A "blood theft" after liver transplantation: the role of interventional radiology in the management and treatment of splenic artery steal syndrome

Federica Riva[1*], Enrico M Garanzini[2], Tommaso Cascella[2], Alfonso Marchianò[2], Carlo Spreafico[2]

1. Post-graduate School in Radiodiagnostics, University of Milano, Milan, Italy

2. Department of Radiology and Radiotherapy, National Cancer Institute of Milan, Italy

* **Correspondence:** Federica Riva, Department of Radiology, Sant'Anna Hospital, Via Ravona, 22042 San Fermo della Battaglia CO, Italy (*federica.riva@asst-lariana.it*)

Radiology Case. 2022 Aug; 16(8):1-7 :: DOI: 10.3941/jrcr.v16i8.4391

ABSTRACT

Splenic artery steal syndrome is a rare complication after liver transplant. It could lead to rapidly evolving major issues such as ischemic cholangiopathy and acute graft failure. Although the pathophysiology is not yet well understood, if diagnosed in time it could be easily managed with interventional radiology treatments. We present a case of a 47-year-old man presented to our institute with radiological findings typical for multifocal hepatocellular carcinoma in a cirrhotic liver. After therapy he underwent transplant. Some days after surgery he developed signs of acute liver failure. Steal syndrome was suspected by laboratory tests and radiology exams. The syndrome was confirmed by angiography and treated. The graft was saved, and the patient is still alive and free of disease. The purpose of this paper is to explain the importance of radiology exams in the diagnosis of splenic steal syndrome, explain its pathogenesis and describe the interventional management of this complication.

CASE REPORT

CASE REPORT

A 47-year-old man presented to our institute in July 2016 with radiologic findings of multifocal hepatocellular carcinoma (HCC) in a cirrhotic macronodular liver. He was diagnosed with chronic hepatitis C 26 years prior initial presentation. He was an active smoker, with insulin-dependent diabetes, with a history of intravenous drug and alcohol abuse. He was classified with the Barcelona Clinic Liver Cancer (BCLC) staging system as BCLC B and Child-Pugh (CP) A6 and was treated with sorafenib according to the European Association for the Study of the Liver Clinical Practice Guideline for hepatocellular carcinoma [1]. After 6 months of therapy, radiology exams showed complete response according to Modified Response Evaluation Criteria in Solid Tumors. Patient disease classification changed to BCLC A and CP B8 so he became eligible for orthotopic liver transplantation (OLT).

On the fifth day after transplant, the patient suddenly developed signs of acute liver failure. Laboratory analysis demonstrated a sudden spike in transaminases (99 U/L to 142 U/L; normal range < 41 U/L), gamma-glutamyltransferase (35 U/L to 698 U/L; normal range 3-6 U/L) and bilirubin (1.8 mg/dL to 8.9 mg/dL; normal range 0.15-1.10 mg/dL).

Color-Doppler Ultrasound (CD-US) revealed a reduced main hepatic artery resistance index (RI: 0.47) with a low peak systolic velocity (PSV:31.8 cm/s) and a flattened peak waveform. Portal vein RI was 0.34 with an elevated PSV of 86 cm/s (Figure 1A, 1B).

The most likely diagnoses with these data were hepatic artery thrombosis or stenosis. Acute graft rejection was less likely given the lack of appropriate clinical symptoms. A contrast-enhanced computer tomography (CT) scan ruled out both hepatic artery stenosis and thrombosis (Figure 2).

In the end, a celiac angiography confirmed the absence of stenosis and showed a preferential flow into the splenic artery with a scarce and slow filling of hepatic artery branches suggesting the diagnosis of splenic artery steal syndrome (SASS). (Figure 3).

Treatment options were discussed in a multidisciplinary conference with surgeons and interventional radiologists. The two main options of treatment were splenic artery surgical ligation or splenic artery embolization. We proceeded with the latter, the splenic artery was engaged with a 7 Fr introducer (Flexor Tuohy-Borst Side-Arm, Cook, USA) and one vascular trilobar plug of nitinol (Amplatzer Vascular Plug type II, Abbott, USA) of 10 mm was placed proximally. The procedure was safe and effective resulting in the complete occlusion of the main splenic artery resulting in the diversion of blood flow to the hepatic artery. Hepatic artery flow returned to normal rates (Figure 4A, 4B).

