

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

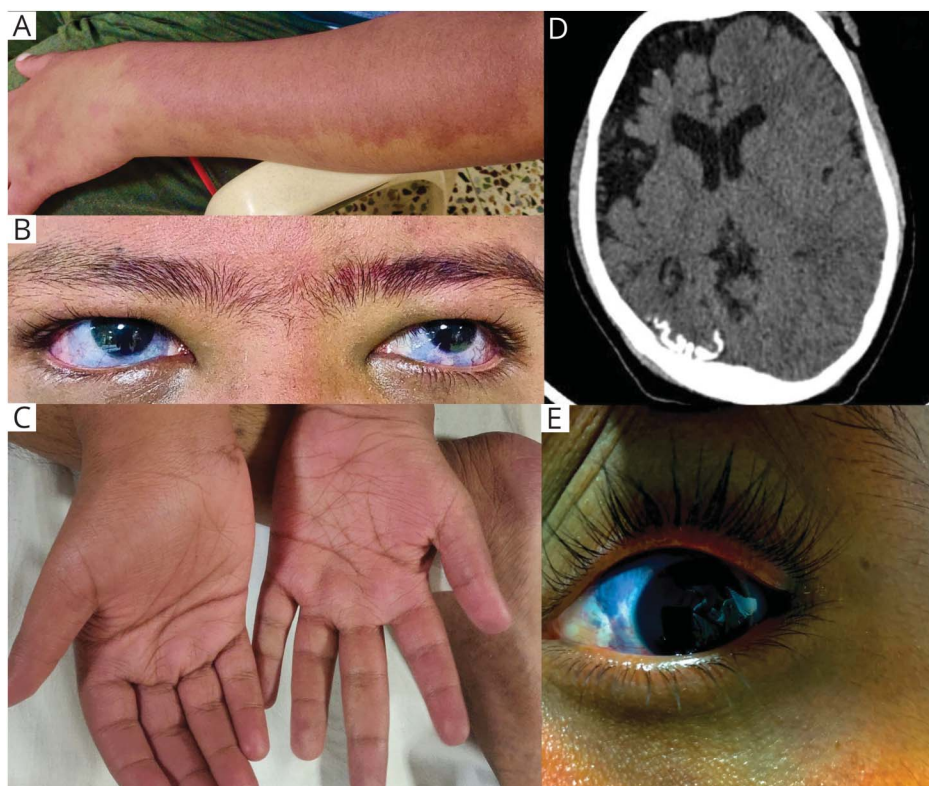
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Figure Features of Sturge-Weber and Klippel-Trenaunay Overlap Syndrome With Ocular Melanosis



(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.¹ The presence of glaucoma, epilepsy, and cerebral gyral calcification are unique to SW, while the limb hyperplasia is exclusive to KT,¹ with ocular melanosis being additional.²

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.

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