



HAL
open science

Hemorrhagic and necrotic adenoma associated with a congenital portosystemic shunt

J. Dumortier, A. Lachaux, S. Collardeau-Frachon, P. J. Valette

► **To cite this version:**

J. Dumortier, A. Lachaux, S. Collardeau-Frachon, P. J. Valette. Hemorrhagic and necrotic adenoma associated with a congenital portosystemic shunt. *Digestive and Liver Disease*, 2020, 10.1016/j.dld.2020.04.026 . hal-02901929

HAL Id: hal-02901929

<https://hal.science/hal-02901929v1>

Submitted on 22 Aug 2022

HAL is a multi-disciplinary open access archive for the deposit and dissemination of scientific research documents, whether they are published or not. The documents may come from teaching and research institutions in France or abroad, or from public or private research centers.

L'archive ouverte pluridisciplinaire **HAL**, est destinée au dépôt et à la diffusion de documents scientifiques de niveau recherche, publiés ou non, émanant des établissements d'enseignement et de recherche français ou étrangers, des laboratoires publics ou privés.



Distributed under a Creative Commons Attribution - NonCommercial 4.0 International License

Hemorrhagic and necrotic adenoma associated with a congenital portosystemic shunt

Jérôme DUMORTIER (1,2), Alain LACHAUX (2,3), Sophie COLLARDEAU-FRACHON (2,4),
Pierre-Jean VALETTE (2,5)

(1) Hospices civils de Lyon, Edouard Herriot hospital, Department of Digestive Diseases,

(2) Claude Bernard Lyon 1 University,

(3) Hospices civils de Lyon, Femme-Mère-Enfant hospital, Department of Pediatrics,

(4) Hospices civils de Lyon, Edouard Herriot hospital, Department of Pathology,

(5) Hospices civils de Lyon, Edouard Herriot hospital, Department of Digestive Radiology,

Lyon, France

Keywords: congenital portosystemic shunt; liver adenoma; necrosis; hemorrhage.

Running title: Complicated adenoma and congenital portosystemic shunt

Word count = 220

Conflicts of interest and sources of funding: None to declare.

Authors' contributions: J.D. and P.J.V. clinically managed the patient and wrote the manuscript; A.L., and S.C.F. clinically managed the patient. All authors read and approved the final version of the manuscript.

Abbreviations: CPSS: congenital portosystemic shunt.

Address for correspondence:

Jérôme Dumortier

pavillon L, Hôpital Edouard Herriot,

69437 Lyon Cedex 03, France

fax number : (33) 4 72 11 01 47

phone number : (33) 4 72 11 01 11

e-mail : jerome.dumortier@chu-lyon.fr

A 10-year-old girl was referred to our institution in September 2005 because of hyperammonemia. Abdominal Doppler ultrasound and CT scan led to the diagnosis of congenital portosystemic shunt (CPSS), associated with multiple liver nodules (figure 1A). Radiological follow-up disclosed progressively increasing size of liver nodules in May 2011 (figure 1B). The diagnosis of telangiectatic liver adenoma was made from a nodule biopsy. In March 2012, the patient was admitted because of acute right abdominal pain. A CT scan disclosed features of non-active intra-tumoral hemorrhage in the largest liver nodule (figure 1C). In June 2014, the patient was admitted again because of recurrence of acute right abdominal pain due to partial necrosis of the largest liver nodule (figure 1D). After the complication episodes in 2012 and 2014, clinical evolution was uneventful and the last MRI showed the persistent largest liver nodule (figure 1E).

Hepatic complications of CPSS include the development of benign liver tumors in 25% of reported cases (1). Since the spectrum of liver tumors in the context of CPSS is very large, liver lesions may, to some degree, mimic one another on imaging, and mischaracterization by imaging can frequently occur (2). Transformation of β -catenin-activated adenoma to HCC has been reported. We report here the first case of hemorrhagic and necrotic complications of a large liver adenoma complicating a CPSS.

References

1. Sokollik C, Bandsma RH, Gana JC, van den Heuvel M, Ling SC. Congenital portosystemic shunt: characterization of a multisystem disease. *J Pediatr Gastroenterol Nutr* 2013;56:675-81.
2. DiPaola F, Trout AT, Walther AE, et al. Congenital Portosystemic Shunts in Children: Associations, Complications, and Outcomes. *Dig Dis Sci* 2019.

Legends for figures

Figure 1: Time course of radiological appearance of liver nodules associated with CPSS

A: T1 MRI image after contrast injection. Only one lesion is visible in liver segments VII-VIII (30 mm diameter), strongly enhanced at arterial time (arrow). The mesenterico-hepatic intrahepatic shunt (*) is nearby, not enhanced at this timepoint.

B: T1 MRI after contrast injection. A second hepatic nodule (arrows) has appeared in segments IV-V (95 mm diameter), less markedly hypervascular than the first nodule which has otherwise decreased in size. This new lesion is slightly heterogeneous.

C: CT-scan, arterial phase after contrast injection. The nodule in segments IV-V (115 mm diameter) has complicated by intratumoral hemorrhage visible as hyperdense clotting (arrows) within a large necrotic area. Non-necrotic portions of the tumor (*) are still present in the periphery of the necrosis.

D: T1 MRI after contrast injection. The tumor (arrows), decreased in size (120 mm diameter) after intratumoral haemorrhage, contained both necrotic (*) and tissue portions.

E: T1 MRI after contrast injection. Six years after intratumoral haemorrhage, the nodule (arrows) in the segments IV-V has significantly re-increased in size (130 mm diameter), again slightly enhanced by the contrast injection. It has recovered its homogeneity, apart from a central scar image (*) that could suggest the diagnosis of focal nodular hyperlasia, without radiological features suggesting hepatocellular carcinoma.