The efficacy of the treatment was confirmed by post-surgical CD-US that showed improvement of hepatic artery PSV (from 31.8 cm/s to 72.1 cm/s) with a regularly pointed apex waveform, of portal vein PSV (from 86 cm/s to 54.4 cm/s) and portal vein RI (from 0.34 to 0.18) (Figure 5A, 5B).

After embolization liver function tests rapidly improved and normalized. Before the discharge, the patient underwent contrast-enhanced US that showed homogeneous enhancement of the spleen without necrotic areas, confirming a preserved splenic flow allowed by the presence of splenic collateral vessels (Figure 6).

DISCUSSION

Etiology & Demographics:

SASS is an under-recognized cause of graft failure after OLT. The incidence of SASS after liver transplantation varies considerably between 0.6-10.1[2] [3] and usually is a diagnosis of exclusion after eliminating the more frequent diagnoses of thrombosis or stenosis.

Clinical & Imaging findings:

The clinical presentation of this syndrome is non-specific. Usually, in the early posttransplant period the patient can suffer from graft dysfunction with abnormal liver function tests, >80% is diagnosed within 60 days of liver transplantation[2]. Rarely a patient can develop ascites after two months from OLT as the main non-specific sign of the syndrome [2]. The SASS was first described by Manner et al. in 1991 and named by Langer et al. in 1992 [4]. The most accepted pathogenetic hypothesis, even if not completely demonstrated, is that the hepatic artery hypoperfusion is due to a relatively portal venous hyperperfusion mediated by an alteration of the physiological compensatory mechanism defined as hepatic arterial buffer response (HABR) [5]. The HABR is based on the concept that adenosine, a powerful vasodilator molecule, is secreted at a constant rate and washed out of the space of Mall into the portal vein. A decreased portal flow washes less adenosine away, which then accumulates leading to marked vasodilation of the hepatic artery. Conversely, a portal overflow will lead to hepatic artery constriction and liver hypoperfusion [6], [7].

Other authors have suggested that multiple factors might be involved in the development of this condition [2] also to explain the existence of clinical varieties of this syndrome such as some cases of preferential flow to the gastroduodenal artery [8] and left gastric artery [9]. Spleen's augmented volume could represent a risk factor for this syndrome [10].

A reduced PSV and augmented IR in CD-US of hepatic vessels could raise the suspicion of SASS diagnosis [11], [12] but these indices aren't pathognomonic because of the CD-US operator-dependent nature.

For detecting or excluding vascular complications the most sensitive exam is contrast-enhanced CT [13].

Digital subtraction angiography is the gold standard for the diagnosis of SASS because dynamically shows hepatic and splenic vessels flow alterations previously described [2], [3], even if without any measurable or quantitative data.

Differential Diagnoses:

The most common cause of post-OLT graft ischaemia and failure consists primarily of acute thrombosis, which occurs in about 10% of liver transplant recipients, and secondly in stenosis of the hepatic artery [9]. Other causes consist of medical complications such as peri-transplantation infections (account for almost 60% of deaths or graft losses in the first operative year) [14] or the development of graft-versus-host-disease (infrequent, with an incidence of 0.5–2%)[15].

Treatment & Prognosis:

Regarding treatment options in SASS, embolization with a minimally invasive method has demonstrated a lower rate of complications in the post-OLT setting than surgical splenic artery ligation. Transcatheter embolization is considered the best choice of treatment and a lower rate of local and systemic complications is reported if the embolization is performed in the proximal splenic artery rather than distally [16], [17]. The embolic agent of choice for the treatment could be different, usually coils or plugs, and the choice is guided by the material availability as well as by the experience of the interventional radiologist with that specific embolic agent.

TEACHING POINT

Splenic artery steal syndrome is a rare but important complication after liver transplant.
This entity should be known and considered when the most common causes of graft complications are already excluded. Contrast-enhanced computer tomography, Color-Doppler ultrasound and diagnostic angiography are the main imaging modalities to use to reach the diagnosis.

