Acute liver failure and experience with therapy using the molecular absorbent recirculation system (MARS)

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Introduction and Objective: Acute liver failure (ALF) is a cause of urgent liver transplantation (LT). The molecular absorbent recirculation system (MARS) is an extracorporeal liver replacement device, considered bridging therapy for LT.

Case report: 24-year-old man with no relevant history was admitted due to asthenia, adynamic, hyporexia, jaundice, oral intolerance and transaminasemia. His laboratory studies are shown in Table 1 and include positivity for IgM hepatitis A (HAV). Other causes are excluded. Development of ALF with two minor criteria (King's College), received N-acetylcysteine, without response, management with MARS/PRISMA is started, one session (initial dialysate 1300 mL, increasing to 1800 mL, maintaining a flow of 150 mL/L, and eight bottles of 25% albumin (400 mL)). His evolution towards neurological, hepatic, and renal improvement. Discharged for improvement.

Discussion: MARS therapy is based on removing molecules, including medium-sized ones, especially those that are binding by albumin and, therefore, cannot be purified conventionally. The relative simplicity, the good tolerance and the results obtained so far make MARS the most promising alternative. There is some experience with the use of MARS in ALF due to HAV.

Conclusion: MARS therapy is useful in the management of patients with ALF due to HAV; its use has shown positive results impacting patient survival and even, in some cases, avoids liver transplantation. The number of sessions will depend on the clinical response.

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Table 1 Laboratory values before and after the use of MARS therapy.

	Before	After
Hemoglobin (g/L)	17.90	13
Leukocytes (cell/10^3)	10.4	6.10
Platelets (cell/10^3)	213	207
Glucose (mg/dL)	86	71
Creatinine (mg/dL)	3.2	1.93
Total bilirrubin (mg/dL)	16.1	10
Direct bilirrubin (mg/dL)	14	5.8
ALT (U/L)	2409	824
AST (U/L)	772	257
ALP (U/L)	110.5	77
GGT (U/L)	501	136
ALB (g/dL)	3.90	2.8
INR	3.4	1.1
PT (Sec)	36.9	21.8
aPPT (Sec)	45	31
	Hepatitis A virus IgM	

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Congenital hepatic fibrosis as a rare cause of non-cirrhotic portal hypertension

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Introduction and Objective: a case of congenital hepatic fibrosis is presented

Case report: A 19-year-old female with a medical story relevant only to epistaxis of 3 years long was referred to the liver clinic because of thrombocytopenia and transaminasemia. She denied data of decompensated advanced chronic liver disease and hepatic transition elastography was performed: S0 224, F4 (19.9 kPa). Portal Doppler ultrasound was performed: a diffuse liver disease with hepatomegaly, signs of portal hypertension, splenomegaly and ascites, and probable thrombosis of the distal splenic vein. Given the suspicion of hereditary thrombophilia, a genetic profile was requested (negative Leiden Factor V PCR, negative JAK 2 PCR, negative lupus anticoagulant, normal antithrombin III, normal protein C, normal protein S). Abdominal-pelvic angiotomography was performed: enlarged liver with no focal lesions, no dilatation of the bile duct, adequate permeability of the portal venous system, and enlarged spleen. The rest of the antibodies and tests for congenital metabolic disorders were requested (normal ANAS, normal ASMAs, normal Anti LKM1, normal AMA, normal IgG, normal ceruloplasmin, normal urine copper, low ferritin, normal transferrin). Active infection by hepatitis B, C and HIV viruses was ruled out. During follow-up, the patient developed variceal gastrointestinal bleeding, endoscopic variceal ligation was performed and management with a non-selective beta-blocker was initiated. A percutaneous liver biopsy was performed, reporting in histopathology: morphological changes consistent with malformation of the ductal plate of a congenital hepatic fibrosis type.

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Sclerosing cholangitis associated with IgG4 disease. Case Report

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Introduction and Objectives: This study aimed to present the case of a patient with sclerosing cholangitis and autoimmune pancreatitis associated with IgG4 disease.

Materials and Methods: A 44-year-old male with a history of allergic rhinitis, diabetes, and dyslipidemia. His clinical picture began with diffuse abdominal pain and jaundice; on physical examination, jaundice, a painful abdomen in the right upper quadrant and hepatomegaly. Liver biochemistry with transaminasemia and cholestasis. In auxiliary studies US with bile duct dilatation of 12.4 mm, ColangioMR was performed, reporting concentric wall thickening in T2 of the intra and extrahepatic bile duct that condition focal stenoses of the right hepatic duct and intrahepatic bile duct approximately 16 mm from the ampulla, conditioning segmental dilatations of the common hepatic and extrapancreatic bile duct up to 8.9mm. According to these findings, IgG4 serum levels were requested, reporting 1200mg/dl. Steroid treatment was started, presenting a favorable response.

Discussion: IG-G4-related disease is an autoimmune relapsing chronic multiorgan fibroinflammatory syndrome. Its maximum incidence is in Japan, with 336 to 1300 patients diagnosed/year; the estimated prevalence is 62/million subjects between 50 and 70 years of age. The main and most commonly affected organs are the pancreas, bile duct, salivary and lacrimal glands, retroperitoneum and lymph nodes.

Conclusions: The relevance as Gastroenterologists recognize the disease associated with IgG4 because of the multi-organ involvement as part of the approach to a patient with jaundice syndrome; despite the low prevalence reported in our country, knowing this entity will make its timely treatment and subsequent recognition easier in other patients.

