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Unusual Findings in Sturge-Weber Syndrome

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Unusual Findings in Sturge-Weber Syndrome

Sturge-Weber syndrome is a neurocutaneous condition characterized by nevus of the face in the territory of one or more divisions of the trigeminal nerve on the same side as the cerebral lesion, intracranial calcifications, convulsive disorders, glaucoma, hemiparesis, hemiatrophy, and mental deficiency. In 1987, Chaudary and Brudnicki [1] described a case of Sturge-Weber syndrome in which intracranial calcifications were contralateral to the bulk of the facial nevus, the intelligence was normal, and the patient had no history of seizure disorders. We recently observed a similar case.

A 54-year-old man was admitted because he complained of two episodes of transient left hemianopia. The patient had an extensive facial nevus affecting the right supraorbital area, normal intelligence, and no history of seizures. Neurologic examination was normal. Doppler examination of the neck arteries and transcranial Doppler sonography were normal. CT examination of the brain showed gyriform calcifications in the left occipital region (Fig. 1). Some enhancement was seen after the IV injection of contrast medium. Enlargement, calcification, and slight enhancement of the left choroid plexus also were observed, thus confirming the report of Stimac et al. [2], which described these as common findings in Sturge-Weber syndrome.

As in the case of Chaudary and Brudnicki [1], in our patient the intracranial calcifications were contralateral to the nevus of the face, the patient was of normal intelligence, and seizure disorders were absent. Furthermore, cerebral hemiatrophy was absent, and the presenting symptom was by transient ischemic attacks.

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2. Stimac GK, Solomon MA, Newton TH. CT and MR of angiomatous malformations of the choroid plexus in patients with Sturge-Weber disease. *AJNR* 1986;7:623-627

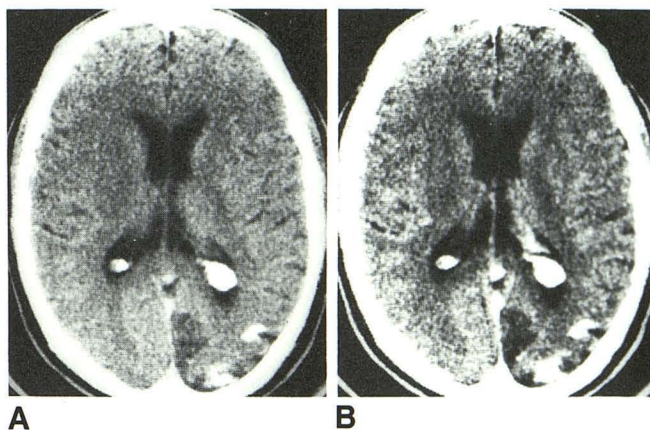


Fig. 1.—Unusual findings in Sturge-Weber disease.
A, CT scan of brain shows gyriform calcifications in left occipital region.
B, CT scan performed after IV injection of contrast medium shows some enhancement.