REFERENCES

1. Galle PR, Forner A, Llovet JM, Mazzaferro V, Piscaglia F, Raoul JL, Schirmacher P-95-e3837. pd., Vilgrain V. EASL Clinical Practice Guidelines: Management of hepatocellular carcinoma. J Hepatol. European Association for the Study of the Liver; 2018;69(1):182-236. PMID: 29628281

2. Saad WEA. Nonocclusive hepatic artery hypoperfusion syndrome (splenic steal syndrome) in liver transplant recipients. Semin Intervent Radiol. 2012 Jun;29(2):140-6. PMID: 23729985

3. Mogl MT, Nüssler NC, Presser SJ, Podrabsky P, Denecke T, Grieser C, Neuhaus P, Guckelberger O. Evolving experience with prevention and treatment of splenic artery syndrome after orthotopic liver transplantation. Transpl Int. 2010 Aug;23(8):831-41. PMID: 20180930

4. Li C, Kapoor B, Moon E, Quintini C, Wang W. Current understanding and management of splenic steal syndrome after liver transplant: A systematic review. Transplant Rev. Elsevier Inc.; 2017;31(3):188-192. PMID: 28254530

5. Lautt WW. Mechanism and role of intrinsic regulation of hepatic arterial blood flow: Hepatic arterial buffer response. Am J Physiol - Gastrointest Liver Physiol. 1985;12(5). PMID: 3904482

6. Lautt WW. Regulatory processes interacting to maintain hepatic blood flow constancy: Vascular compliance, hepatic arterial buffer response, hepatorenal reflex, liver regeneration, escape from vasoconstriction. Hepatol Res. 2007;37(11):891-903. PMID: 17854463

7. Quintini C. "Splenic Artery Steal Syndrome" Is a Misnomer: The Cause Is Portal Hyperperfusion, Not Arterial Siphon. Liver Transplant. 2008;14:374-379. PMID: 17563933

8. Nishida S, Kadono J, DeFaria W, Levi DM, Moon JI, Tzakis AG, Madariaga JR. Gastroduodenal artery steal syndrome during liver transplantation: Intraoperative diagnosis with Doppler ultrasound and management. Transpl Int. 2005;18(3):350-353. PMID: 15730497

9. Sevmis S, Boyvat F, Aytekin C, Gorur SK, Karakayali H, Moray G, Haberal M. Arterial Steal Syndrome After Orthotopic Liver Transplantation. Transplant Proc. 2006;38(10):3651-3655. PMID: 17175358

10. Grieser C, Denecke T, Steffen IG, Avgenaki M, Fröhling V, Mogl M, Schnapauff D, Lehmkuhl L, Stelter L, Streitparth F, Langrehr J, Rothe JH, Hamm B, Hänninen EL. Multidetector computed tomography for preoperative assessment of hepatic vasculature and prediction of splenic artery steal syndrome in patients with liver cirrhosis before transplantation. Eur Radiol. 2010;20(1):108-117. PMID: 19662418

11. Sanyal R, Shah SN. Role of imaging in the management of splenic artery steal syndrome. J Ultrasound Med. 2009;28(4):471-477. PMID: 19321675

12. Uslu N, Aslan H, Tore HG, Moray G, Karakayali H, Boyvat F, Arslan G, Haberal M. Doppler ultrasonography findings of splenic arterial steal syndrome after liver transplant. Exp Clin Transplant. Exp Clin Transplant; 2012 [cited 2022 Feb 6];10(4):363-367. PMID: 22757943

13. Low G, Crockett AM, Leung K, Walji AH, Patel VH, Shapiro AMJ, Lomas DJ, Coulden RA. Imaging of vascular complications and their consequences following transplantation in the abdomen. Radiographics. Radiological Society of North America; 2013 May 3 [cited 2021 Sep 5];33(3):633-52. PMID: 23674767

14. European Association for the Study of the Liver. EASL Clinical Practice Guidelines: Liver transplantation. J Hepatol. 2016 [cited 2021 Sep 5];64(2):433-485. PMID: 26597456

15. Murali AR, Chandra S, Stewart Z, Blazar BR, Farooq U, Ince MN, Dunkelberg J. Graft Versus Host Disease After Liver Transplantation in Adults: A Case series, Review of Literature, and an Approach to Management. Transplantation. 2016 Dec;100(12):2661-2670. PMID: 27495762

16. Nüssler NC, Settmacher U, Haase R, Stange B, Heise M, Neuhaus P. Diagnosis and treatment of arterial steal syndromes in liver transplant recipients. Liver Transplant. 2003;9(6):596-602. PMID: 12783401

17. Madoff DC, Denys A, Wallace MJ, Murthy R, Gupta S, Pillsbury EP, Ahrar K, Bessoud B, Hicks ME. Splenic arterial interventions: Anatomy, indications, technical considerations, and potential complications. Radiographics. 2005;25(SPEC. ISS.):191-212. PMID: 16227491

