

Teaching NeuroImage: Sturge-Weber Syndrome in an Adult

Fábio A. Nascimento, MD, John R. McLaren, MD, M. Brandon Westover, MD, PhD, Sahar F. Zafar, MD, and Steven M. Stuffelbeam, MD

Neurology® 2022;98:814-815. doi:10.1212/WNL.0000000000200512

Correspondence

Dr. Nascimento
nascimento.fabio.a@gmail.com

Figure 1 Skin Examination



Facial capillary malformation (port-wine stain) involving the first division of the trigeminal nerve in the right hemiface.

We report a 19-year-old right-handed man with a history of Sturge-Weber syndrome (SWS) based on port-wine stain involving the first division of the trigeminal nerve in the right hemiface (Figure 1) and leptomeningeal capillary-venous malformations associated with calcification involving the ipsilateral occipital lobe (Figures 2, E–G) and resultant refractory epilepsy. He was referred to our center for a presurgical epilepsy evaluation. Video-EEG data showed electroclinical and electrographic seizures arising from the right frontotemporal region and right posterior quadrant, respectively. In addition, there were frequent right temporal interictal discharges. He also had evidence of right hippocampal sclerosis (Figures 2, C and D), suspected to be the result of longstanding refractory epilepsy (i.e., dual pathology). SWS is a neurocutaneous disorder characterized by ipsilesional facial and leptomeningeal capillary-venous malformations with regional atrophy, gyral calcification, focal leptomeningeal enhancement, and bony changes (Figures 2, A, B, E–H).^{1,2}

