 2022	Retrospective case/control	19/57 (33.3%)	Cold ischemia time Warm ischemia time Blood transfusions Anastomosis type	• Gastroduodenal artery preservation8/19 (42.1%) • MAL division 3/19 (15.8%) • Aorto-hepatic reconstruction 1/19 (5.3%) • No treatment 7/19(36.8%) • Controls: no treatment	No differences were found in the prevalence of biliary and arterial complications between the presence of MAL or no MAL in LT patients.
Shu -Xuan Li et al. 2022	Retrospective cohorts	8/288 (2.7%)	Cold ischemia time Warm ischemia time Blood loss Blood transfusion Plasma transfusion Operative time Hepatic artery flow	• No intervention 2/8 (25%) • Gastroduodenal preservation with splenic artery ligation 3/8 (37.5%) • Gastroduodenal artery preservation2/8 (25%) • Reoperation with MAL division 1/8 (12.5%)	The conservation of collateral circulation between the superior mesenteric artery and the celiac artery through the gastroduodenal artery with or without ligation of the splenic artery is a safe and feasible method for MAL division.
Viñas et al. 2023	Retrospective cohorts	4/394 (1.01%)	Operative time Cold ischemia time Warm ischemia time Hepatic artery flow Portal flow	• MAL division 4/4 (100%)	No morbidity associated with MAL division No vascular complications after LT

summarized in Table 1A. The published series on LT and MALS that we reviewed are presented in Table 1B.

MALS is an uncommon condition in the general population. According to published series,[5] the reported incidence in post-LT patients ranges from 1.6%–33%, although our series had a slightly lower incidence (1%). The symptoms of MALS are non-specific; 2 of our patients presented abdominal pain, which was accompanied by vomiting in one case. The usual symptoms of MALS can be attributed to chronic liver disease.[10] Therefore, its diagnosis is difficult and requires thorough radiological evaluation of the pre-transplantation CT scan, thus allowing the surgeon to establish an appropriate intraoperative surgical strategy.[1,5,9,10]

To date, there are no established treatment recommendations for MALS in patients undergoing LT.[1,5] There is a classification of MALS in patients undergoing pancreaticoduodenectomy, and the treatment is adapted according to the percentage and length of the stenosis.[10] However, it has not been determined whether this algorithm is applicable in LT. Surgical treatment is generally accepted as being required in symptomatic patients, but there is no consensus on treatment in cases with no obvious symptoms.[1,5,9,10] Advocates of

treating any MALS in patients undergoing LT argue that, if left untreated, the risk of post-LT vascular and biliary complications may increase, while treatment does not increase morbidity.[1] Nevertheless, the few published series provide no data confirming a higher incidence of vascular and biliary complications after LT if there is asymptomatic MALS that is not treated.[1,6]

When treating MALS in patients undergoing LT, therapeutic options include: division of the MAL, preservation of the gastroduodenal artery, or more complex techniques, such as aorto-hepatic bypass or endovascular techniques.[5,6,9,10] In our series, we performed division of the MAL in all cases, and no morbidity was observed associated with this technique. One of the possible complications indicated associated with the division of the MAL is that hemorrhage occurs due to the existence of varices and abundant collateral circulation in the area. We believe that the portacaval shunt that we perform in all of our patients minimizes this risk, facilitating division of the MAL.

In conclusion, MALS in patients undergoing LT is rare, usually asymptomatic, and requires a thorough pre-transplantation CT study. There are no therapeutic algorithms with high scientific evidence, but since its treatment does not increase morbidity, we believe that MAL division is advisable to avoid possible postoperative vascular and/or biliary complications.

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