FIGURES

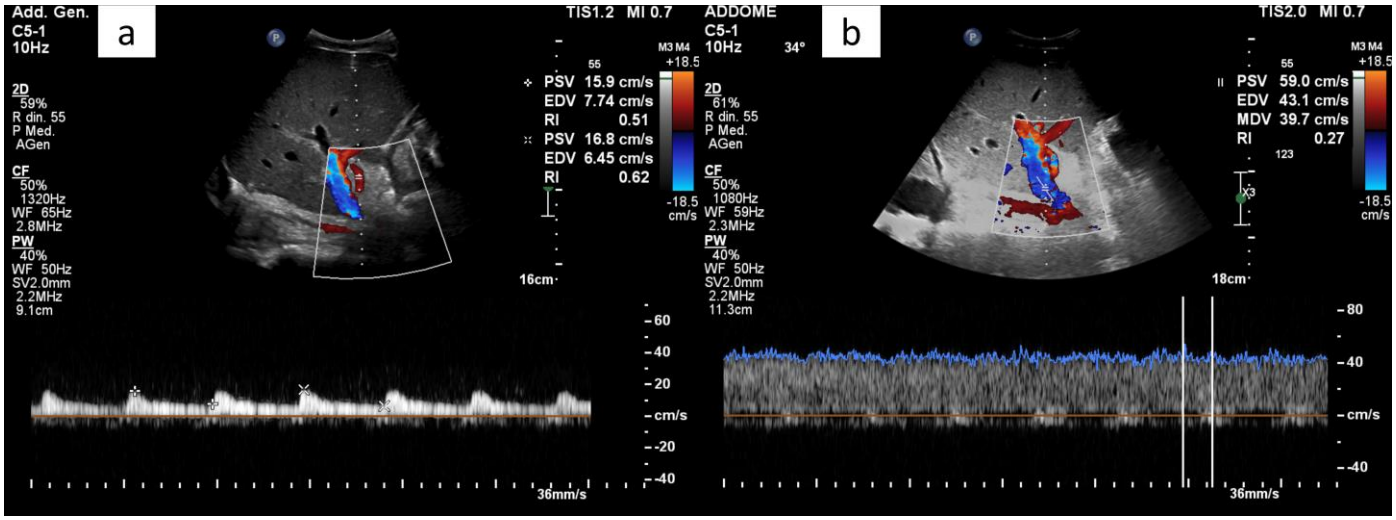

Figure 1: 47 years old male with acute liver graft failure.

FINDINGS: Doppler US obtained after 5 days from transplant showing alteration of PS velocity and RI respectively in the hepatic artery (a) and the portal vein (b); hepatic artery velocity is very low.

