Improved Detection and Diagnosis of Sturge-Weber Syndrome Using MR Susceptibility Weighted Imaging (SWI)

Y. Yu[1], C. Juhasz[2], Z. Kou[3], Y. Xuan[3], Z. Latif[3], K. Kudo[3], H. T. Chugani[2,3], M. E. Haacke[1,3], and J. Hu[3]

[1]MRI Institute of Biomedical Research, Detroit, MI, United States, [2]Department of Neurology, Wayne State University, Detroit, MI, United States, [3]Department of Radiology, Wayne State University, Detroit, MI, United States

INTRODUCTION: Sturge-Weber Syndrome (SWS) is a rare neurocutaneous disease. It is characterized by a vascular anomaly, leptomeningeal angiomatosis and abnormal cortical venous drainage. Cortical damage underlying the leptomeningeal angiomatosis can result from venous stasis leading to progressive hypoxia, necrosis and gliosis as well as cortical dystrophic calcification. Calcification is often observed in meningeal arteries and in cortical and subcortical veins underlying the leptomeningeal angiomatosis. Venous stasis causes hypoxia and an increase of local deoxyhemoglobin concentration. Deoxyhemoglobin (paramagnetic) and calcifications (diamagnetic) cause a small distortion of the local magnetic field. Susceptibility weighted imaging (SWI) is an imaging modality which exploits the magnetic susceptibility effects generated by local field inhomogeneities due to the presence of venous vasculature, blood products, iron depositions and calcifications. SWI is an exquisitely sensitive technique to detect the tissue intrinsic susceptibility property change without administration of a contrast medium. The purpose of our study was to evaluate the use of high resolution SWI technique in the detection and diagnosis of SWS.

METHODS: Thirteen children with the diagnosis of SWS were recruited prospectively. MR studies were carried out on a 1.5 T Siemens Sonata system using the standard head coil. The MR protocol includes 3D high resolution SWI and gadolinium-enhanced T1W. The gadolinium-enhanced T1W was performed after injection of 0.1mmol/kg Gd-DTPA, Magnevist. The SWI acquisition was performed with a turbo 3D gradient echo sequence with FA= 20°, TR/TE=89/40 ms, EPI factor= 5, acquisition matrix= 512x256x48, FOV= 256x256x96 mm^3. Flow compensation was applied in all 3 directions. Both magnitude and phase images were saved for further post-processing and analysis. Two investigators evaluated both SWI and T1-Gd images and made a consensus agreement on six types of imaging findings by using a four-grade scoring system. The performance of SWI vs. T1-Gd images was then compared for each type of abnormality.

RESULTS: SWI was superior to T1-Gd in identifying the enlarged transmedullary veins (p=0.0020), abnormal periventricular veins (p=0.0078), cortical gyriform abnormalities (p=0.0020), and grey matter/white matter junction abnormalities (p=0.0078). Conversely, T1-Gd was better than SWI in identifying enlarged choroid plexus (p=0.0050) and leptomeningeal abnormalities (p=0.0050).

DISCUSSION AND CONCLUSION: High resolution SWI offers the potential to visualize the presence of subtle venous abnormalities and typical cortical gyriform calcification. It provides useful diagnostic information which could complement to the standard contrast enhanced T1W in the clinical setting for SWS patients. SWI is particularly suitable to monitor the progression of the SWS disease longitudinally.

ACKNOWLEDGEMENTS: Grant support by Michigan Technology Tri-Corridor Fund, MTTC 085P5200251 (to EMH) and The National Institute of Neurological Disorders and Stroke, grant RO1-NS041922 (to CJ).

REFERENCES: 1) KA Thomas-Sohl, *et al.*, Pediatric Neurology 2004, 30:303-310.
2) EM Haacke, *et al.*, Magn Res Med, 2004,52:612-8. 3) JR Reichenbach *et. al*, Radiology 1997,204:272-7

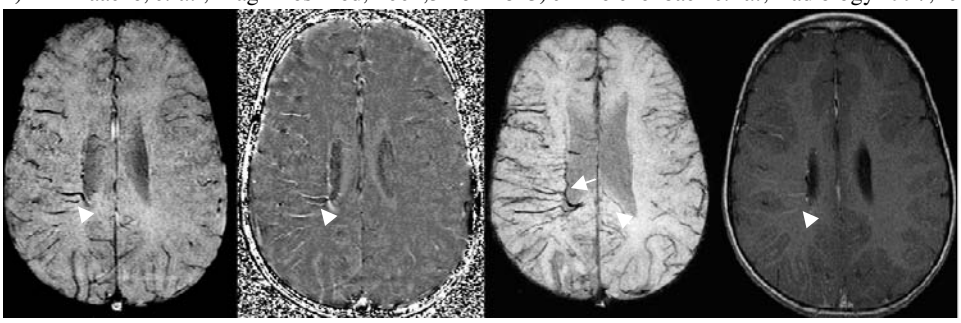

Fig. 1: SWI images magnitude (a), phase (b), mIP image (over 8 adjacent slices with slab thickness of 16mm; (c), T1W post-gadolinium (d). The abnormal transmedullary veins (arrow head) and periventricular veins (arrow) are shown on the SWI phase and magnitude images, and better visualized on SWI mIP image than the T1 post.

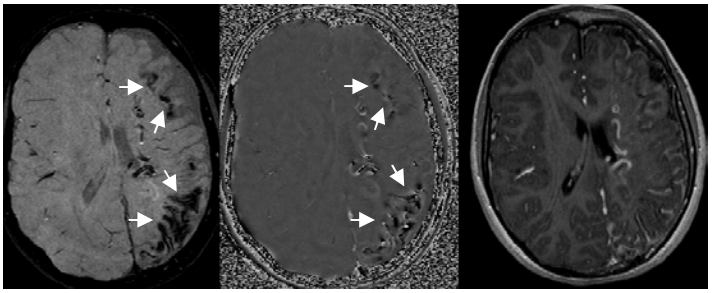

Fig. 2: SWI magnitude (left), phase (middle) demonstrates cortical gyriform hypointense lesions (arrow) which are not shown on T1 post-contrast image (right). The gyriform hypointense lesions presumably reflect the typical calcification.