Teaching NeuroImages: Neurocutaneous melanosis

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Figure Axial and coronal views of multiplanar reconstructed images of the brain MRI and photograph of the melanocytic nevus

(A) Axial and (B) coronal views of multiplanar reconstructed images of the brain MRI demonstrate a single 8 mm × 7 mm × 6 mm lesion within the right amygdala with T1 shortening with no evidence of necrosis, hemorrhage, or edema. (C) The giant congenital melanocytic nevus.

A 3-month-old boy presented with a giant congenital melanocytic nevus involving his back. He was developing normally with no neurologic symptoms. Brain MRI demonstrated a T1-bright lesion in right amygdala without necrosis, hemorrhage, edema, or enhancement (figure). MRI spine was normal. The imaging findings in the clinical setting are consistent with a diagnosis of neurocutaneous melanosis.

First described by Von Rokitsanski in 1861, neurocutaneous melanosis is a rare sporadic embryonic neuroectodermal dysplasia characterized by congenital melanocytic nevi and melanotic tumors of the leptomeninges or brain parenchyma.¹ It most commonly results from an embryonic postzygotic somatic mutation in *NRAS*.²

AUTHOR CONTRIBUTIONS

Dr. Kumar: drafting the manuscript, chart review, analysis and interpretation of data. Dr. Dhamija: acquisition of data, study supervision, critical revision of the manuscript.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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Neurology 2015;84:e207

DOI 10.1212/WNL.0000000000001689

This information is current as of June 15, 2015

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