Sturge-Weber Syndrome: An Illustration

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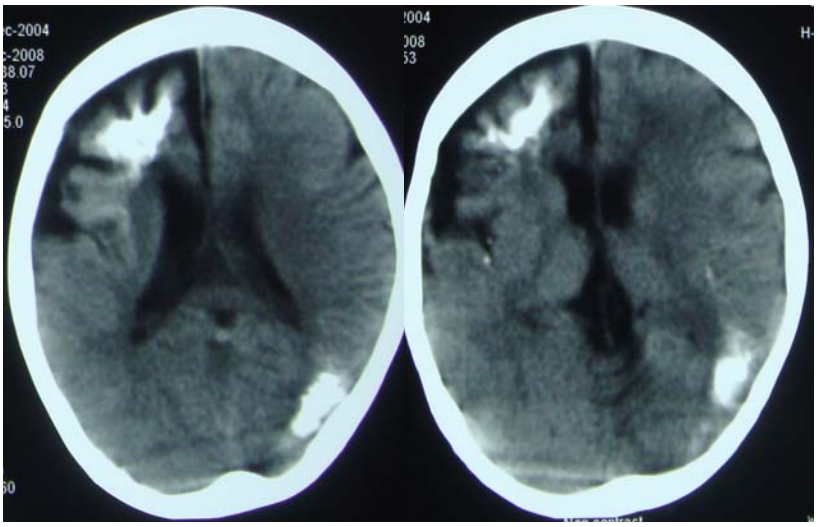

Figure 1. Computerized tomographic image showing calcification of cerebral cortex on right frontal and left parieto occipital region with cortical atrophy on right frontal region.

Sturge–Weber syndrome (SWS) is a neurocutaneous syndrome commonly associated with epilepsy and port wine nevus. Its prevalence is about 1 per 50,000 live births, men and women are equally affected and there is no racial bias. Familial cases are rare and thus are thought to be due to somatic mutation. The identification of milder forms has recently led to an increase in incidence.

Here we present a case of 5 year old boy with right sided facial port-wine nevus, who had Generalized Tonic Clonic seizure associated with psychomotor disturbance . The symptoms were gradually worsening since 2 years back. He was not evaluated by any specialist doctor and was not under any medication. The frequency of seizure was increasing up to 3 times per week, sometimes lasting for more than 10 minutes. Plain and enhanced CT head was advised which showed huge calcified lesion in right frontal and left parietal lobe with underlying cortical atrophy **(Figure 1)**. The scalp EEG showed generalized pattern waves without obvious localization of epileptogenic zone.

Childhood seizure disorder is one of the common medical problems in our society as anywhere else in the world. However, due to inadequate and improper evaluation we often miss the underlying cause of seizure such as SWS which is a rare entity. The case that we reported was thought to be an ordinary case of childhood epilepsy and was further evaluated as usually with CT of head and EEG. His clinical features and CT findings strongly suggested SWS.

The syndrome is associated with epilepsy in 75–90% of the cases and are resistant to medical treatment in almost 60% of cases. Therefore, they may require some sort of epilepsy surgery. However, the localization of epileptic foci may be very difficult.[1] It has been reported that the onset of seizures in the first year of life is correlated with a higher rate of resistance to medical treatment.[2] Approximately half of the children with SWS have frank mental retardation. Psychomotor delay is more common in children whose seizures begin before the age of 2 years or who have seizures not controlled with antiepileptic drugs .

A common way of presentation is attention deficits and hyperactivity. Imaging is important in SWS mainly to determine the extent of cerebral pathology. Findings on CT and MRI include superficial calcification, atrophy and leptomeningeal angiomatosis. In some cases arterial and venous MR angiography may be helpful to locate vascular anomalies or to demonstrate a prominent deep collateral venous system and/or lack of superficial cortical veins.[3]

References

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