



STURGE WEBER SYNDROME WITH EARLY CORTICAL CALCIFICATION

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ABSTRACT

A seven and a half month old female baby with facial mole, developmental delay and history of fits was referred for CT scan of the brain with clinical suspicion of Sturge weber syndrome. CT scan showed right cerebral hemispheric atrophy with cortical calcifications. Contrast enhanced MRI of the brain showed classical leptomeningeal enhancement in the right cerebral hemisphere along with prominence of the choroid plexus of the right lateral ventricle thus confirming the diagnosis of Sturge weber syndrome. In patients with Sturge weber syndrome, calcifications are rarely demonstrable before 2 years of age. The presence of early calcification signifies considerable parenchymal atrophy and carries poor prognosis in terms of developmental mile stones. Our patient presented with early calcifications and developmental delay. The purpose of presenting this case is to emphasize the importance of early calcification and its significance in the development of the infant.

INTRODUCTION

Sturge weber syndrome (encephalo trigeminal angiomatosis) is one of the commonest sporadic neurocutaneous syndromes characterized by angiomas in the distribution of the trigeminal nerve as well as in the cranial cavity [1]. The patients classically present with epilepsy, facial portwine stain, hemiparesis, mental retardation, developmental delay, glaucoma, choroidal angioma and homonymous hemianopia [2]. The syndrome shows classical imaging manifestations and radiology plays a major role in its diagnosis and prognostication.

CASE HISTORY

A seven and a half month old female baby was brought to the paediatric OPD with history of fits and congenital facial mole. Physical examination revealed a large port wine stain along the distribution of the ophthalmic and maxillary divisions of the right trigeminal nerve (Fig.1). The developmental milestones were delayed with lack of head control and inability to sit with support.

The central nervous system examination was unremarkable and the laboratory investigations were within normal limits. The baby was clinically diagnosed to have Sturge weber syndrome and referred for CT scan of the brain.

CT scan of the brain showed atrophy of the right cerebral hemisphere with hyperdense calcifications (CT attenuation of 96-100HU) in the cortex and subcortical white matter (Fig.2). The extra axial spaces in the right side of the cranial cavity were widened.

MRI of the brain was performed. It showed atrophy of the right cerebral hemisphere with widened extra axial spaces. Parenchymal signal intensity alterations were not observed on plain MR sequences. After administration of i.v. contrast, there was striking supratentorial pial enhancement in the right cerebral hemisphere (Figure 3). The choroid plexus of the right lateral ventricle was prominent and showed intense enhancement (Figure 3). There were a few prominent deep medullary veins. The calvarium and paranasal sinuses were normal. The eye



balls were normal. There were no choroidal angiomas. MR venography revealed reduced number of cortical veins over the right cerebral hemisphere. There was no intra cranial bleed. Based on the clinical features and imaging findings, the diagnosis of Sturge weber syndrome was made. As this is a classical case of sturge weber syndrome with rare feature of early cortical calcification, it is presented here.

DISCUSSION

Sturge weber syndrome was described by Sturge in 1879. The radiological manifestations were described by weber in 1922 [1]. It is one of the commonest sporadic phacomatoses characterized by multiple angiomas of the skin, eye and the meninges. It occurs due to the failure of the normal development of the cortical veins with formation of angiomas. The deep cerebral veins and the medullary veins become prominent and form collaterals [14]. As the duration of the disease increases, the deep veins may also get occluded resulting in ischemia.

Imaging features are the sequelae of progressive venous occlusion and chronic venous ischemia. The angiomas are seen ipsilateral to the side of the portwine stain (nevus flammeus). Portwine stain is present in about 98% cases of Sturge weber syndrome. The neurological features and pial angiomas are observed only in those cases which have the stain over the upper face and the eye lids, in the distribution of the ophthalmic division of the trigeminal nerve [2].

Plain radiograph of the skull reveals parallel curvilinear calcifications giving tram track appearance. The calcifications are not demonstrable until 2 years of age [12]. In our case, the hyperdense early calcifications were demonstrated at the age of less than 1 year itself. Presence of early calcifications signifies considerable early parenchymal atrophy and carries poor prognosis [12]. Non contrast CT detects the cortical calcifications better than the MRI. Hence non contrast CT, followed by contrast enhanced MRI help in accurate diagnosis of all the manifestations. FLAIR – contrast MRI improves the conspicuity of the leptomeningeal angiomatosis.

Contrast enhanced MRI is the imaging modality of choice to evaluate the complete spectrum of the syndrome. Post contrast T1W MR Images reveal intense supratentorial pial enhancement on the side of the portwine stain which is diagnostic of the syndrome [3]. Pial enhancement is classically seen in the occipital and posterior parietal cortex. However, frontal lobe, temporal lobe and the brain stem may also show enhancement. Enhancement may also be seen in the periorbital soft tissues, bony orbit, choroidal angioma and the frontal bone diploic space. Infratentorial pial enhancement is unusual.

