Teaching NeuroImage: Sturge-Weber Syndrome in an Adult

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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]

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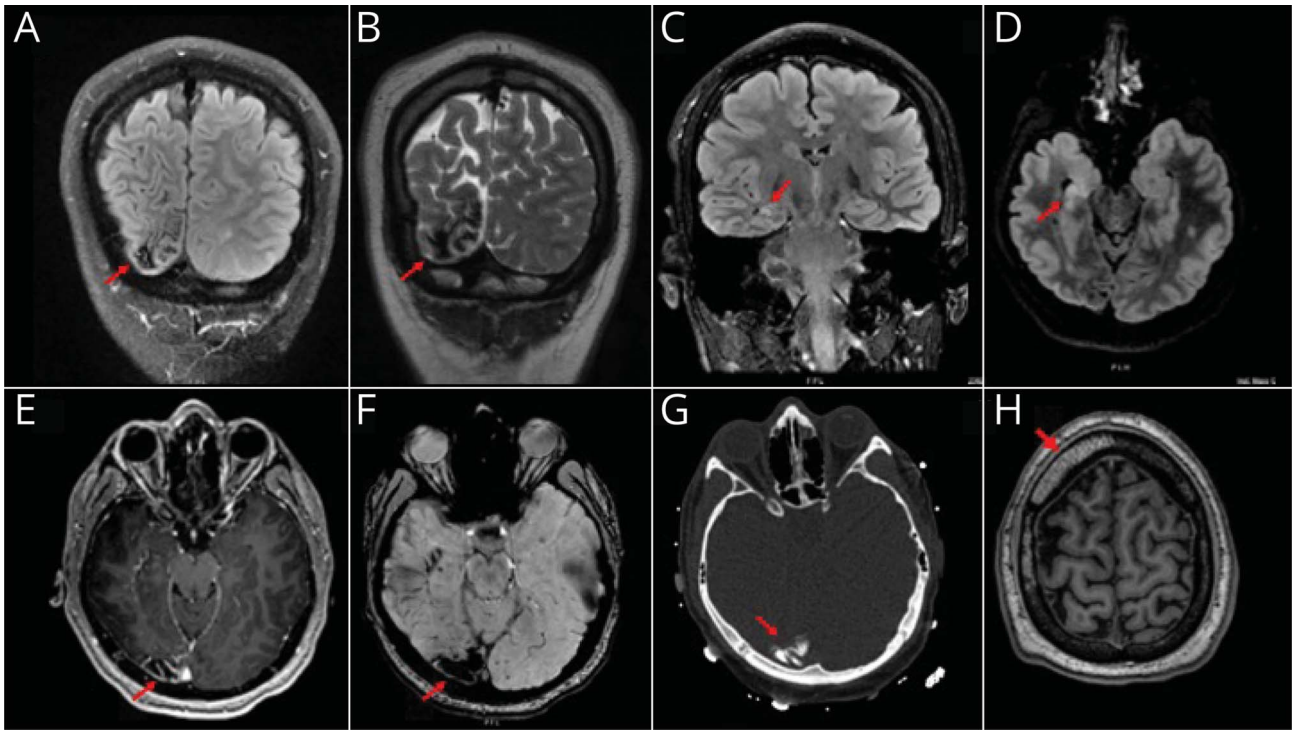
Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

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).

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Appendix *(continued)*

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