Nodular regenerative hyperplasia in chronic Budd-Chiari syndrome

The patient, who was affected by Budd-Chiari syndrome, was referred for evaluation of multiple focal liver lesions detected at US examination.

Imaging Findings:

The patient was admitted for evaluation of asthenia, hyporexia, low fever, jaundice and hepatosplenomegaly. Laboratory results were: total bilirubin 2.1 mg/dL, sALT 417 IU/L, sAST 99 IU/L, sGGT 99 IU/L, total protein 6.1 g/L, sAFP 2.5 ng/mL, prothrombin ratio 50%. Markers for hepatotropic viruses were negative. Sonographic examination and computed tomography did not show the hepatic veins: on the basis of these findings the diagnosis of Budd-Chiari syndrome was made.

Eight months later the patient underwent further investigations because of worsening of the general clinical status. Ultrasonography showed multiple hepatic nodules with a hypoechoic pattern whose diameter ranged from few millimetres to 2.5 centimetres. On spiral CT the nodules showed a relatively hyperdense appearance on unenhanced scans, marked contrast enhancement both in arterial and portal-venous phase, and wash-out of the contrast media in the delayed phase. On MR imaging the nodules were hyperintense on T1-weighted SE images and hypointense on T2-weighted FSE images; on dynamic scanning the nodules showed contrast enhancement in arterial and portal-venous phase, and wash-out in the delayed phase.

The treatment of choice is orthotopic liver transplantation.

Discussion:

Nodular regenerative hyperplasia (NRH) is a rare condition that is associated with myeloproliferative syndromes, lymphoproliferative syndromes, collagenopathies, graft versus host disease, and drugs. It consists of multiple regenerative nodules composed of large hyperplastic hepatocytes. The surrounding parenchyma is compressed and atrophic. There is no significant fibrosis in or around the nodules (1-5).

Patients with NRH are often asymptomatic; in half of cases portal hypertension can be demonstrated; in some instances NRH may cause bleeding (1,3).

The association of NRH with chronic Budd-Chiari syndrome has been described in isolated cases. The
pathogenesis remains unclear: a hypothesis by De Sousa et al. is that persistent sinusoidal congestion due to hepatic outflow obstruction leads to increased exposure of the hepatocytes to circulating hepatopoietins (3,4).

Imaging findings in NRH are variable, ranging from a normal hepatic appearance to multiple hepatic nodular lesions (1,2,3). On sonography, NRH appears as hypoechoic or isoechoic masses, with an anechoic centre if there is intranodular haemorrhage (1,2,3,5). On CT, NRH has an isoattenuating or relatively hyperattenuating pattern on precontrast scans. After administration of contrast material most nodules show contrast enhancement in arterial phase; in porto-venous phase some lesions have persisting enhancement, other become isodense (2,3,5). On MR imaging, the nodules have a hyperintense appearance on T1-weighted scans. On T2-weighted scans the signal intensity is variable: the lesions may be hyperintense, isointense or hypointense (2,3,4).

When multiple hepatic masses are found in patients affected by chronic Budd-Chiari syndrome, differential diagnosis is required between multinodular hepatocellular carcinoma (HCC) and NRH; for this purpose imaging may be helpful.

Both NRH and HCC show marked enhancement in arterial phase on spiral CT, but HCC typically shows rapid wash-out in portal-venous phase. Furthermore HCC usually displays low signal intensity on T2-weighted MR images and high signal intensity on T1-weighted MR images (3). HCC has a heterogeneous appearance, in contrast with the homogeneity of NRH (2).

In patients with Budd-Chiari syndrome multiple (more than 10), small (less than 4 cm) lesions are suggestive of benignity (2).

**Differential Diagnosis List:** Nodular regenerative hyperplasia

**Final Diagnosis:** Nodular regenerative hyperplasia

**References:**

Description: In the unenhanced CT scans, several focal liver lesions are visible. The nodules appear relatively hyperattenuating with respect to the surrounding parenchyma. Origin:
Description: In the arterial phase the hepatic lesions show clear contrast enhancement. **Origin:**

Description: In the portal-venous phase the nodules present persisting contrast enhancement. **Origin:**
Description: In the delayed phase the lesions show wash-out of the contrast medium. Origin:
Description: On T1-weighted SE images the multiple liver lesions have a hyperintense pattern. Origin:
Description: On T2-weighted FSE images the nodules appear hypointense.

Origin: