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

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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 malformation and dysfunctional cortical perfusion. It is usually diagnosed in infancy, but it may occasionally present in adulthood with seizures or stroke-like episodes.

REFERENCES

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.

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.

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