

Clinical findings of the phakomatoses: Sturge–Weber syndrome

Mark Quigg, MD, MSc; Robert S. Rust, MD; and James Q. Miller, MD†



Figure 1. (A) Ophthalmic port-wine stain (PWS) in an infant. Glaucoma is a frequent complication. PWS that spares ophthalmic trigeminal distribution is usually not indicative of underlying Sturge–Weber syndrome.¹ (B) Bilateral or more lower facial involvement occurs in 15%.¹ As in this child, about half have an extracranial nevus that typically respects segmental anatomy.

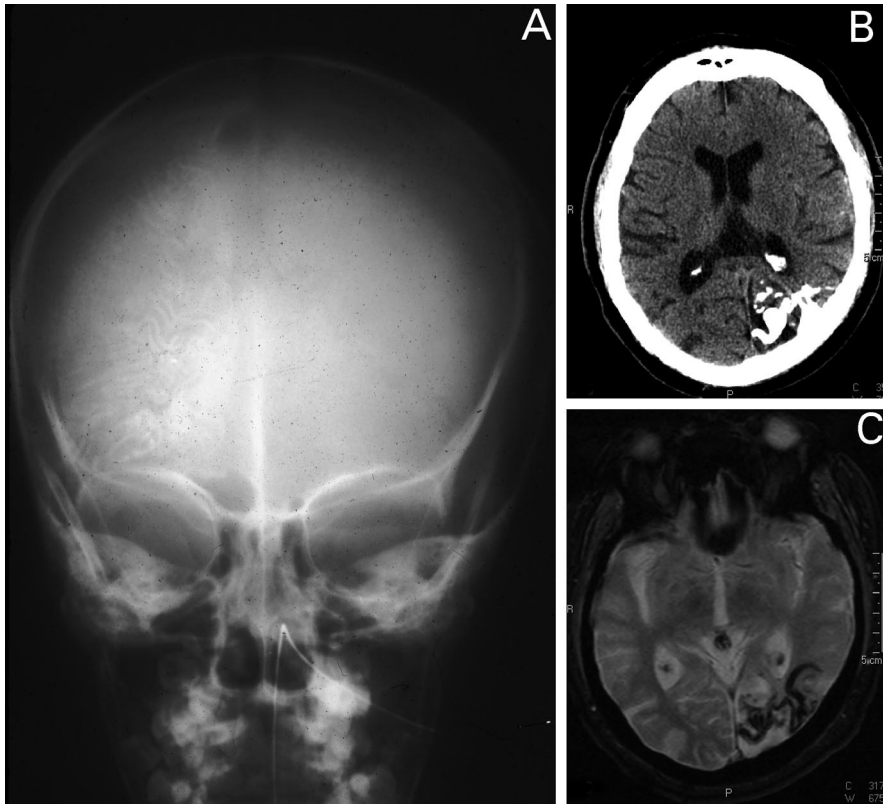


Figure 2. (A) A-P skull film that demonstrates the gyral calcifications of the subarachnoid angiomas of Sturge-Weber syndrome, tortuous parallel “tram tracks” across most of the right hemisphere. (B) In another patient, uncontrasted axial CT and (C) gradient-echo axial MRI show gyral calcifications, atrophy, and gliosis of the underlying cortex due to leptomeningeal involvement.

The dermatologic hallmark of Sturge-Weber syndrome (SWS) is the nevus flammeus (port-wine stain, PWS), a angiomatous lesion usually distributed in the region innervated by the ophthalmic branch of the trigeminal nerve¹ caused by lack of normal regression of embryonic vascular plexus of the cephalic neural tube. The

variable constellation of clinical findings of hemiparesis, mental retardation, and epilepsy is caused by the resulting leptomeningeal and cortical angiomatosis and calcification. SWS is a congenital and nonhereditary disorder. Physical findings of SWS are shown in figure 1 and neuroimaging in figure 2.

Acknowledgment

The authors thank C. Douglas Phillips of the Division of Neuroradiology for his contributions of MRI images.

Reference

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Disclosure: The authors report no conflicts of interest.

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Neurology[®]

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Neurology 2006;66;E17-E18

DOI 10.1212/01.wnl.0000197789.72780.a9

This information is current as of February 27, 2006

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