MR Imaging findings in patients with hereditary hemorrhagic telangiectasia and liver involvement

G. K. Schneider[1], A. Massmann[1], M. A. Kirchin[2], R. Seidel[1], P. Fries[1], U. W. Geisthoff[3], and A. Buecker[1]

[1]Clinic for Diagnostic and Interventional Radiology, Saarland University Hospital, Homburg/Saar, Germany, [2]Bracco Imaging S.p.A., Milan, Italy, [3]Department of Otorhinolaryngology, Hospitals of the City of Cologne, Cologne, Germany

Purpose: As yet there have been comparatively few reports describing MR imaging findings in the liver in patients with hereditary hemorrhagic telangiectasia (HHT, Rendu-Osler-Weber disease) despite the spectrum of liver involvement in affected patients ranging from discrete changes to life threatening right heart insufficiency and the possibility of significant morbidity and mortality. The aim of our study was therefore to characterize the spectrum of hepatic changes on MRI and the involvement of the hepatic vasculature in patients with HHT.

Materials and Methods: 230 patients (96 male, mean age: 47.9 years; 134 female, 45.8 years) with diagnosed HHT (according to established Curaçao criteria), or first-degree relatives, underwent preliminary screening MRI for cerebral, hepatic and pulmonary manifestations of the disease. Patients with positive findings for liver involvement on preliminary screening (enlarged hepatic artery [HA], inhomogeneous parenchyma, increased liver size, nodular lesions, dilated portal vein [PV]) thereafter underwent an additional targeted MRI examination of the liver before (non-contrast T1- and T2-weighted sequences) and after (contrast-enhanced dynamic and delayed T1-weighted sequences) intravenous administration of gadobenate dimeglumine (MultiHance, Gd-BOPTA) at a dose of 0.05 mmol/kg bodyweight. Image evaluation involved determinations of the diameter of HA and PV, the overall liver size (normal, moderate, considerable hepatomegalia), the dilation and tortuosity of the intrahepatic vessels (none, moderate, considerable), the presence of nodular hyperplastic changes of liver parenchyma and indirect signs of right heart insufficiency (dilated inferior caval vein, dilated right atrium/ventricle).

Results: Overall, 38/230 (16.5%) patients (8 [21.1%] male, 60.3 years; 30 [78.9%] female, 53.7 years) showed changes of the liver caused by HHT on screening studies. Hepatomegalia was present in 28/38 (73.7%) patients while enlarged, tortuous arterial vessels in the liver were observed in 21/38 (55.3%) patients. In addition to hepatic arteriovenous malformations (HAVMs), complex direct shunts to epigastric and retroperitoneal arterial vessels were observed in 3 patients. Nodular hyperplastic changes of the liver parenchyma characterized by arterial phase hypervascularization and late uptake of gadobenate dimeglumine in the liver-specific phase similar to that seen in FNH was observed in 19 (47%) patients. These changes were found in 3/14 (21.4%) patients with a HA diameter of <10 mm, in 10/18 (55.6%) patients with a HA diameter of 10-12 mm and in 5/6 (83.3%) patients with a HA diameter of >12 mm. Nine (23.7%) patients showed signs of right heart insufficiency caused by left-to-right shunting via the hepatic vascular malformations. Signs of right heart insufficiency were noted in 3/14 (21.4%) patients with HA diameter of <10 mm, 5/18 (27.8%) patients with a HA diameter of 10-12 mm and 1/6 (16.7%) patients with a HA diameter >12 mm. Notably the diameter of the PV showed an inverse correlation to the HA diameter with mean PV diameters of 15.4 mm, 15.7 mm and 12.7 mm for HA diameters of <10 mm, 10-12 mm and >12 mm, respectively. An increased rate of nodular hyperplastic changes of the liver parenchyma was noted with increasing diameter of the HA although the rate of right heart insufficiency decreased.

Conclusion: Arterio-portal and arterio-venous shunting in patients with HHT involving the liver may result in a variety of imaging findings among which hepatomegalia, nodular hyperplastic changes of the liver parenchyma, dilated tortuous intrahepatic vessels, increased HA and PV diameter as well as right heart insufficiency are the main imaging manifestations. A larger (>12 mm) diameter of the HA seems to correlate with an increased number of nodular hyperplastic changes of the liver parenchyma and a lower rate of right heart insufficiency. Conversely, an increased diameter of the HA seems to correlate with a smaller diameter of the PV. These findings could be explained by different microscopic vascular shunts in the liver in patients with HHT. Patients with considerable nodular hyperplastic changes of the liver parenchyma may have shunts predominantly at the sinusoidal level whereas in the case of only discrete nodular hyperplastic changes increased direct arterio-venous and arterio-portal shunts may be present. On the one hand an increased arterial blood supply to the liver tissue would explain the nodular hyperplastic changes of the liver parenchyma (i.e. a focal overgrowth of liver tissue similar to the proposed mechanism of development of FNH) while on the other hand this mechanism could also explain the decreased rate of patients with right heart insufficiency in the presence of nodular hyperplastic changes due to a tissue supply and no direct shunting of AV-malformations. Additional studies to evaluate the perfusion of liver parenchyma in patients with HHT may help to further understand the underlying mechanisms.