TECHNIQUE: Doppler ultrasound imaging

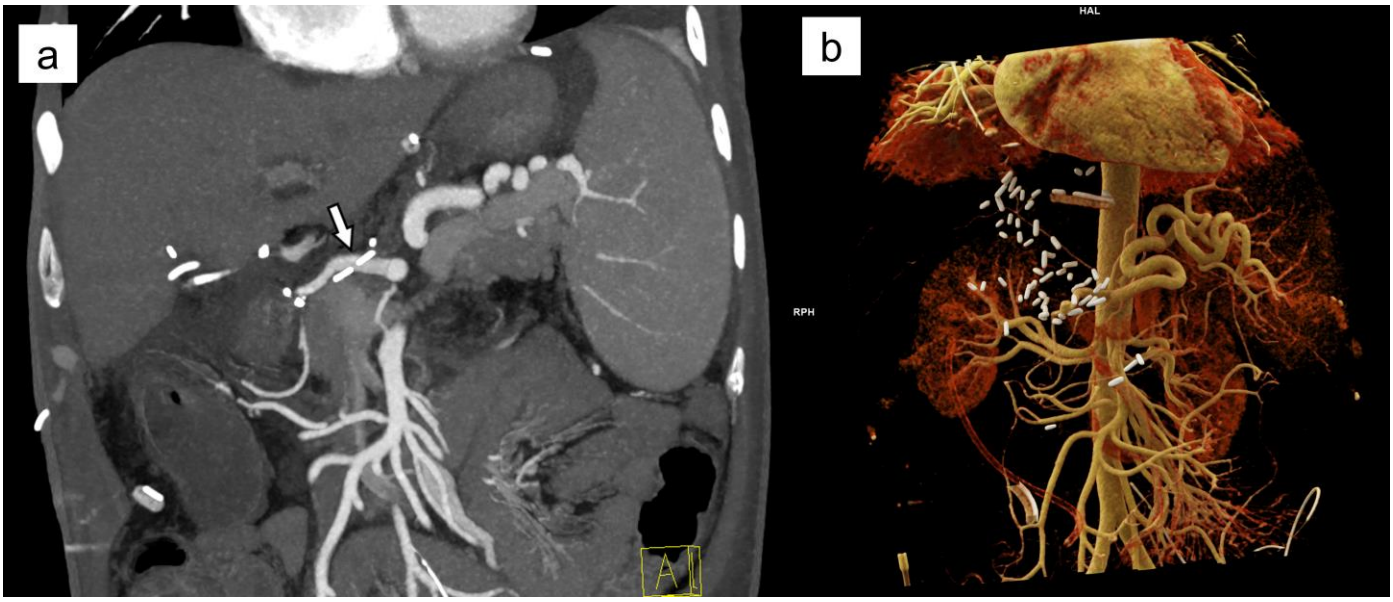

Figure 2: 47 years old male with acute liver graft failure.

FINDINGS: Contrast-enhanced coronal CT scan reconstruction (a) documented patency of the hepatic artery (arrow). Reformat 3D (b).

TECHNIQUE: Axial CT, 190 mAs, 120 kV, 1mm slice thickness, 90 ml Iopamiro 370.

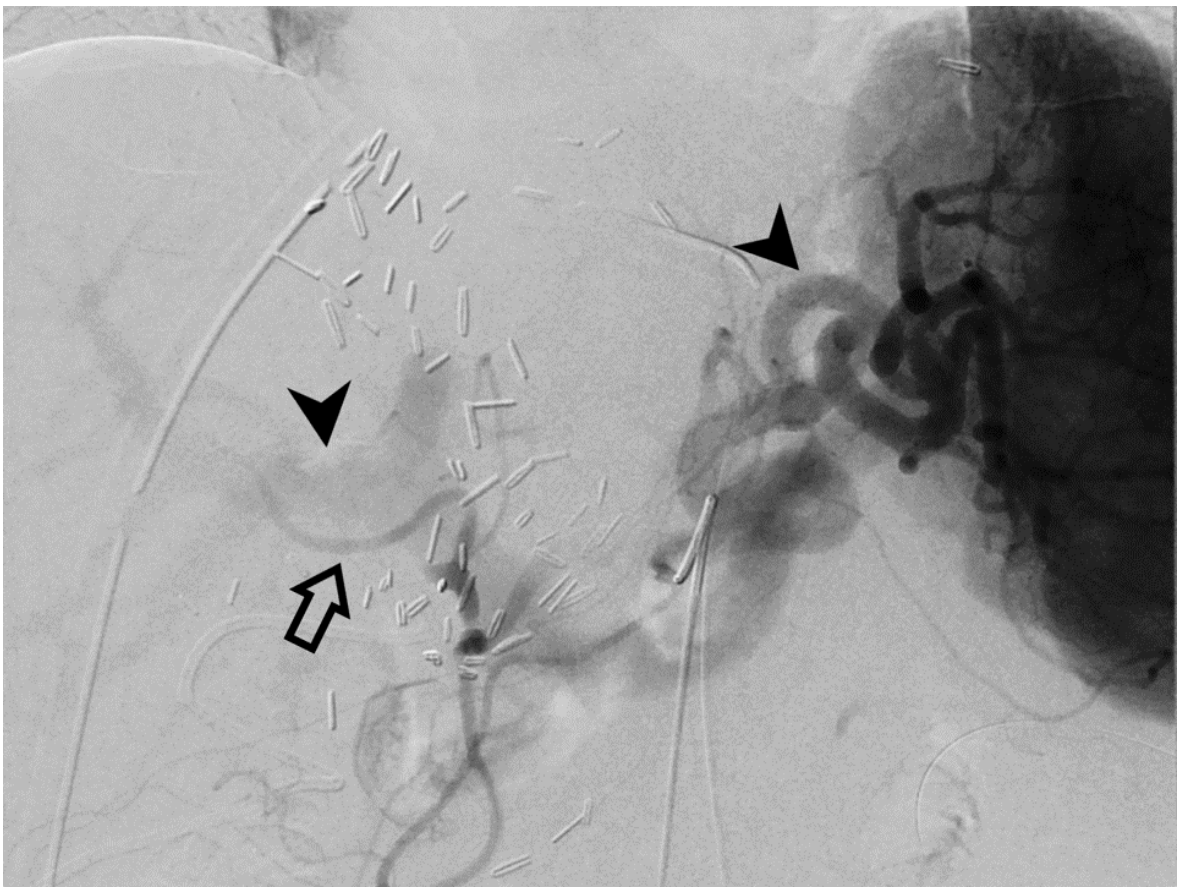

Figure 3: 47 years old male with acute liver graft failure.

