Sturge-Weber Syndrome with Posterior Fossa Involvement

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Summary: We present a case of Sturge-Weber syndrome with involvement of the cerebellum in addition to the occipital and temporal lobes.

Index terms: Phakomatoses; Posterior fossa, abnormalities and anomalies; Cerebellum, abnormalities and anomalies

Sturge-Weber syndrome, encephalotrigeminal angiomatosis, is a neurocutaneous disorder characterized by a facial cutaneous vascular nevus, seizures, mental retardation, hemiplegia, homonymous hemianopia, and buphthalmos or glaucoma (1). The clinical findings are related to vascular anomalies of the face, ocular choroid, and leptomeninges. The imaging findings in Sturge-Weber syndrome may include cerebral calcifications, cerebral atrophy, choroid plexus anomalies, pial and/or cortical enhancement, and prominent deep-draining veins. The leptomeningeal vascular anomaly is usually located in the anterior occipital, posterior parietal, and temporal cerebrum.

Case Report

The subject of this report is a 6-year-old boy with a facial port-wine nevus involving the first and second divisions of the fifth cranial nerve, the right scalp, right pinna, and right posterior auricular region. He also had right conjunctival telangiectasias and hypertrophy of the upper gingiva. At 2 years of age he had a petit mal seizure consisting of a staring spell with prolonged eye deviation. He was subsequently admitted to the community hospital where he had a Jacksonian seizure with left body involvement and eyes deviated to the left. An electroencephalogram at that time showed decreased beta activity and slowing over the right hemisphere. The computed tomographic (CT) and magnetic resonance (MR) studies were reportedly normal. The patient discontinued anticonvulsive therapy after 1 year. The patient suffered no further seizures and was developing normally until approximately 2 years later when he was found staggering about and had a left eye gaze paralysis with nystagmus. CT showed calcification in the right temporal and anterior occipital lobes (Fig 1). A contrast-enhanced CT showed prominent foliaform and vascular enhancement of the right cerebellum (Fig 1). MR showed increased T2 signal intensity in the right cerebellar hemisphere and prominent vascular flow voids (Fig 2). The postcontrast T1-weighted images showed enhancement conforming to the folia of the right cerebellum and over the cortical surface of the right temporal lobe and prominent vascular enhancement over the right temporooccipital region (Fig 2). An arteriogram demonstrated superficial and deep venous anomalies of the right cerebellum and of the right occipital and temporal lobes (Fig 3). The arterial and capillary phases were normal.

Discussion

Sturge-Weber syndrome was clinically first described by W. Allen Sturge in 1879 (2). The syndrome was confirmed pathologically by Kalischer in 1874, histologically by Volland in 1913, and radiographically by Weber in 1922, and Dimitri in 1923 (3). The syndrome of encephalotrigeminal angiomatosis consists of congenital facial nevus in the distribution of the fifth cranial nerve, classically with involvement of the ophthalmic division, along with ipsilateral or bilateral leptomeningeal vascular anomalies of the occipital, parietal, or temporal lobes, and vascular anomalies of the ocular choroid. Clinically there are seizures, mental retardation, hemiplegia, hemianopia, or glaucoma (1, 4–6).

Reported CT and MR findings include those consistent with cerebral atrophy, calcifications, infarction, gliosis, and demyelination (7–15). Contrast enhancement of the pial vascular malformation and underlying cortex has been reported, along with contrast enhancement of associated deep venous and choroid plexus anomalies (7–15). Angiography characteristically shows delayed opacification or nonopacification of the

Received September 10, 1992; accepted pending revision November 23; revision received December 11.

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AJNR 15:389–392, Feb 1994 0195-6108/94/1502–0389

