Pediatric Neurology Part I: Chapter 37. Sturge-Weber syndrome (Handbook of Clinical Neurology)

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Sturge–Weber syndrome (SWS) is a rare sporadic neurocutaneous syndrome defined by the association of a facial capillary malformation in the ophthalmic distribution of the trigeminal nerve, with ipsilateral vascular glaucoma and vascular malformation of the eye, and a leptomeningeal angioma. SWS is suspected at birth in the presence of facial angioma in the trigeminal nerve area. MRI with gadolinium enhancement and pondered T1, T2, FLAIR and diffusion sequences is today the technique of choice to visualize the leptomeningeal angioma or to suspect it by indirect signs, even before the development of neurological signs, from the first months of life. The prognosis of SWS with leptomeningeal angioma is related to the severity of neurological signs that are absent at birth and develop later in life (epilepsy, hemiparesis, and mental delay). Seizures are usually the presenting neurological symptom. Status epilepticus might inaugurate the epilepsy and remains frequent in infancy. Repetitive seizures are thought to increase the atrophy of brain tissue in regard to the leptomeningeal angioma. Preventive presymptomatic treatment with antiepileptic drugs is often recommended, and parents are trained to use rescue benzodiazepines in case of seizures. After epilepsy onset, in patients intractable to antiepileptic drugs, surgery should be considered.

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