HAEMANGIOMAS AND VASCULAR MALFORMATIONS

STURGE-WEBER SYNDROME: ABOUT A CASE

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Background: Sturge-Weber syndrome (SWS) is a neurocutaneous syndrome characterized by angiomas involving the face, choroid, and leptomeninges. The facial capillary vascular malformation is also known as "port wine stain" or "nevus flammeus" and usually is seen in the territory of the trigeminal nerve. The early diagnosis and prompt treatment may reduce the incidence of neurologic sequelae, and may prevent irreversible blindness.

Observation: We report a case of a 32-year-old man, who was treated for epilepsy for 29 years with a developmental delay and mental retardation. A large, purple, hemangiomatous lesion was noted over left half of the face in the distribution of the ophthalmic and maxillary divisions of the trigeminal nerve. Ophtalmologic examination found a conjonctival hemangioma and the MRI of the scalp objectived a cerebral atrophy. In view of the constellation of findings, a diagnosis of SWS was made.

Key message: The port wine stain may remain static in extent but can undergo progressive hypertrophy, darken and become nodular in up to 65% of the patients by the fifth decade. About 8% of the individuals may have an underlying SWS and the association is up to 78% if the lesion involves the entire V1 distribution. The leptomeningeal angiomas cause vascular steal and cortical ischemia leading to the cerebral atrophy and/or dystrophic calcifications. Glaucoma can affect 30-70% of patients and ipsilateral eye is mostly affected. Other ocular abnormalities could be choroidal, conjunctival hemangiomas and heterochromia of the iris. Treatment is symptomatic with antiepileptics, antiglaucoma drugs and laser therapy for portwine stain. Low dose Aspirin has been studied in the prevention of stroke like episodes and seizures. Surgical intervention is reserved for patients with refractory seizures and uncontrolled glaucoma.