Teaching NeuroImages:

Sturge-Weber syndrome presenting in a 58-year-old woman with seizures

Rebecca Traub, MD
Claire Riley, MD
Susanna Horvath, MD,
FAHA

Address correspondence and
reprint requests to Dr. Rebecca
Traub, Neurological Institute,
Columbia University Medical
Center, 710 West 168th Street,
New York, NY 10032
rt2238@columbia.edu

Figure 1 Facial angioma of Sturge-Weber syndrome

Facial angioma (port wine birthmark) over the left V1 and V2 and right V2 distribution in Sturge-Weber syndrome.

A 58-year-old woman with a history of migraine presented with headache, aphasia, right homonymous hemianopsia, and right hemiparesis, followed by 2 seizures. On examination, she was noted to have a facial

angioma (figure 1) that was present from childhood. MRI findings (figure 2, A–C) were consistent with leptomeningeal angiomatosis, making the diagnosis of Sturge-Weber syndrome. Sturge-Weber syndrome is a sporadically occurring (nonhereditary) neurocutaneous syndrome, hypothesized to be caused by somatic mutations in fibronectin expression causing capillary malformation¹ and dysfunctional cortical perfusion.² It is usually diagnosed in infancy, but it may occasionally present in adulthood with seizures or strokelike episodes.

REFERENCES

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Figure 2 MRI in Sturge-Weber syndrome

(A) T2 fluid-attenuated inversion recovery hyperintensity in the left temporal and occipital cortex; (B) T1 postcontrast leptomeningeal enhancement over the left parieto-occipital region; and (C) 3-dimensional SWAN magnetic susceptibility showing enlarged left parietal draining veins consistent with leptomeningeal angiomatosis.

From the Neurological Institute, Columbia University Medical Center, New York, NY.

Disclosure: Dr. Traub reports no disclosures. Dr. Riley has received speaker honoraria from Teva Pharmaceutical Industries Ltd. and Bayer Schering Pharma and has served as a consultant for EMD Serono, Inc. and Questcor Pharmaceuticals, Inc. Dr. Horvath serves on the editorial board of *Clinical Neurology News*.

Neurology®

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Neurology 2010;75:e52

DOI 10.1212/WNL.0b013e3181f39a52

This information is current as of September 20, 2010

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