Tram Track Sign in Sturge-Weber Syndrome Type 3

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Clinical Summary

A 36-year-old male, without any previous comorbidities, presented with new-onset recurrent left focal Seizure with secondary generalization for 48 h. He developed refractory status epilepticus, which was controlled after infusion of anesthetic agents. The MRI brain revealed volume loss in the right parieto-occipital lobe with gyriform cortical hypointensity in T2-WI [arrow in Figure 1a] and susceptibility-WI [arrow in Figure 1b] hyperintensity in PHASE images suggestive of calcification (tram-track calcifications). Postgadolinium T1-WI showed gyriform enhancement [arrow in Figure 1c] in the right parieto-occipital lobe suggesting pial angiomas and an enlarged enhancing right choroid plexus [arrow in Figure 1d]. The imaging features were suggestive of Sturge-Weber syndrome (SWS).[1] In the absence of facial angiomas, and glaucoma on ocular examination, we made the diagnosis of SWS Type 3. Type 3 is the rarest variety of SWS spectrum disorders, which can present in adults for the first time, usually with seizures[2] or migraine.[3] Despite achieving adequate seizure control, the present case succumbed to in-hospital respiratory complications. As there are no neurocutaneous manifestations, and the clinical presentation can be highly variable, from migraine-like attacks with visual aura,[4] to hemiparesis and status epilepticus, a high index of suspicion is required to diagnose SWS type 3, and the diagnosis is usually clinched with the help of characteristic imaging findings.

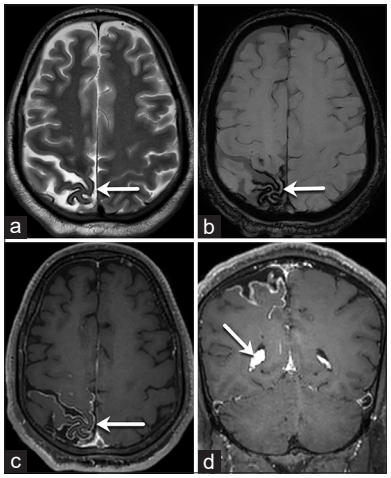

Figure 1: The MRI brain revealed volume loss in the right parieto-occipital lobe with gyriform cortical hypointensity in T2-WI (arrow in a) and susceptibility-WI (arrow in b); hyperintensity in PHASE images (not shown) suggestive of calcification (tram-track calcifications). Postgadolinium T1-WI showed gyriform enhancement (arrow in c) in the right parieto-occipital lobe suggesting pial angiomas and an enlarged enhancing right choroid plexus (arrow in d). The imaging features were suggestive of Sturge-Weber syndrome (SWS)

Commentary or discussion on the imaging findings

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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