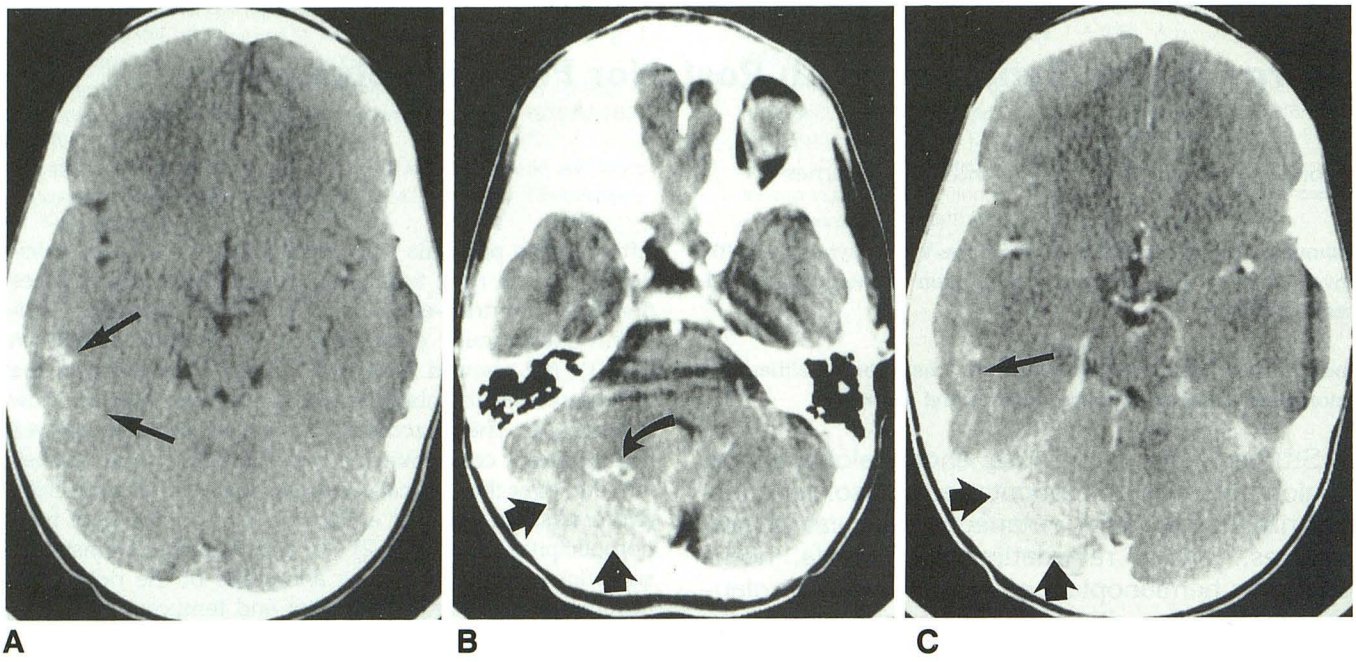

Fig. 1. CT before (*A*) and after (*B* and *C*) contrast enhancement shows right temporal cortical calcification (*long arrows*) plus right cerebellar hemispheric foliaform enhancement (*block arrows*) and deeper vascular enhancement (*curved arrow*), the latter similar to that seen with venous angiomas. There is also right temporal cortical enhancement (*long arrows*).