M.E. Adams et al have described infratentorial enhancement in 10% of the cases in their study [1]. They have also described unusual imaging features like bilateral pial enhancement, brain stem enhancement, ipsilateral cerebral pial enhancement & contra lateral cerebellar pial enhancement etc. However in our case infratentorial pial

enhancement was not seen. Griffith et al [7] studied the relation between the choroid plexus size and the leptomeningeal enhancement. They found that the ipsilateral choroid plexus enlargement & intense enhancement were constantly associated with extensive leptomeningeal enhancement. In our case also there was significant ipsilateral choroid plexus enlargement and intense enhancement.

Paul D Griffiths et al [5] studied fifteen children with Sturge weber syndrome for abnormal ocular enhancement. They correlated the fundoscopy findings, meningeal enhancement, ocular enhancement and glaucoma. They found that the likelihood of ocular enhancement was more with bilateral disease, extensive facial portwine stain or glaucoma. No correlation was observed between ocular enhancement and the extent of the hemispheric disease. Ocular hemangiomas, choroidal and retinal detachments were observed in patients with severe ocular disease [6]. In our case no abnormal ocular enhancement was seen in spite of large facial portwine stain and significant right hemispheric involvement.

The diagnosis of congenital glaucoma on imaging modalities is not widely described. Choudhary et al [8] observed that the enlargement of the anterior chamber in the absence of trauma and ipsilateral optic nerve thinning might represent congenital glaucoma. However, there was no evidence of optic nerve atrophy / congenital glaucoma in our patient.

George et al [10] have described accelerated myelination in a case of Sturge weber syndrome due to cerebral transient hyperperfusion. However, in our case no such accelerated myelination was observed. Migrational anomalies, cortical dysplasias and gyral abnormalities like pachygyria might be associated with Sturge weber syndrome [13]. There was no evidence of such abnormalities in our case.

Oromaxillofacial hypertrophy and sclerosis were described by Linn et al [11] in one patient with Sturge weber syndrome. However, no such finding was observed in our case. Very rarely, lack of leptomeningeal enhancement (burnt out disease) with progressive atrophy and cortical calcification might be observed. It might be either due to thrombosis of the feeding vessel [4] or regression of the abnormal leptomeningeal vessels due to lack of capillary run off as a result of severe parenchymal calcification. However our case showed classical leptomeningeal enhancement. Nuclear medicine studies show diploic involvement, progressive glucose hypometabolism and hypoperfusion of the cortex in late phases of the disease [14]. However, nuclear medicine tests were not performed in our case.

Prophylactic anticonvulsants and prophylactic low dose aspirin are prescribed to prevent further parenchymal atrophy [14]. Intractable seizures might require surgeries like callosotomy/ lobectomy/ hemispherectomy. Reduced visual acuity due to diffuse choroidal hemangiomas, exudative retinal detachment and subretinal fluid collection



might be treated by plaque brachytherapy as described by Ramesh et al [9]. The treatment of Sturge weber syndrome is symptomatic with anti-epilepsy medications, physical & mental rehabilitation and neurosurgery. There is scope for interventional radiology with embolization of the angiomas

to prevent epilepsy and further atrophy of the brain. Presence of early calcifications signifies considerable parenchymal damage and requires prophylactic anticonvulsants and low dose aspirin to prevent further damage to the brain.

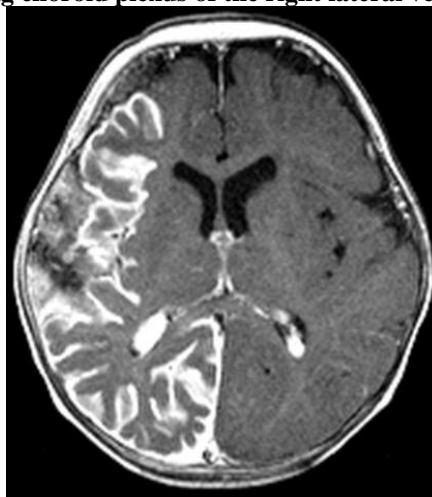
Figure 1. Photograph of the patient showing facial portwine stain along the distribution of the ophthalmic and maxillary divisions of the right trigeminal nerve.



Figure 2. Axial CT image showing atrophy of the right cerebral hemisphere with prominent extra axial spaces and hyper densities in the cortex & subcortical areas representing calcifications.



Figure 3. Axial post contrast T1 weighted MR image showing intense enhancement of the leptomeninges of the right cerebral hemisphere. Note the enlarged intensely enhancing choroid plexus of the right lateral ventricle.



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