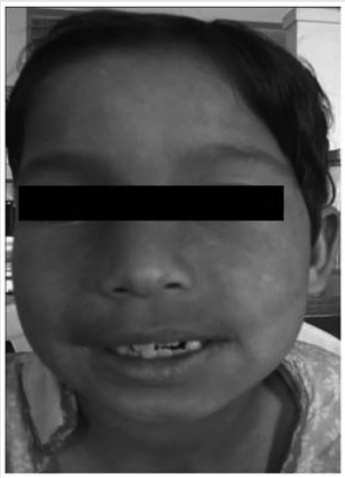
SPOT DIAGNOSIS (IMAGE GALLERY)



BILATERAL FACIAL NEVUS AND TRANSIENT RECURRENT HEMIPARESIS

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An 8 year old female born of non-consanguineous marriage presented with weakness of left side upper and lower limb, generalized headache, bodyache for one day. There was also one episode of loss of consciousness for few minutes. Past medical history revealed that patient had transient weakness of left side of body and convulsive disorder since the age of 8 months for which she was under treatment. Child also had delayed developmental milestones. On examination a large port-wine stain involving both sides of face along the ophthalmic and mandibular divisions of trigeminal nerve and neck region was present. (Figure 1) On ophthalmological evaluation, she had bilateral megalocornea with vertical diameter of 14mm. Neuro retinal rim was unhealthy and nasal shifting of vessels was seen. There was hypotonia on left side of body and deep tendon reflexes were decreased over left side of the body. Other systemic examination was normal. CT brain showed right cerebral hemisphere and left occipital atrophy with linear and clumped gyral calcification. MRI brain showed right hemispheric atrophy with diffuse gyral enhancement involving right cerebral hemisphere & left occipital lobe, enhancing choroid in both eyeballs suggestive of choroidal angioma.

What is the diagnosis?

Sturge-Weber syndrome (SWS). It is a rare sporadically occurring, congenital neurocutaneous disorder with an estimated frequency of approximately 1 per 50,000 live births. (1) This syndrome is characterized by intracranial leptomeningeal vascular angioma with unilateral facial nevus but in our patient, port wine stain involved both sides of the face and extended up to the neck. Other clinical findings associated with SWS are seizures, glaucoma, hemiparesis and mental retardation. About 25 to 56 percent of patients experience recurrent episodes of paroxysmal focal neurological deficits in form of transient hemiparesis which may be due to vascular ischemia or may be post-ictal in origin. (2) The radiological hallmark is "Tram-line" or "Gyriform" calcification. SWS is believed to be caused by residual embryonal blood vessels and their secondary effects on the surrounding brain tissue leading to hypoxia, ischemia, venous occlusion, thrombosis, infraction or vasomotor phenomenon. (3,4) The management of SWS is symptomatic and aimed at controlling of seizures, preventing stroke like episodes, monitoring for glaucoma and using laser therapy for cutaneous capillary malformation. (1)

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