Teaching Neuro*Image*

Clinical findings of the phakomatoses: Sturge–Weber syndrome

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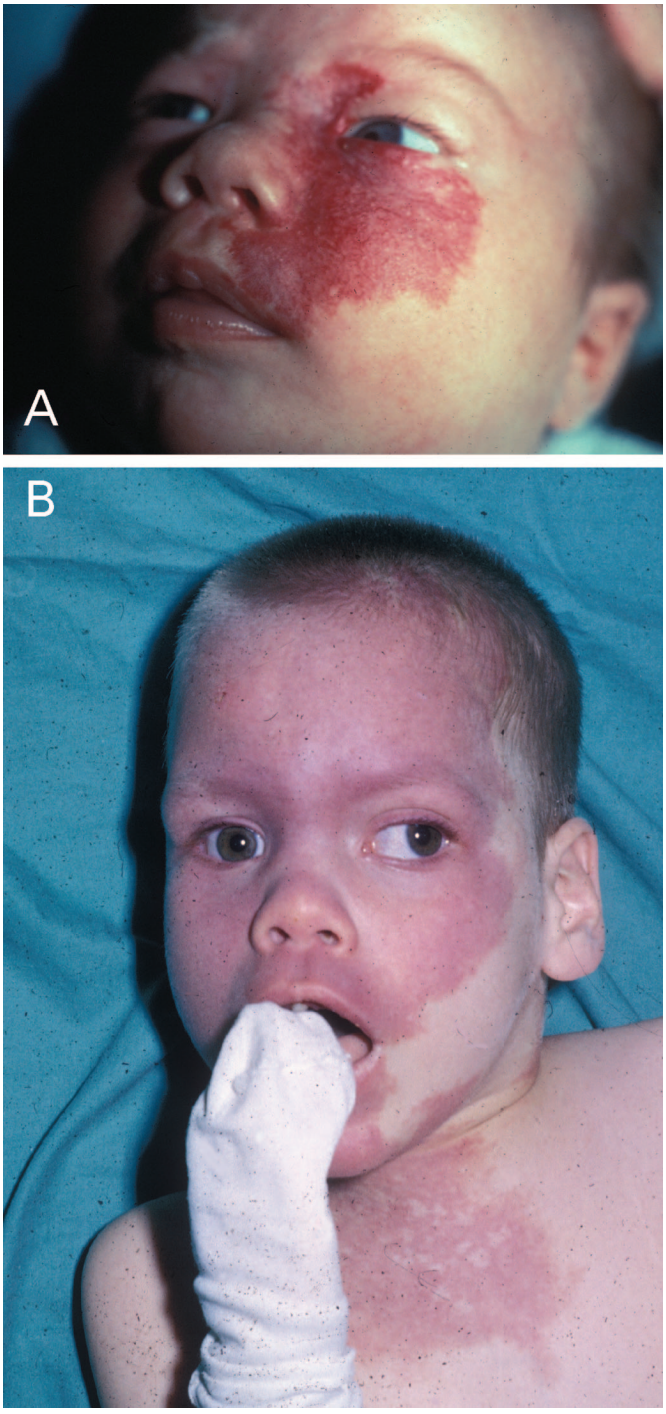

Figure 1. (A) Ophthalmic port-wine stain (PWS) in an infant. Glaucoma is a frequent complication. PWS that spares ophthalmic trigeminal distribution is usually not indicative of underlying Sturge–Weber syndrome.[1] (B) Bilateral or more lower facial involvement occurs in 15%.[1] As in this child, about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

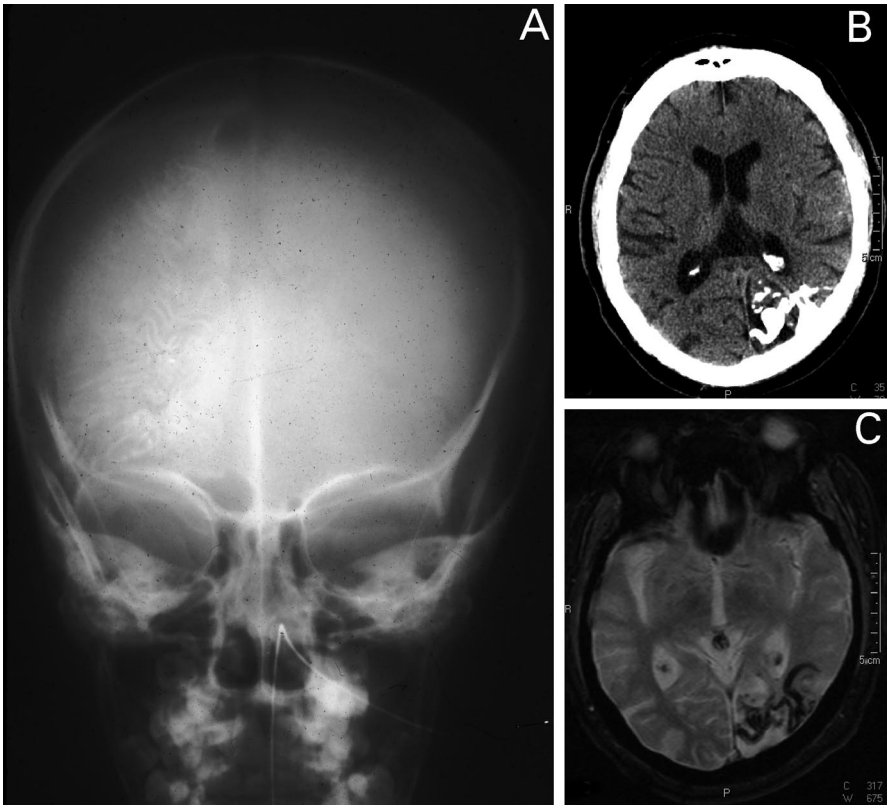

Figure 2. (A) A-P skull film that demonstrates the gyral calcifications of the subarachnoid angiomas of Sturge–Weber syndrome, tortuous parallel "tram tracks" across most of the right hemisphere. (B) In another patient, uncontrasted axial CT and (C) gradient-echo axial MRI show gyral calcifications, 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 nerve[1] 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.

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Reference

1. Tallman B, Tan OT, Morelli JG, et al. Location of port-wine stains and the likelihood of ophthalmic and/or central nervous system complications. Pediatrics 1991;87:323–327.

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