MR Imaging the Hepatic Manifestations and Complications of Hereditary Hemorrhagic Telangiectasia: A Pictorial Review

Christina Marie Chingkoe1, Errol Colak1, Vikram Prabhudesai1, Marie Faughnan2, and Anish Kirpalani1

1Department of Medical Imaging, St. Michael's Hospital, Toronto, Ontario, Canada, 2Division of Respiratory and Director of HHT Program, St. Michael’s Hospital, Toronto, Ontario, Canada

BACKGROUND:
Hereditary hemorrhagic telangiectasia (HHT), also known as Rendu-Osler-Weber syndrome, is an autosomal dominant disease characterized by cutaneous and visceral vascular abnormalities. While the cerebrovascular system and lungs are frequently involved, liver involvement is seen in up to 40% of patients, and is characterized by the presence of vascular lesions and intrahepatic shunting.1 Patients are often asymptomatic, and only present when later complications occur. MRI has an increasing role in the screening, diagnosis, and prognostication of liver disease among HHT patients.

PURPOSE: To review the MRI findings of the hepatic manifestations of HHT.

OUTLINE OF CONTENT
MRI Protocol
At our institution, MRI has replaced ultrasound as the most reliable imaging test for HHT patients with suspected liver disease. Our standard MRI protocol for hepatic HHT evaluation includes 3D fat-suppressed coronal high-resolution MRCP, axial fat-suppressed T1W 3D GRE before and after GBCA administration in multiple phases, phase contrast imaging through the hepatic artery and celiac axis ($V_{ref} = 150$ cm/s), plus other routine liver sequences at 1.5T.

Intrahepatic Shunts
Arteriovenous shunts have early opacification of the hepatic vein during arterial phase imaging (Figure 1). Arterioportal shunts are demonstrated by early and prolonged enhancement of the portal veins on arterial and late arterial phase imaging. Portovenous shunts are characterized by a dilated portal vein in communication with an enlarged hepatic or systemic vein.

Discrete Vascular Malformations and Focal Nodular Hyperplasia
Capillary telangiectases (small lesions with avid arterial enhancement, fading to isointensity) and large confluent vascular masses (> 10 mm size, avid arterial enhancement, with persistence on delayed phases, seen on Figure 2) are commonly seen.2 Large arteriovenous malformations (mass-like, tangle of vessels) are seen more rarely. Focal nodular hyperplasia (with typical MRI features) are commonly found in these patients as well.

Ischemic Biliopathy
Ischemic biliopathy is thought to occur from intrahepatic shunting of blood away from the biliary epithelium. MRCP and post-gadolinium imaging may demonstrate irregular biliary dilatation and strictures.

Complications
Hemosiderosis is demonstrated by diffuse hypointensity of the liver and spleen (Figure 3). Bilomas and intrahepatic abscesses commonly occur secondary to ischemic biliary strictures and vascular malformations, leading to sepsis. Hepatic encephalopathy, a rare complication, can be elucidated by T1 hyperintensities within the basal ganglia and globus pallidus.

SUMMARY
MRI has an increasingly important role in hepatic evaluation due to the benefits of simultaneous assessment of parenchymal, biliary, and vascular abnormalities without the use of ionizing radiation.

REFERENCES: