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Sturge Weber Syndrome with Bilateral Portwine Stain

Medical Science

KEYWORDS : Sturge weber syndrome ,bilateral port-wine stain, seizures, cerebral atrophy

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ABSTRACT

Sturge weber syndrome is a sporadic vascular disorder consisting of unilateral facial capillary malformation (port-wine stain), abnormal blood vessels of the brain (lepto meningeal angioma) and abnormal blood vessels of the eye leading to glaucoma. Incidence is approximately 1 in 50,0001. Here we present a case of this rare syndrome with a varient of bilateral port-wine stain and with seizures on contralateral side to the side of involvement of ophthalmic division of trigeminal nerve.

Introduction:
The Sturge–Weber syndrome, also known as encephalofacial angiomatosis, is a neurocutaneous disorder that occurs as a sporadic congenital condition; it is characterized by unilateral port-wine stain that affects the skin in the distribution of the ophthalmic branch of the trigeminal nerve and is associated with venous-capillary abnormalities of the leptomeninges and the eye[2].

Case report:
A 5.5month old male child of Indian origin, born of a non-consanguineous marriage presented to our hospital with chief complaints of seizures since four days.

These seizures were tonic clonic movements occurring in left upper limb and left side of face involving the eyelid but not in left leg. Episode lasted for about 2minute followed by loss of consciousness for 30 minutes. The child completely regained his consciousness thereafter. These episodes would occur multiple times a day. Two oral antiepileptic drugs phenobarbitone and sodium valproate had to be started to control the seizure activity. Patient did not have any history suggestive of head trauma, birth asphyxia or meningitis.

Family history is unremarkable and both parents and elder siblings are healthy.

On physical examination, port-wine stain is present along ophthalmic division of trigeminal nerve on right side of face and along the maxillary division on left side of face. Development age of the child is 5 months with no other significant neurological or systemic examination. Opthalmological examination tested negative for glaucoma presently.

Routine blood and CSF investigations were normal. MRI suggestive of changes of diffuse cerebral atrophy with gyriform abnormal signals in the right frontotempoparietal region.no calcifactions were present.

Discussion:
Sturge weber syndrome includes unilateral localisation of aberrant vasculature in the facial skin, eyes and meninges. The surface capillary malformations are usually present at birth and seldom progress. Seizures most commonly begin between 2 to 7 months of age and are grand mal in type. The degree of CNS involvement is variable with 30% having paresis and approximately 83%having seizures, 39%have normal intelligence. A poor prognosis of cognitive development is predicted by the number of seizures, an early age of onset, a poor response to treatment, bilateral cerebral involvement or severe unilateral lesion.

Facial PWSs are a far more common occurrence than Sturge weber syndrome, with an incidence of approximately 1 in 300 for the former and an estimated incidence of between 1 in 20 000 and 1 in 50 000 for the latter[3].

Aggressive control of seizures is recommended. Stroke like episodes are common. Focal resection may be needed.

Glaucoma develops before 2years of age if tissues of the anterior chamber angle are affected.

Cerebral calcification is usually not evident in radiography until later infancy [4].

Fig: port-wine stain present along ophthalmic division of trigeminal nerve on right side of face and along the maxillary division on left side of face

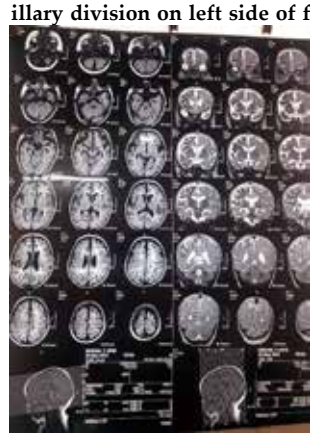

Fig: MRI suggestive of changes of diffuse cerebral atrophy with gyriform abnormal signals in the right frontotempoparietal region.

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