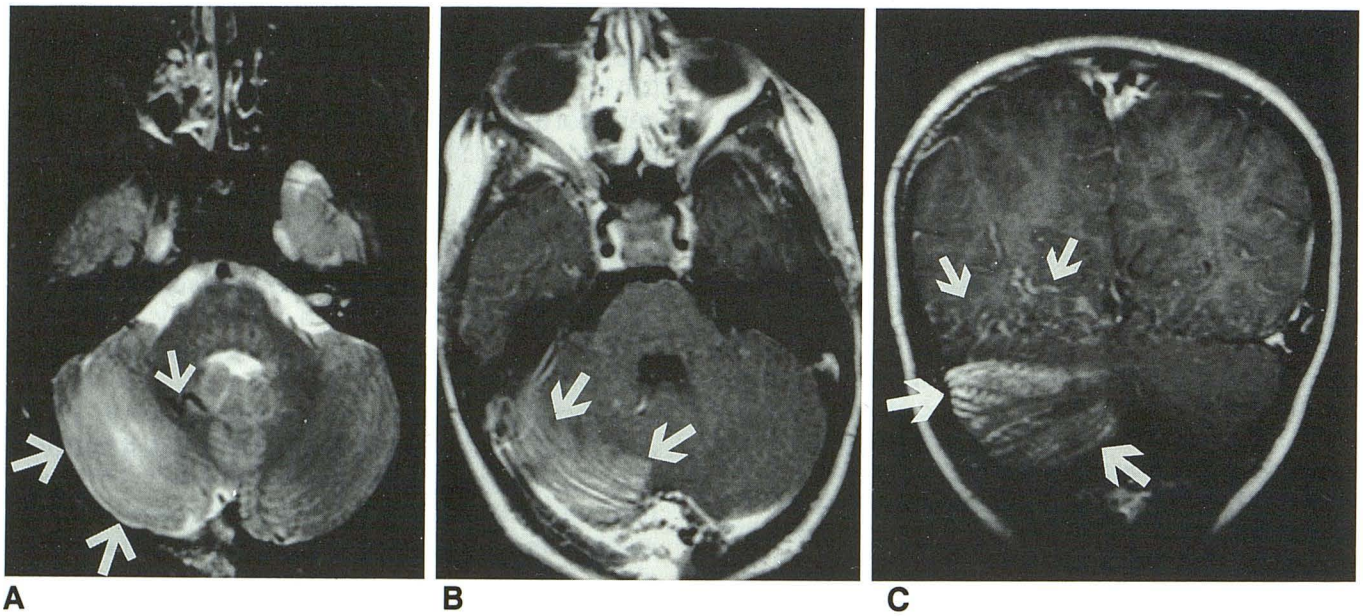

Fig. 2. Axial (*A*) T2-weighted MR (2000/80/2 [repetition time/echo time/excitations]) shows right cerebellar hemispheric high intensities (*large arrows*) and deeper vascular flow signal void (*small arrow*). Axial (*B*) and coronal (*C*) gadolinium-enhanced T1-weighted images (600/15/2) show right cerebellar enhancement (*large arrows*) and right temporooccipital cerebral enhancement (*small arrows*).

dysplastic or thrombosed superficial cortical veins with shunting and opacification of dilated medullary and deep veins (16, 17). Other rare findings in Sturge-Weber syndrome include migrational anomalies (pachygyria and polymicrogyria) underlying the leptomeningeal vascular anomaly (10, 18).

The patient described in this report demonstrated many of the characteristic clinical and imaging findings of Sturge-Weber syndrome. In

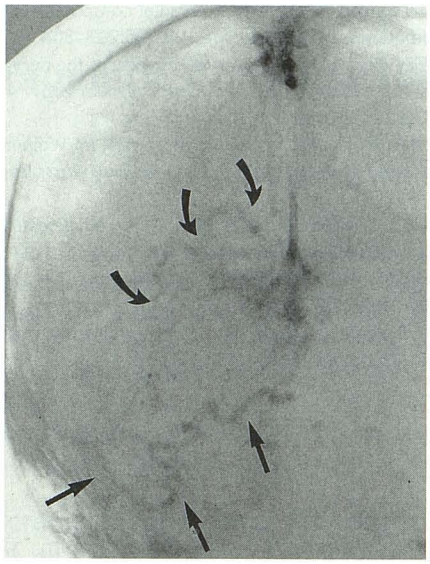

Fig. 3. Frontal subtracted projection of the venous phase of the left vertebral angiogram opacifies the anomalous right cerebellar (*straight arrows*) and temporooccipital cerebral (*curved arrows*) venous structures.

addition, there was the unusual finding of cerebellar involvement demonstrated by CT, MR, and angiography (Figs 1–3) without associated symptoms or signs specific to that involvement. Although several sources acknowledge the rare occurrence of posterior fossa Sturge-Weber syndrome, there are no specific descriptions of the clinical or imaging findings of this involvement (19–22). According to embryonic theory (22, 23), combined facial and pial vascular anomalies result from architectonic persistence of primordial sinusoidal vascular channels normally present only from the fourth to the eighth gestational weeks. During that period, the ectoderm destined to become the skin of the upper face overlies that portion of the dorsal neural tube destined to form the occipital and adjacent lobes of the cerebral hemispheres. Normally, with growth of the cerebrum, the superficial and deep vascular systems are widely separated.

The persistence of intervening primordial vascular channels may result in the association of facial nevi frontally, along the ophthalmic division of the trigeminal nerve, with leptomeningeal vascular anomalies in the occipital cerebrum; the association of maxillary division nevi with parietal anomalies; and the association of nevi along all three trigeminal divisions with cerebral hemispheric involvement. We speculate that occasionally facial nevi in the trigeminal distribution may be associated with cerebellar leptomeningeal vascular anomalies because of the persistence of primordial channels between that portion of the ectoderm forming the face and that portion of the underlying ventral neural tube that forms the rhombencephalon.

The imaging findings for cerebellar involvement in Sturge-Weber syndrome in this case are to be distinguished from meningioangiomatosis (23) and Lhermitte-Duclos syndrome (24). Meningioangiomatosis occurs in neurofibromatosis and is a dysplastic meningeal and vascular proliferation with infiltration of brain. Imaging demonstrates calcification and irregular enhancement. Lhermitte-Duclos syndrome is a dysplastic hypertrophy of cerebellar cortex. On imaging there is thickening of the cortex with a striated or laminated appearance and occasional calcification but no abnormal enhancement.

Acknowledgments

We thank Virginia Grove for manuscript preparation and Don Sucher for photography.

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