Case report

Sturge-Weber syndrome presenting with intractable seizures

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Abstract:

Sturge-Weber syndrome (SWS) also known as encephalotrigeminal angiomatosis is one of the many neurocutaneous syndromes consisting of port wine stain (facial nevus flammeus), congenital glaucoma and cortical calcification with anomalous leptomeningeal venous plexus. A child with intractable seizures, facial port wine stain and occipital cortical calcification is presented in this case report.

Keywords: Childhood seizures, neurocutaneous syndrome, port wine nevus, Sturge-Weber syndrome

Introduction

Sturge-Weber syndrome, also known as encephalotrigeminal angiomatosis is one of the many neurocutaneous syndromes consisting of port wine stain (facial nevus), congenital glaucoma and cortical calcification with anomalous leptomeningeal venous plexus.[1,2] Other features involving the dermatologic and oral systems are also present. It is a rare syndrome that presents with facial port wine stain or more commonly with seizures.

Case report

A 4-year old male child was brought to the emergency with history of intractable seizures that recurred every hour for the last 24 hours. He had past history of such on and off seizures for the last two years. Family history was negative for any seizure disorders. He had been previously started on phenytoin with poor control of the attacks. There was history of gross developmental, school and sociological compromise due to the recurrent attacks. On examination the child was in status epilepticus with a forehead port wine stain over the left first trigeminal division (Fig. 1). The pupils were equal bilaterally and reactive to light. There was clenching of the teeth with Glasgow coma score of 6/15. Further examination showed gingival hyperplasia with staining of the teeth suggestive of chronic high phenytoin toxicity.

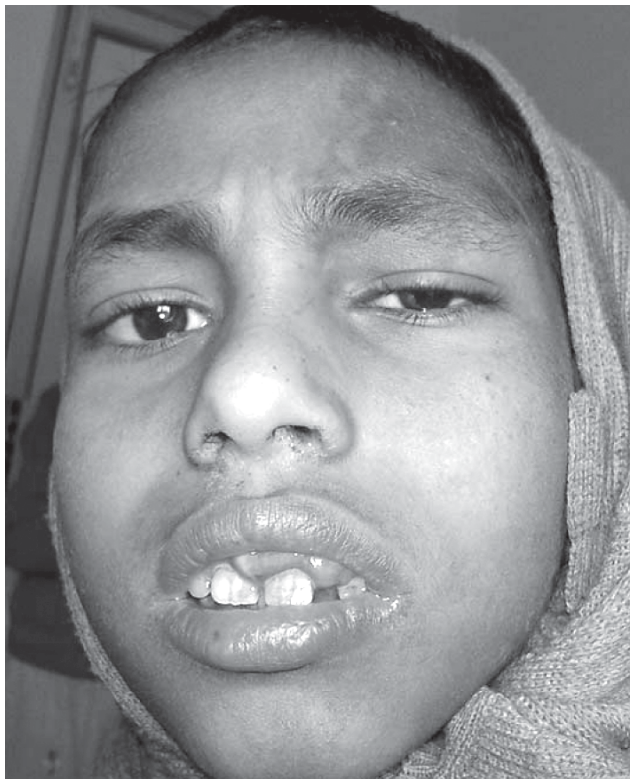

Fig. 1: Distribution of the port wine stain over the left first trigeminal division and gross gingival hyperplasia secondary to phenytoin toxicity.

Seizures were controlled with intravenous diazepam bolus and then he was maintained on valproic infusion (30 mg/kg weight loading followed by 20 mg/kg weight as maintenance). Routine bloods and liver function tests were normal. Electroencephalogram (EEG) was done which showed features of status epilepticus. Computed Tomogram (CT) showed a large serpigenous calcification in the left occipital lobe with no perilesional edema (Fig. 2). The Hounsfield unit was suggestive of calcification. Interictal EEG on the third day was definitive for generalized tonic clonic seizures arising from the left occipital area. With the triad of childhood seizures, port wine facial stain and occipital cortical calcification he was diagnosed as a case of SWS. On the third day he was started and maintained on oral valporate. He has since then no recurrence of seizure for the last three months of follow up.

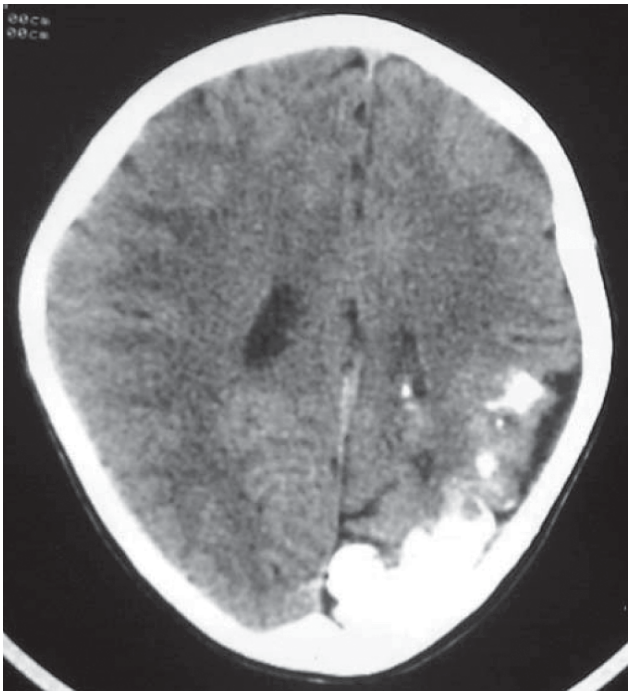

Fig. 2: Left occipital cortical serpigenous calcification- the same side as the dermal nevus.

Discussion

Sturge-Weber Syndrome is one of the many neurocutaneous syndromes consisting of port wine stain (facial nevus), congenital glaucoma and cortical calcification with anomalous leptomeningeal venous plexus.[1,2] This syndrome can manifest as intractable seizures, headaches, stroke-like symptoms, mental retardation, learning and behavioral problems or hemiparesis.[3] Bupthalmos and glaucoma can lead to progressive visual loss with blindness.[4] Vascular steal phenomenon due to the cortical angioma results in cortical ischemia which is aggravated by recurrent seizures and other vascular events leading to gliosis, atrophy and calcification.[5,6] The Roach scale is used to classify whether the SWS is complete or incomplete (Table-1).[7]

Table 1: Roach scale classification of SWS.

Type-I	Both facial and leptomeningeal angiomas; may have glaucoma
Type II	Facial angioma alone (no CNS involvement); may have glaucoma
Type III	Isolated LA; usually no glaucoma

CT or magnetic resonance imaging with contrast helps to confirm the brain involvement in suspected cases of SWS. Seizures are present in almost 75% of cases and early medical intervention is must to prevent the development of neurological deficits and impairment. Medical management is sufficient in most of the cases although some may need lobectomy to control their seizures. Early surgery leads to good control of the seizures with less satisfactory improvement in hemiparesis or intellectual deficits. Poor medical control, early age of onset of seizures and bilateral cerebral involvement are indicators of poor prognosis.[3] Counseling regarding the benign nature of the disease along with the education for the need to control the seizures is a must for the parents and caregivers to help overcome their problems and improve the overall treatment outcome.

Conclusions

Sturge-Weber Syndrome is one of the many neurocutaneous syndromes which may manifest as intractable seizures, headaches, stroke like symptoms, mental retardation, learning and behavioral problems. It is a benign disease where seizure control remains the mainstay of its treatment. With proper drug dosage and patient counseling, good long term results can be achieved in these cases.

References

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