

Neurosciences Quiz

Submitted by: Ramachandiran Nandbagopal, MBBS, DM, Abdullah Al-Asmi, MD, FRCPC.

From the Neurology Unit, Department of Medicine, College of Medicine and Health Sciences, Al-Khod, Muscat, Oman.

Address correspondence to: Dr. Ramachandiran Nandbagopal, Consultant Neurologist, Neurology Unit, Department of Medicine, College of Medicine and Health Sciences, PO Box 35, SQU Al-Khod, Zip 123, Muscat, Oman. Tel. +968 24143404. Fax. +968 24413419. E-mail: drrnandagopal@gmail.com

Notice: Authors are encouraged to submit quizzes for possible publication in the Journal. These may be in any field of Clinical Neurosciences, and should approximately follow the format used here. Please address any submissions to the Assistant Editor, Neurosciences Journal, Riyadh Military Hospital, PO Box 7897, Riyadh 11159, Kingdom of Saudi Arabia.
E-mail: sdouglas@rmh.med.sa

A young adult with seizure and visual field defect

Case Presentation

A 27-year-old man presented with recurrent episodes of focal motor seizure with secondary generalization. Visual field charting revealed left homonymous hemianopsia. **Figure 1** shows the clinical photograph of face. His cranial CT is shown in **Figure 2**.



Figure 1 - Photograph of face.

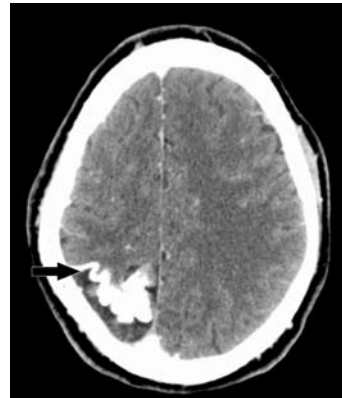


Figure 2 - Axial CT of brain (non-contrast scan).

Questions

1. What finding is observed in **Figure 1**?
2. What does the CT brain (**Figure 2**) show?
3. What is the diagnosis?

Neurosciences Quiz

Answers

1. Facial examination reveals port-wine stain (**Figure 1**) in the cutaneous distribution of the ophthalmic and maxillary division of the right trigeminal nerve.
2. Non-enhanced CT brain shows gyriform calcification in the right parieto-occipital region.
3. The presence of facial angioma (port-wine stain), seizure disorder, focal neurological deficit (hemianopsia), and the characteristic gyriform calcification (calcified lepto-meningeal angioma) on brain CT favors the diagnosis of encephalo-trigeminal angiomatosis, eponymously known as Sturge-Weber syndrome.

Discussion

Sturge-Weber syndrome is a neuro-cutaneous disorder that manifests with seizure, focal neurological deficits including visual field defect and stroke-like episodes, developmental delay, and glaucoma.¹ The intracranial involvement is due to leptomeningeal angioma that is usually unilateral and undergoes calcification resulting in the characteristic gyriform hyperdensity on CT scan. Bilateral intracranial involvement is rarely observed in only 15% of cases.² The syndrome is usually sporadic in occurrence. Although the condition is congenital, progressive neurological injury has been attributed to impaired regional blood flow.^{3,4} The present patient was started on anticonvulsants and had no recurrence of seizure during the follow-up period of 6 months. In patients with refractory seizures, resective surgery could be considered as an option for optimal seizure control.² Low dose aspirin has been suggested for the prevention of stroke-like episodes.⁵ The case illustrates the diagnostic importance of extra-neural signs, especially cutaneous signs, in the clinical evaluation of seizure disorders.

References

1. Puttgen KB, Lin DD. Neurocutaneous vascular syndromes. *Childs Nerv Syst* 2010; 26: 1407-1415.
2. Alkonyi B, Chugani HT, Karia S, Behen ME, Juhasz C. Clinical outcomes in bilateral Sturge-Weber syndrome. *Pediatr Neurol* 2011; 44: 443-449.
3. Pinton F, Chiron C, Enjolras O, Motte J, Syrota A, Dulac O. Early single photon emission computed tomography in Sturge-Weber syndrome. *J Neurol Neurosurg Psychiatry* 1997; 63: 616-621.
4. Evans AL, Widjaja E, Connolly DJ, Griffiths PD. Cerebral perfusion abnormalities in children with Sturge-Weber syndrome shown by dynamic contrast bolus magnetic resonance perfusion imaging. *Pediatrics* 2006; 117: 2119-2125.
5. Bay MJ, Kossoff EH, Lehmann CU, Zabel TA, Comi AM. Survey of aspirin use in Sturge-Weber syndrome. *J Child Neurol* 2011; 26: 692-702.

www.neurosciencesjournal.org

Neurosciences Journal Online features

- * Instructions to Authors
- * Uniform Requirements
- * STARD
- * Free access to the Journal's Current issue
- * Future Contents
- * Advertising and Subscription Information

All Subscribers have access to full text articles in HTML and PDF format. Abstracts and Editorials are available to all Online Guests free of charge.