

# Teaching NeuroImage: Nevus Flammeus, Ocular Melanosis, and Seizures in Young Adult With Sturge-Weber and Klippel-Trenaunay Overlap Syndrome

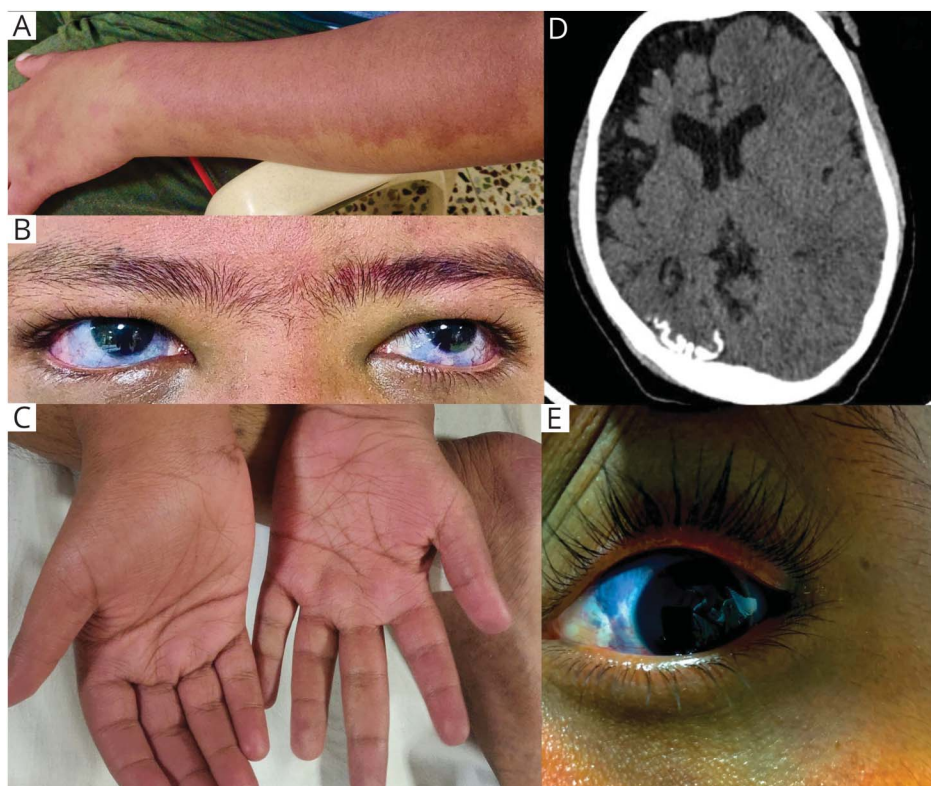
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*Neurology*® 2023;100:984-985. doi:10.1212/WNL.0000000000206901

**Figure** Features of Sturge-Weber and Klippel-Trenaunay Overlap Syndrome With Ocular Melanosis



(A) Port-wine stain over the patient's forearm, (B) bluish-grey ocular melanosis, (C) limb-length discrepancy with right upper-limb hyperplasia, (D) axial CT image showing right frontoparietal atrophy with gyral calcification in the parietal region, and (E) a close-up picture of the patient's left eye.

An 18-year-old man with delayed gross motor and language development and congenital glaucoma presented with recurrent seizures since 3 months of age. The seizures were characterized by left focal onset with secondary generalization and complicated by frequent drug noncompliance. Examination revealed a port-wine stain over the face and extremities (Figure, A), bluish-grey discoloration of the eyes and periocular region (Figure, B and E), and limb-length discrepancy with right upper-limb hyperplasia (Figure, C). These examination findings along with frontal atrophy and gyral calcifications seen on CT (Figure, D) prompted a diagnosis of Sturge-Weber (SW) and Klippel-Trenaunay (KT) overlap syndrome with

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ocular melanosis. Both SW and KT syndromes are mesodermal phacomatoses which share port-wine stain as a common clinical feature.<sup>1</sup> The presence of glaucoma, epilepsy, and cerebral gyral calcification are unique to SW, while the limb hyperplasia is exclusive to KT,<sup>1</sup> with ocular melanosis being additional.<sup>2</sup>

### Author Contributions

K. Lavanya: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data. R. Ramesh: drafting/revision of the manuscript for content, including medical writing for content; study concept or design; analysis or interpretation of data. S. Shanmugam: drafting/revision of the manuscript for content, including medical writing for content; analysis or interpretation of data. D. Avadhani: analysis or interpretation of data. P. Hazeena: analysis or interpretation of data.

### Study Funding

No targeted funding reported.

### Disclosure

The authors report no relevant disclosures. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

### Publication History

Received by *Neurology* October 14, 2022. Accepted in final form December 23, 2022. Submitted and externally peer reviewed. The handling editor was Resident & Fellow Deputy Editor Ariel Lyons-Warren, MD, PhD.

### References

1. Mandal RK, Ghosh SK, Koley S, Roy AC. Sturge-Weber syndrome in association with Klippel-Trenaunay syndrome and phacomatosis pigmentovascularis type IIb. *Indian J Dermatol Venereol Leprol*. 2014;80:51-53.
2. Prabhu PB, Soman D, Babitha V. Conjunctival melanosis with retinal vessel tortuosity: an unusual presentation of Sturge-Weber syndrome. *Kerala J Ophthalmol*. 2017;29:226-229.

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*Neurology* 2023;100:984-985 Published Online before print January 31, 2023

DOI 10.1212/WNL.0000000000206901

**This information is current as of January 31, 2023**

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