FINDINGS: Pre-treatment digital subtraction angiography of celiac trunk which confirmed an altered flow distribution. Decreased flow in the hepatic artery (arrow). Portal early and splenic artery increased flow (arrowheads).

TECHNIQUE: anteroposterior angiography with 30ml of Iopamiro 370.

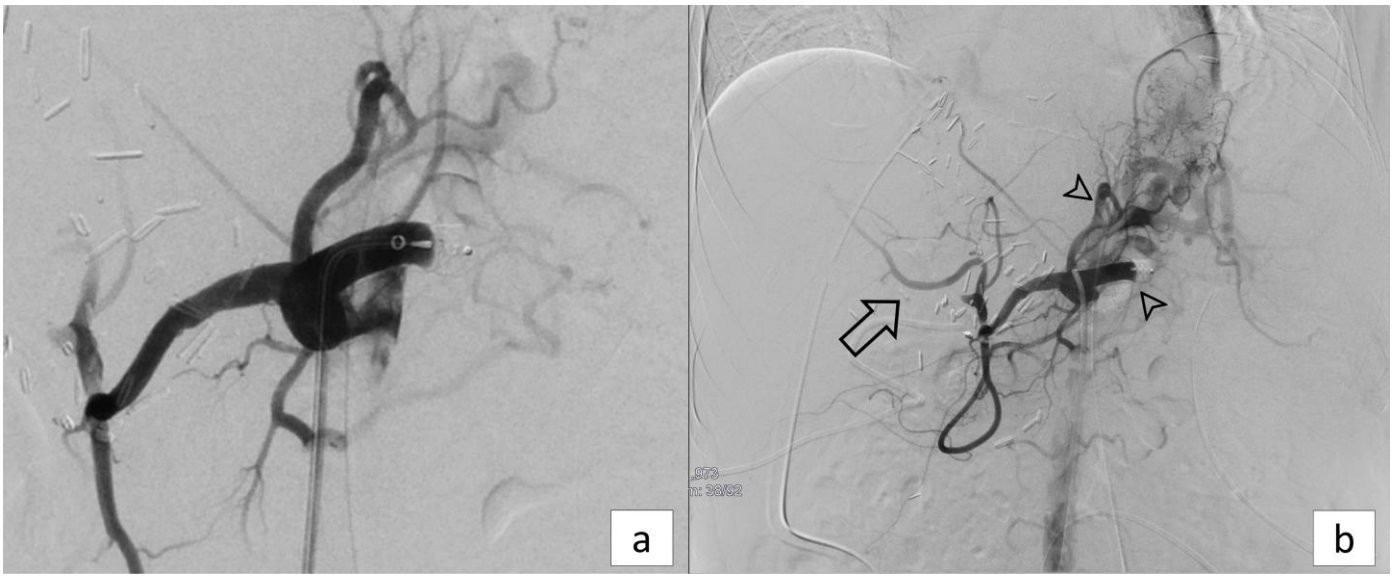

Figure 4: 47 years old male with acute liver graft failure.

FINDINGS: Digital subtraction angiography immediately after plug placement (a). Splenic collateral vessels and plug in the proximal splenic artery (arrowheads); hepatic artery flow restored (arrow). Detail of the positioning of the endovascular plug in the proximal splenic artery (b).

TECHNIQUE: anteroposterior angiography with 30ml of Iopamiro 370.

