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Hemorrhagic and necrotic adenoma associated with a congenital portosystemic shunt

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Abbreviations: CPSS: congenital portosystemic shunt.

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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

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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.









