

A Young Female with Left Sided Focal Seizure

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Fig. 1 : Portwine stain in the region of ophthalmic and maxillary division of trigeminal nerve.

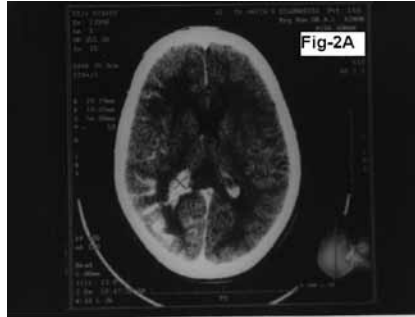


Fig. 2A : Gyral calcification in right parieto-occipital lobe

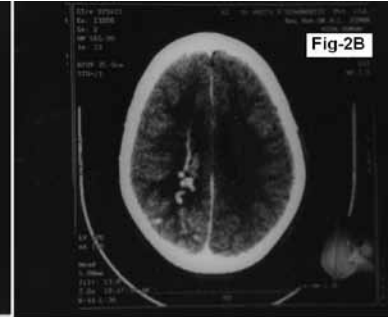


Fig. 2B : Dilated and tortuous vessels in the ventricular system with contrast enhancement.

A 23 years old female presented with recurrent episode of left hemiparesis for 6 months. Each episode lasted for 10 to 20 min at interval of 15 to 20 days. These episodes of weakness were not associated with any seizure like activity, unconsciousness or other focal neurological deficit. Three days back patient had recurrent left sided focal motor seizure with secondary generalization. She had past history of seizure at age of six months, since then she was asymptomatic till six month back. Physical examination revealed left sided face arm leg weakness and a left extensor planter response. A Portwine stain present on the right side of face in the region of ophthalmic and maxillary division of trigeminal nerve (Fig. 1). The examination of right eye revealed megalocornea, deep anterior chamber, increased intraocular pressure, glaucomatous optic atrophy with 90% cupping and choroidal hemangiomas. Left eye was normal on examination. Baseline investigation including hematology, serum biochemical and coagulation profile were within normal limits. Computerized tomography (CT) Scan of cranium showed gyral calcification in parieto-occipital lobe of right hemisphere (Trolley-Track appearance), dilated and tortuous vessels in ventricular system with contrast enhancement (Fig. 2A & 2B). The case was diagnosed as Sturge Weber syndrome.

Sturge Weber syndrome is characterized by facial cutaneous angioma (Portwine stain typically involving the ophthalmic division of trigeminal nerve) and an associated ipsilateral brain angioma with gyral calcification in parieto-occipital lobe. Our case was associated with glaucoma of ipsilateral eye. Principle neurological manifestations are epileptic seizure and focal neurological deficit both. Some patients may have refractory seizure, while others have long seizure free intervals without medications. Some patient may present with repeated episode of focal weakness similar to transient ischemic attacks. Generally the more extensive the intracranial lesion, the more difficult is it to fully control the seizure with medications.¹ Sujansky et al² reported 50% patients fully controlled and additional 35-40% had partial control of seizure with medication. In refractory seizures, hemispherectomy, resection of the area predominately affected or corpus callosum section sometime improves seizure control.

References

1. Bodensteiner JB, Roach ES. Sturge-Weber Syndrome: Introduction and Overview. In: Bodensteiner JB, Roach ES, eds. Sturge-Weber Syndrome. Sturge Weber Foundation. Mt Freedom, New Jersey. 1999.
2. Sujansky E, Conradi S. Outcome of Sturge Weber Syndrome in 52 adults. Am J Med Genet 1995;57:35-45.

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