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Image Gallery

Sturge Weber Syndrome

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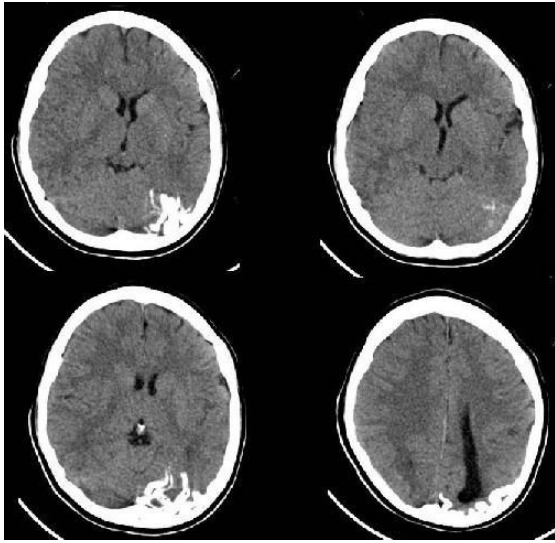
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A 3 year old female child presented with complaints of multiple episodes of seizures since infancy. Patient was already receiving antiepileptic drugs but compliance was not good. There was history of 3 episodes of focal seizures in last 15 days. On examination there was presence of a port wine stain on the face. On Developmental assessment baby has

developmental delay. Ophthalmological examination revealed an enlarged corneal diameter. There was no significant clinical history of similar illness in any other sibling or any other family member of the patient. Routine investigation like CBC, electrolytes, CRP and blood sugar levels were normal. Metabolic workup was also found to be normal. A CT Scan was done which is shown below.

Fig 1 : Tram Track Calcification is seen on CT Scan.



What is the diagnosis?

- Sturge-Weber syndrome**, also called as **encephalotrigeminal angiomatosis**, is a rare neurological disorder. It is one of the condition where neurocutaneous marker ie port wine stain over the face is seen. Other associated abnormalities may include glaucoma, bupthalmos, Bupthalmos, Conjunctival and episcleral hemangiomas, choroidal hemangiomas and heterochromia iridis. seizures, mental retardation, and ipsilateral leptomeningeal angioma may be present. It is characterized by abnormal blood vessels on the brain surface. Normally, only one side of the brain is affected. And focal seizures if seen are usually seen on the opposite side of port wine stain.

CT scan appearance of sturge-weber syndrome is classically called tram track calcification. Cortical atrophy and enlarged choroid plexus may also be seen on CT Scan¹ Other features on CT scans include ipsilateral cortical atrophy, enlargement of the ipsilateral ventricle, and loss of volume of the ipsilateral cranial cavity³.

Management is symptomatic. Antiepileptic drugs for seizures, laser treatment for port wine stain. Glaucoma can initially be managed by medicine but later may require surgical intervention. A multidisciplinary approach is needed while managing these patients.

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