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

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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_{enc} = 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.[2]

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.[3] 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.[4]

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.

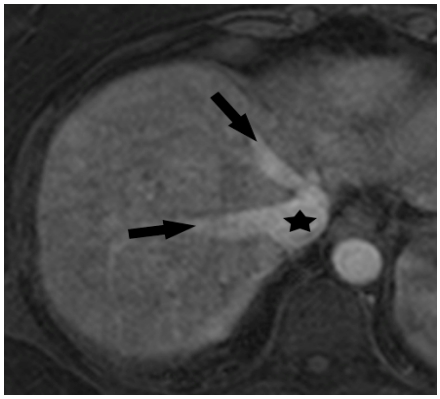

Figure 1: Axial arterial phase gadolinium-enhanced T1W FS images demonstrate simultaneous enhancement of the hepatic veins (black arrows), inferior vena cava (black star), and aorta, consistent with arteriovenous shunting.

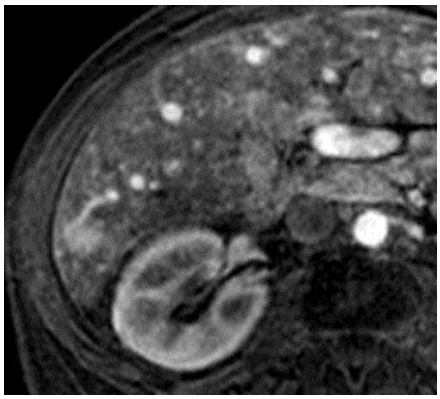

Figure 2: Confluent vascular mass on contrast-enhanced T1W, arterial phase, in the right hepatic lobe.

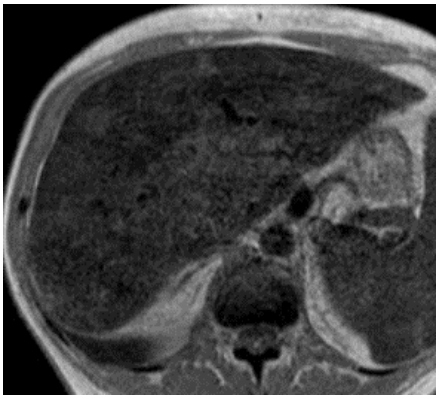

Figure 3: T1W opposed phase image demonstrating low signal within the liver and spleen, consistent with hemosiderosis.

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:

1. Abdalla SA and Letarte M. Hereditary haemorrhagic telangiectasia: current views on genetics and mechanisms of disease. *Journal of medical genetics*, 2006 Feb; 43(2):97-110.
2. Memeo M, Stabile Ianora AA, Scardapane A, Buonamico P, Sabba C, Angelelli G. Hepatic involvement in hereditary hemorrhagic telangiectasia: CT findings. *Abdom Imaging* 2004 Mar-Apr;29(2):211-220
3. Sabbà C, Pompili M. Review article: the hepatic manifestations of hereditary haemorrhagic telangiectasia. *Aliment Pharmacol Ther.* 2008 Sep 1;28(5):523-33.
4. Milot L, Gautier G, Beuf O, Pilleul F. Hereditary hemorrhagic telangiectases: magnetic resonance imaging features in liver involvement. *J Comput Assist Tomog*. 2006 May-Jun; 30(3):405-411.