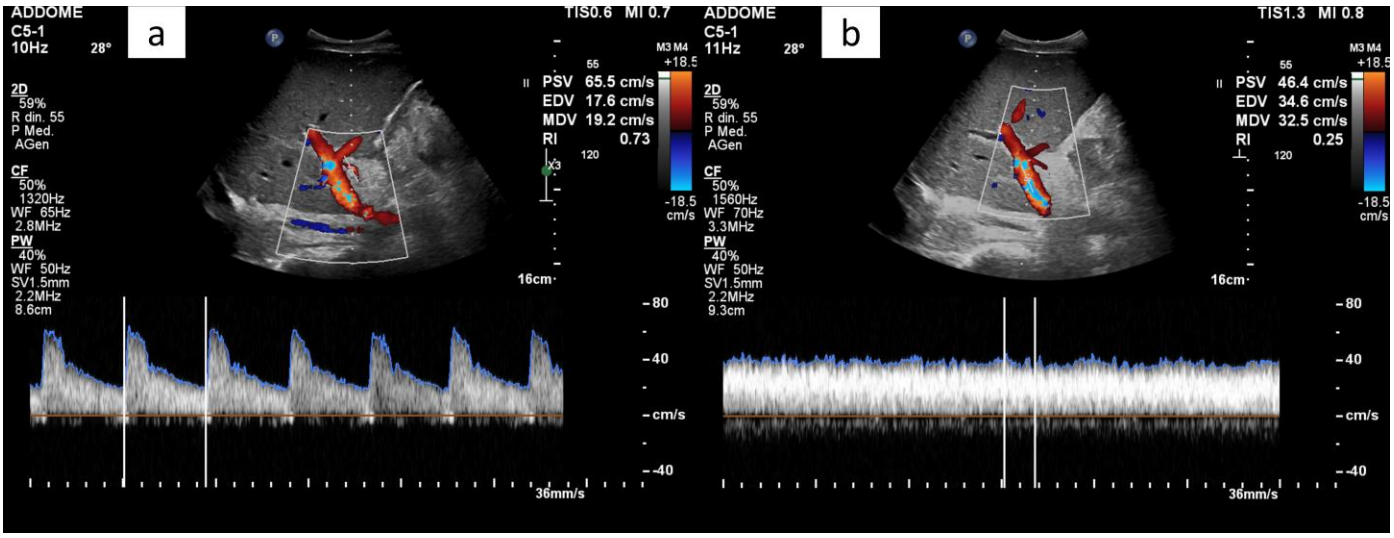

Figure 5: 47 years old male with acute liver graft failure.

FINDINGS: doppler US at 24 hours post-procedure showed an improvement of velocity and RI in the hepatic artery (a) and reduction of portal vein velocity (b).

