Sturge-Weber Syndrome: Study of Eight Patients and Review of Literature

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Citation

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Abstract

Introduction:
Sturge-Weber syndrome (SWS) is a rare congenital sporadic disease with neuro-ocular and cutaneous vascular findings. Clinically, the full-blown condition consists of a facial port-wine stain (PWS) involving the V1 facial trigeminal skin area, alone or in combination with V2 and V3PWS, seizures and ocular abnormalities (glaucoma and choroidal angioma). Radiologically, a leptomeningeal (pial) capillary and venous malformation, mostly located in the parieto-occipital area, cerebral atrophy and calcifications are demonstrated.

Methods:
A retrospective review of the records of patients diagnosed of SWS (facial nevus flammeus at least over the first branch of the trigeminal nerve and ipsilateral leptomeningeal angioma) was performed. Eight patients with Sturge-Weber studied.

Results:
The nevus flammeus was unilateral in 7 patients (twice as often on left side) and bilateral in 1 patient and in one patient on the same side of the leg. All of these patients had cerebral lesions. Seizures, most of which were focal, were present in 6 patients (75%) usually contralateral to the nevus flammeus. Seizure presentation coincided with febrile episodes in none of these patients. Total seizure control was obtained in all patients. Abnormality of the cerebral parenchyma can be detected from birth in some patients and has a progressive character; at the same time, progressive atrophy and parenchymatous hyper density of the affected hemisphere is evident, as well as a decrease in arterial size, especially during the first decade of life. No relationship exists between the size of the facial nevus flammeus or its unilateral or bilateral location and clinical neurologic impairment. Conversely, a direct relationship exists between greater anatomic manifestations (i.e., atrophy, calcification) in the involved hemisphere when the lesion is unilateral as well as the presence of leptomeningeal angiomatosis in both cerebral hemispheres in patients with bilateral facial nevus flammeus and the severity of clinical disorders.

Discussion:
Early evaluation of newborns with nevus flammeus affecting the first branch of the trigeminal nerve must be done, including an MR with contrast and an ophthalmological exam.

References

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