

Enhanced Detection of Vascular Abnormalities in Sturge-Weber Syndrome using SWI

J. Hu¹, J. Neelavalli¹, C. Juhasz², O. Muzik², K. Tong³, S. Ashwal³, Y. Shen¹, Y. Xuan¹, Z. Latif¹, V. Sehgal¹, D. Chugani², H. Chugani², M. E. Haacke¹

¹Radiology, Wayne State University, Detroit, MI, United States, ²Children's Hospital of Michigan, Detroit, MI, United States, ³Loma Linda University, Loma Linda, California, United States

Introduction: Sturge-Weber syndrome (SWS) is an often progressive but rare neurocutaneous disorder. The usual pathologic manifestations are angiomatous facial malformation (port-wine stain), along with leptomeningeal venous angiomatosis. The venous stasis is associated with abnormal collateral venous drainage, and the progressive venous occlusions are believed to be primarily responsible for neurologic deterioration and hemispheric atrophy (5-7). Susceptibility-weighted imaging (SWI) in the recent past has proven to be a new means to greatly enhance visualization of veins, venous malformations and microhemorrhage in brain (1-3). In this study, we present preliminary data, investigating the use of SWI in SWS.

Materials and Methods: Six patients diagnosed with SWS were imaged at 1.5 T using a standard Siemens head coil. The protocol included the conventional contrast enhanced (CE) T1 weighted spin echo (SE) sequence and the susceptibility weighted sequence. SWI is a high resolution, fully flow compensated, 3D gradient echo sequence. One of the patients was imaged pre and post-contrast for both T1 weighted imaging and SWI. The other 5 patients were imaged with a pre and post contrast T1 sequence but only post contrast SWI was used. T1 weighted 2D SE was acquired in interleaved mode with 2 concatenations, an in plane resolution of 0.86 x 0.86 mm and a slice thickness of 3 mm. SWI (3D gradient echo, TR/TE/ α = 60ms/20 and 40ms/20° double echo) was acquired with an in plane resolution of 1.72x 0.86 mm², and slice thickness of 2 mm. To increase the visibility of the venous vessels, a phase mask was constructed from each original phase image and this mask filter was multiplied 4 times with the corresponding magnitude image (1).

Results: Whereas the contrast enhanced T1 weighted images showed leptomeningeal enhancement throughout the involved cortex, susceptibility weighted images clearly delineated the abnormally large collateral veins deep in the white matter as well as ill-defined areas of T2 shortening that could represent increased tissue deoxyhemoglobin or calcification. Figures 1 and 2 show the comparison between information provided by the SWI and conventional CE T1 weighted images. The SWI images are complementary to the conventional T1 weighted images giving clear, specific location and extent information of the venular dysplasia as compared to the diffuse leptomeningeal enhancement. Although the vascular anomalies are seen in CE MRI as regions of enhancement, the SWI images can demonstrate the venous component of these anomalies prominently, even without the need for a contrast agent.



Fig 1.a



Fig 1.b

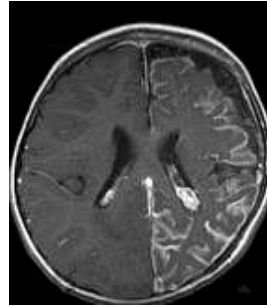


Fig 1.c

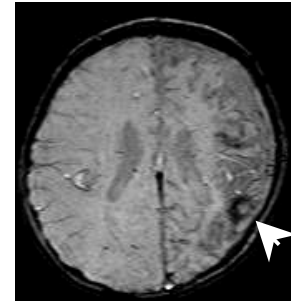


Fig 1.d



Fig 2a



Fig 2.b

Fig.1.a) CE T1 weighted image showing the leptomeningeal enhancement (b) Corresponding post-contrast SWI showing markedly hypointense region in the underlying white matter that could reflect increased tissue deoxyhemoglobin or calcification.

(c) CE T1 weighted image (d) Corresponding SWI image with No-Contrast. SWI shows a portion of the affected region as hypointense even without contrast agent.

Fig.2. a) Maximum intensity projection image of CE T1 weighted images over 6 mm. b) Corresponding minimum intensity projection SWI images. SWI clearly shows the conspicuity of the abnormal collateral venous vasculature.

Discussion: As SWI relies on the BOLD effect of deoxyhemoglobin (1-3), it enhances the visibility of the venous abnormalities to a greater extent than conventional methods. Use of a contrast agent improves the signal cancellation effect in a partial volumed vessel and thus enhances the visibility of vessels of subvoxel dimensions (2). All these characteristics make SWI very much sensitive to even small venous vascular structures. In the scanned patients SWI images clearly showed the specific areas of venous abnormality/possible areas of calcification, in addition to the leptomeningeal enhancement. The regions of venous angiomatosis are known to be causally associated with the areas of neuronal degeneration (4-6) and hence early detection may be important for potential therapeutic intervention. These areas of venous malformations can be seen even without a contrast agent using SWI (1-3) and may be ideally suited for early imaging of small infants. In conclusion, SWI may improve early detection of vascular pathology in patients with SWS, and may contribute to our understanding of the pathophysiology of disease progression.

References:

- 1). Reichenbach JR, Venkatesan R, Schillinger DJ, Kido DK, Haacke EM, Radiology 1997; 204:272-277. 2). J.R. Reichenbach, M. Essig, E. M. Haacke, B. C. Lee, C. Przetak, W. A. Kaiser, L. R. Schad, MAGMA 6:62-69 (1998). 3). W. Lin, P. Mukherjee, H. An, Y. Yu, Y. Wang, K. Vo, B. Lee, D. Kido, E. M. Haacke, JMRI 10:118-123(1999). 4)J. K. Cure, K. R. Holden, P. V. Tassel. AJNR 16:1539-1542. (1995). 5). P. Portilla et al, AJNR 23:490-492 (2002). 6). A. Aydin et al, Pediatr Neurol 22(5):400-402: (2002).