TECHNIQUE: Doppler ultrasound imaging

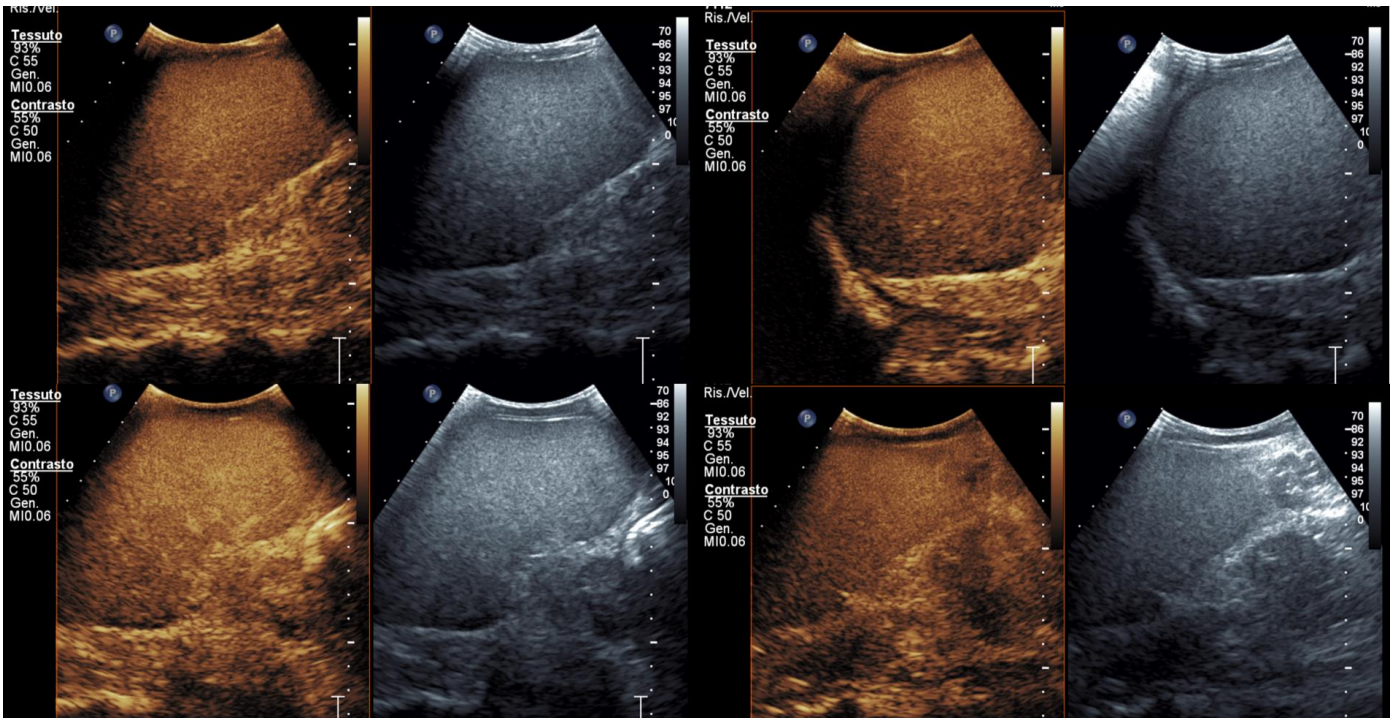

Figure 6: 47 years old male with acute liver graft failure.

FINDINGS: contrast-enhanced US scan that excludes emerging hypoechogenic ischemic areas in the spleen after embolization.

TECHNIQUE: Contrast-enhanced ultrasound imaging with 2ml of SonoVue (Bracco).

Etiology	Unknown. Main hypothesis related to hepatic arterial buffer response.
Incidence	0.6 - 10%
Gender ratio	No differences in gender.
Age predilection	No differences in age. 80% developed SASS in the first 60 days post-transplant.
Risk factors	Augmented spleen volume with pancytopenia and splenic artery enlargement.
Treatment	Splenic artery embolization or surgical banding-ligation. In the post-transplant setting, proximal endovascular splenic artery embolization is safer than surgery and presents minimal complications (i.e. hepatic artery thrombosis, biliary duct ischemia).
Prognosis	The outcome of SASS is related to the duration of the ischemia suffered by the hepatic artery. Early diagnosis and treatment are important.
Imaging findings	DSA is the gold standard test, it shows the early perfusion of the splenic or gastroduodenal artery; the delayed or dim perfusion of the hepatic artery; and the early portal venous contrast filling.

Table 1: Summary table of splenic artery steal syndrome.

	CT	**US**	**DSA**
Vascular complications	Angiographic phase in hepatic artery/anastomosis thrombosis: occlusion of the vessel by a thrombus. In case of hepatic artery stenosis: reduction in calibre of the vessel.	Thrombosis: absence of color-Doppler signal. Stenosis: hepatic artery with antegrade flow, turbulent flow whose maximum velocity is above the normal value. Distally parvus tardus flow.	Angiography: same as CT, with endovascular treatment options.
Splenic artery steal syndrome	Used to exclude other complications.	-low systolic peak in the hepatic artery (<35 cm/s) -increased hepatic artery resistance index (RI >0.8)	-early perfusion of the splenic or gastroduodenal artery -delayed or dim perfusion of the hepatic artery -early portal venous contrast filling
Infections	Diffused low attenuation of liver parenchyma, presence of fluid collection or necrosis	Hypoechogenic heterogeneous parenchyma, presence of fluid collections or necrosis	Not diagnostic for this condition.
Graft versus host disease	US-guided biopsy or Transjugular biopsy (in complicated patients) Histological diagnosis.	Hypoechogenic heterogeneous parenchyma, presence of fluid collections or necrosis.	Used to guide transjugular biopsy.

Table 2: Differential diagnosis table for splenic artery steal syndrome.

ABBREVIATIONS

BCLC = Barcelona Clinic Liver Cancer
CD-US = Color-Doppler ultrasound
CP = Child-Pugh
CT = Computer tomography
HABR = Hepatic arterial buffer response
HCC = Hepatocellular carcinoma
OLT = Orthotopic liver transplantation
PSV = Peak systolic velocity
RI = Resistance index
SASS = Splenic artery steal syndrome

KEYWORDS

Hepatocellular carcinoma; orthotopic liver transplant; angiography; splenic artery steal syndrome; embolization; ultrasound; interventional radiology; SAS; liver; spleen; CT; US

Online access
This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/4391

Peer discussion
Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity
This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad
www.EduRad.org