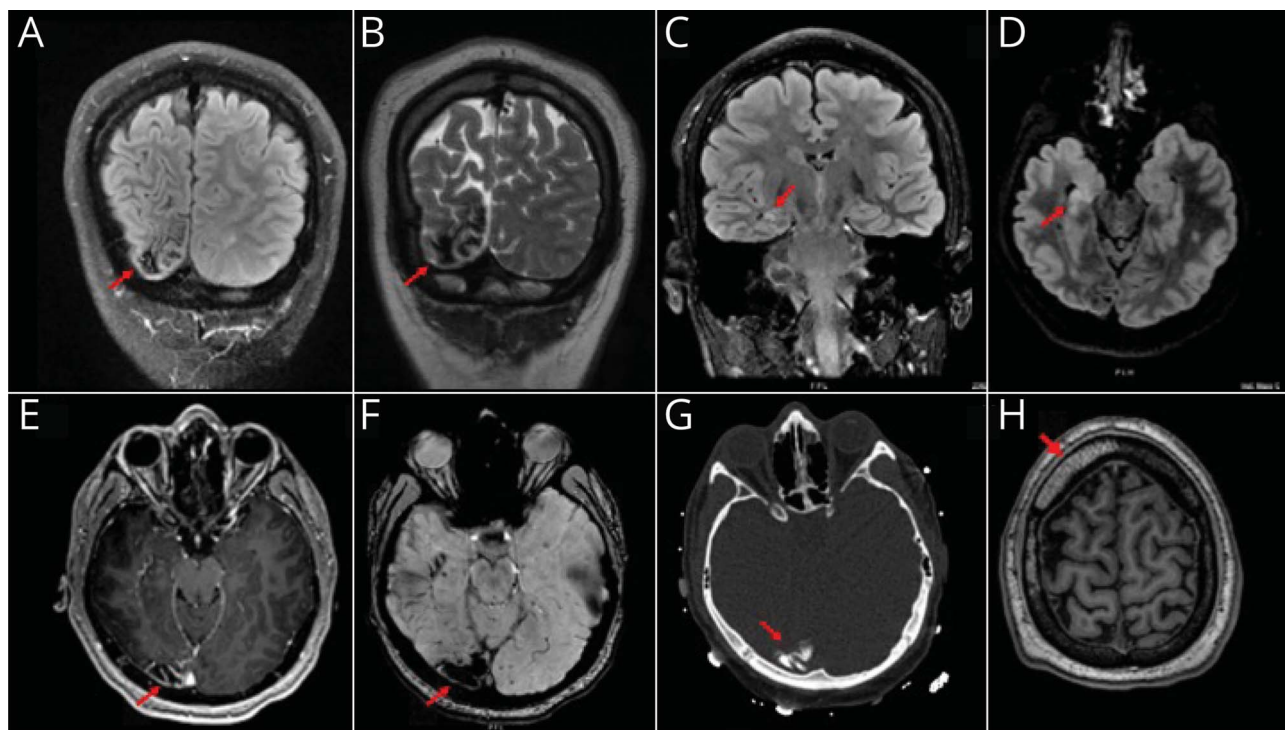
MORE ONLINE

Teaching slides

links.lww.com/WNL/B893

From the Departments of Neurology (F.A.N., J.R.M., M.B.W., S.F.Z.), and Radiology (S.M.S.), Massachusetts General Hospital, Harvard Medical School, Boston, MA. Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Figure 2 Brain MRI Findings Consistent With Sturge-Weber Syndrome: Coronal FLAIR (A), T2 FSE (B), Coronal (C) and Axial (D) FLAIR, Postcontrast Axial T1 (E), Axial SWI (F), Axial CT (G), and Axial T1 (H)



Atrophy predominantly involving the right occipital lobe (A and B, arrows), regional leptomeningeal enhancement consistent with temporo-occipital pial angioma (E, arrow), and local susceptibility blooming and low T1/T2 signal (F, arrow) with corresponding high attenuation (G, arrow) consistent with calcification. Asymmetric thickening and relative T1 hypointensity of the right frontal/parietal bones (H, arrow). Right hippocampal atrophy and hyperintensity (C and D, arrows).

Study Funding

No targeted funding reported.

Disclosure

F. Nascimento is a former member of the Neurology Resident and Fellow Section Editorial Board. J. McLaren, M. B. Westover, S. Zafar, and S. Stufflebeam report no disclosures relevant to the manuscript. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

Publication History

Received by *Neurology* October 16, 2021. Accepted in final form February 28, 2022. Submitted and externally peer reviewed. The handling editor was Roy Strowd III, MD, Med, MS.

Appendix Authors

Name	Location	Contribution
Fábio A. Nascimento, MD	Department of Neurology, Massachusetts General Hospital, Harvard Medical School, Boston, MA	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data
John R. McLaren, MD	Department of Neurology, Massachusetts General Hospital, Harvard Medical School, Boston, MA	Drafting/revision of the manuscript for content, including medical writing for content, and analysis or interpretation of data

Appendix (continued)

Name	Location	Contribution
M. Brandon Westover, MD, PhD	Department of Neurology, Massachusetts General Hospital, Harvard Medical School, Boston, MA	Drafting/revision of the manuscript for content, including medical writing for content
Sahar F. Zafar, MD	Department of Neurology, Massachusetts General Hospital, Harvard Medical School, Boston, MA	Drafting/revision of the manuscript for content, including medical writing for content
Steven M. Stufflebeam, MD	Department of Radiology, Massachusetts General Hospital, Harvard Medical School, Boston, MA	Drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; and analysis or interpretation of data

References

- Comi AM. Presentation, diagnosis, pathophysiology, and treatment of the neurological features of Sturge-Weber syndrome. *Neurologist*. 2011;17(4):179-184.
- Warne RR, Carney OM, Wang G, et al. The bone does not predict the brain in Sturge-Weber syndrome. *AJNR Am J Neuroradiol*. 2018;39(8):1543-1549.

Neurology[®]

Teaching NeuroImage: Sturge-Weber Syndrome in an Adult
Fábio A. Nascimento, John R. McLaren, M. Brandon Westover, et al.
Neurology 2022;98:814-815 Published Online before print April 11, 2022
DOI 10.1212/WNL.0000000000200512

This information is current as of April 11, 2022

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/98/19/814.full
References	This article cites 2 articles, 1 of which you can access for free at: http://n.neurology.org/content/98/19/814.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Education http://n.neurology.org/cgi/collection/all_education Amyotrophic lateral sclerosis http://n.neurology.org/cgi/collection/amyotrophic_lateral_sclerosis_MRI MRI http://n.neurology.org/cgi/collection/mri Other cerebrovascular disease/ Stroke http://n.neurology.org/cgi/collection/other_cerebrovascular_disease__stroke Other neurocutaneous disorders http://n.neurology.org/cgi/collection/other_neurocutaneous_disorders
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2022 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

