Sturge–Weber syndrome clinical features

Often associated with port-wine stains of the face, glaucoma, seizures, mental retardation, and ipsilateral leptomeningeal angioma (cerebral malformations and tumors).

Cardinal features:

a) localized cerebral cortical atrophy and calcifications (especially cortical layers 2 and 3, with a predilection for the occipital lobes):

● calcifications appear as curvilinear double parallel lines (“tram-tracking”) on plain x-rays

● cortical atrophy usually causes contralateral hemiparesis, hemiatrophy, and homonymous hemianopia (with occipital lobe involvement)

b) ipsilateral port-wine facial nevus (nevus flammeus) usually in distribution of 1st division of trigeminal nerve (rarely bilateral) other findings that may be present:

a) ipsilateral exophthalmos and/or glaucoma, coloboma of the iris

b) oculomeningeal capillary hemangioma

c) convulsive seizures: contralateral to the facial nevus and cortical atrophy. Present in most patients starting in infancy

d) retinal angiomas

Epilepsy in Sturge-Weber syndrome

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