

A Case Report of Sturge-Weber Syndrome

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Abstract: Sturge-Weber syndrome is one of the neurocutaneous syndromes and was first described by Sturge in 1879. Weber demonstrated the characteristic intracranial calcifications in 1929. It is a rare, nonfamilial disease that is characterized by the following features: port-wine stain (nevus flammeus), leptomeningeal angiomas, choroidal angioma, buphthalmos, intracranial calcification, cerebral atrophy, mental retardation, glaucoma, seizures, hemiparesis, and hemiatrophy. We report a rare case of Sturge-Weber syndrome in a 6-month-old baby.

I. Materials and methods

A 6-month-old female baby came to Bapuji hospital with chief complaints of seizures since 4 months and port wine stain on the right side of face since birth.

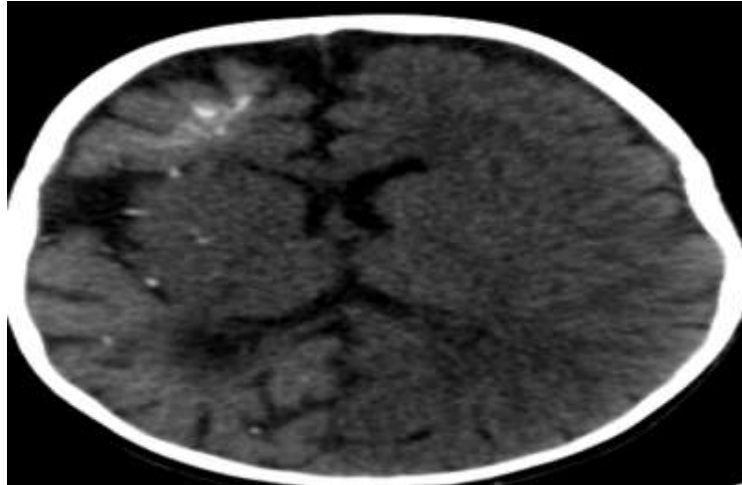
Non-contrast computed tomography of brain was done in Bapuji hospital, Radio diagnosis department using Toshiba Activion 16 slice CT machine.



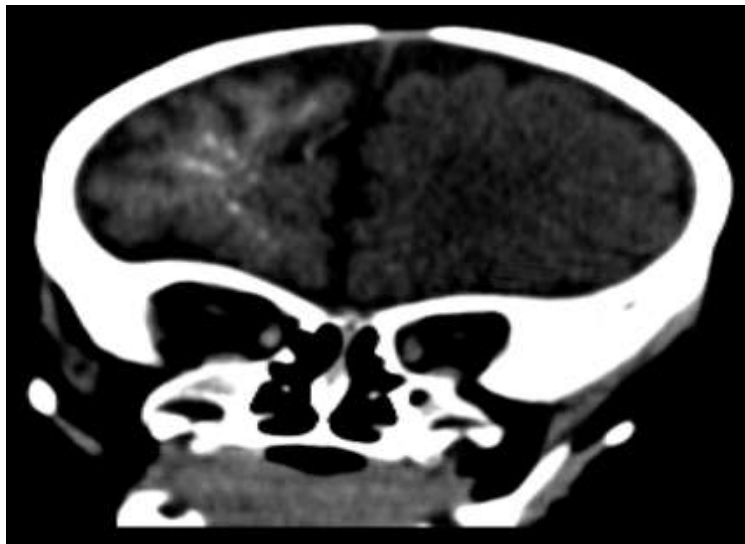
Photograph of baby showing port wine stain on right side of forehead

II. Results

Non-contrast computed tomography showed diffuse gyriform & subcortical calcification. And also showed diffuse cortical atrophy in right cerebral hemisphere with prominent calcified vessels on ipsilateral subarachnoid space with shift of falx and 3rd ventricle towards right side. Cerebellar hemispheres and brain stem are normal.



Axial NECT shows diffuse gyriform& subcortical calcification with diffuse cortical atrophy in right cerebral hemisphere.



Coronal NECT shows diffuse gyriform& subcortical calcification with diffuse cortical atrophy in right cerebral hemisphere.

III. Conclusion

Sturge-Weber Syndrome is usually a sporadic congenital (but not inherited) malformation in which fetal cortical veins fail to develop normally. Imaging features are sequelae of progressive venous occlusion and chronic venous ischemia. Clinically they usually present with "Port-wine" stain, seizures, hemiparesis. Rare: 1:20,000-50,000. Increased extent of lobar involvement and atrophy leading to increased likelihood of seizures. NECT imaging shows Gyral/subcortical white matter (WM) calcifications. Usually progressive calcifications are noted posterior to anterior. Late changes include cerebral atrophy, hyperpneumatization of paranasal sinuses and thick diploe. CECT shows serpentine leptomeningeal enhancement. [1]

Reference

[1] Anne G. Osborn, Karen L. Salzman, and Miral D. Jhaveri. *Diagnostic Imaging: Brain* 3rd ed. (Elsevier, Philadelphia, 2016)