Austin Journal of Radiology

Open Access

Austin Publishing Group

Clinical Image

"Tram-Track" In the Brain: A Radiological Feature of Sturge-Weber Syndrome

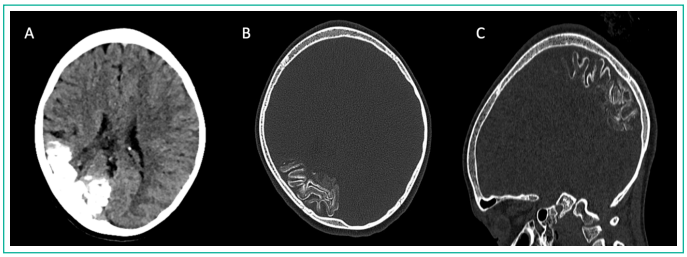
Marrakchi Salma*; El Yousfi Zakia; Hadj Hsain Ihssan; Allali Nazik; Chat Latifa; El Haddad Sihame
Department of Pediatric Radiology, Ibn Sina University Hospital, Rabat, Morocco

***Corresponding author: Marrakchi Salma**
Department of Pediatric Radiology, Ibn Sina University Hospital, Mohamed Ben Abdellah Regragui, Al Irfane, Rabat, Morocco.
Tel: +212650095952
Email: marrakchi.salma@gmail.com

Received: April 27, 2024
Accepted: May 15, 2024
Published: May 22, 2024

Clinical Image

Sturge-Weber Syndrome (SWS), a phakomatosis, is a rare neurocutaneous disorder affecting 1 in 20,000 to 50,000 individuals [1]. First described by Sturge in 1879 then by weber, it is characterized by angiomatosis of the skin, eye, and meninges [2]. Pathophysiologically, it involves somatic mutations in the GNAQ (9q21) gene leading to vascular dysplasia [3]. Clinically, patients exhibit facial port-wine stains, ocular abnormalities, and neurological symptoms. On CT scans, characteristic findings include leptomeningeal enhancement and gyriform calcifications in the cerebral cortex [4]. These calcifications represent chronic ischemia and gliosis [5]. The image below depicts a 7-year-old patient who presents with recurrent seizures. On the non-contrast cerebral CT scan, in axial sections with parenchymal (A), bone (B) window settings, and sagittal reconstruction (C), it demonstrates the typical appearance of gyriform cortical calcifications in the right parieto-occipital lobe, giving rise to a "tram-track" pattern, associated with slight parenchyma volume loss. The patient was subsequently diagnosed as carrying a mutation in the GNAQ gene, confirming the diagnosis of Sturge-Weber Syndrome (SWS).



References

1. Poliner A, Fernandez Faith E, Blieden L, Kelly KM, Metry D. Port-wine Birthmarks: Update on Diagnosis, Risk Assessment for Sturge-Weber Syndrome, and Management. Pediatr Rev. 2022; 43: 507-516.

2. Higueros E, Roe E, Granell E, Baselga E. Sturge-Weber Syndrome: A Review. Síndrome de Sturge-Weber: revisión. Actas Dermosifiliogr. 2017; 108: 407-417.

3. Adams ME, Aylett SE, Squier W, Chong W. A spectrum of unusual neuroimaging findings in patients with suspected Sturge-Weber syndrome. AJNR Am J Neuroradiol. 2009; 30: 276-281.

4. Yeom S, Comi AM. Updates on Sturge-Weber Syndrome. Stroke. 2022; 53: 3769-3779.

5. Sudarsanam A, Ardern-Holmes SL. Sturge-Weber syndrome: from the past to the present. Eur J Paediatr Neurol. 2014; 18: 257-266.

Citation: Marrakchi Salma; El yousfi Zakia; Hadj hsain Ihssan; Allali Nazik; Chat Latifa, et al. "Tram-Track" In the Brain: A Radiological Feature of Sturge-Weber Syndrome. Austin J Radiol. 2024; 11(2): 1234.