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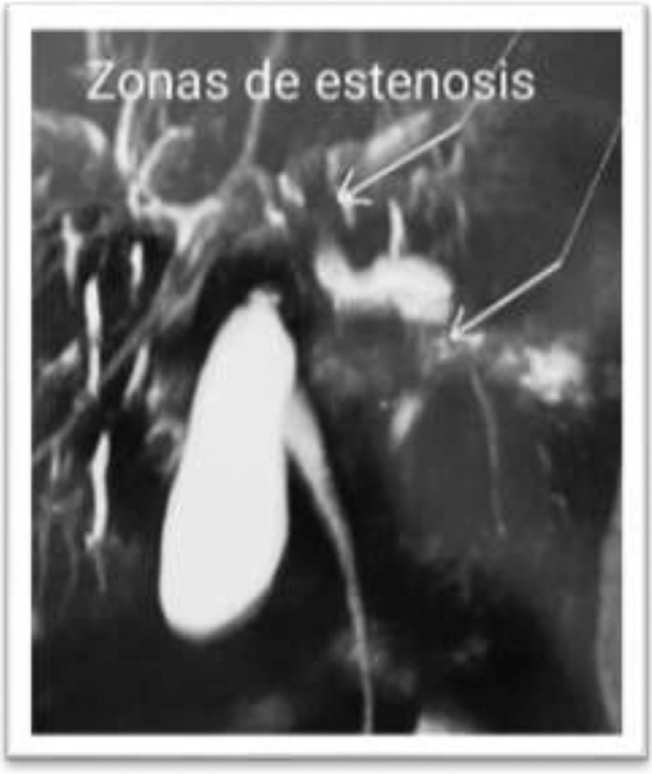

Figure 1.
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Giant simple hepatic cyst, when and how to treat it

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Introduction and Objective: Hepatic cysts are rare, with a prevalence between 0.1 to 0.5%. They are divided into parasitic and non-parasitic, being more common than the last ones. They can be subdivided into simple (<5% of the population) or multiple.

Case Report: 46-year-old woman with no relevant history. She comes due to an increase in abdominal perimeter of 4 months of evolution and weight loss of 9kg in 4 months; asthenia, adynamia, early satiety, postprandial fullness and abdominal pain in the right upper quadrant, oppressive, intensity 9/10, exacerbated by mobilization. CT scan with a giant liver cyst of 219 × 166 × 239mm, a volume of 4544cc. Alkaline phosphatase and GGT >3 times their normal value. She was admitted for percutaneous drainage placement, with a total output of 7480cc and biochemical and clinical improvement, without complications.

Discussion: Simple cysts occur in people over 40 years of age, more frequently in women (4:1 ratio). The differential diagnosis includes liver abscess, tumor, hemangioma, hematoma, parasitic cyst, and polycystosis. They are easy to distinguish by image as they are well-defined; they contain serous fluid and lack septa, papillary projections, and calcifications. They are considered giants when measuring>5 cm and their treatment is only indicated in symptomatic patients, with pain being the usual. Percutaneous drainage has little morbidity and improves compression symptoms. However, recurrence is high (almost 100%), so the administration of a sclerosing agent is recommended.

Conclusions: Conservative procedures have high recurrence rates, so systematized laparoscopic surgery is a good option for definitive treatment.

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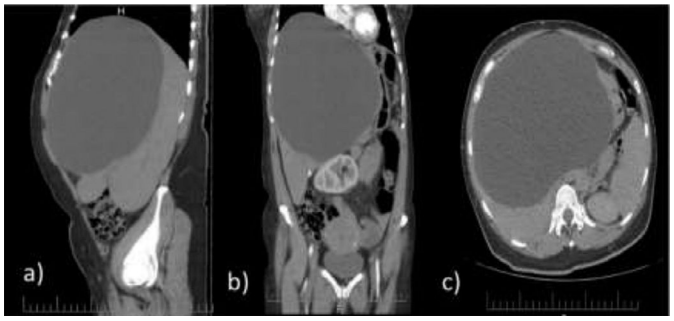

Figure 1.
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Symptomatic giant cavernous hemangioma as an indication for liver transplantation

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Introduction and Objective: Hepatic hemangiomas (HH) are the most common primary benign tumors of the liver, more frequent in women, attributed to estrogens. Its size can reach up to 30 cm, being considered giant when it measures >4 cm.

Case Report: A 65-year-old woman with giant cavernous hemangiomas as an incidental finding on liver USG. The abdominal MRI established the diagnosis by spotting two intrahepatic lesions, one of the right lobe occupying all the segments, measuring 18.5 × 16.6 × 15.7 cm, with a volume of 2527.6 cc; another in the left lobe of 7.8 × 7.8 × 6.5 cm, the volume of 206.8 cc; hypointense on T1 sequence, hyperintense on T2, with enhanced contrast medium in the periphery, later it is centripetal, with focal areas without enhancement at 20 minutes.

Physical examination: painful swelling in the left hypochondrium up to the anterior axillary line and epigastrium. Increased alkaline phosphatase and GGT. Surgical management is contraindicated due to the characteristics of the lesion. We decided to send her for a liver transplant.

Discussion: Histologically, they are vascular malformations characterized by caverns covered by a single layer of endothelium. The gold standard is MRI, where we observe peripheral nodular enhancement followed by central enhancement in a well-defined homogeneous mass. Surgical management is indicated in the symptomatic giant HH, with an increase in size or suspicion of malignancy. They