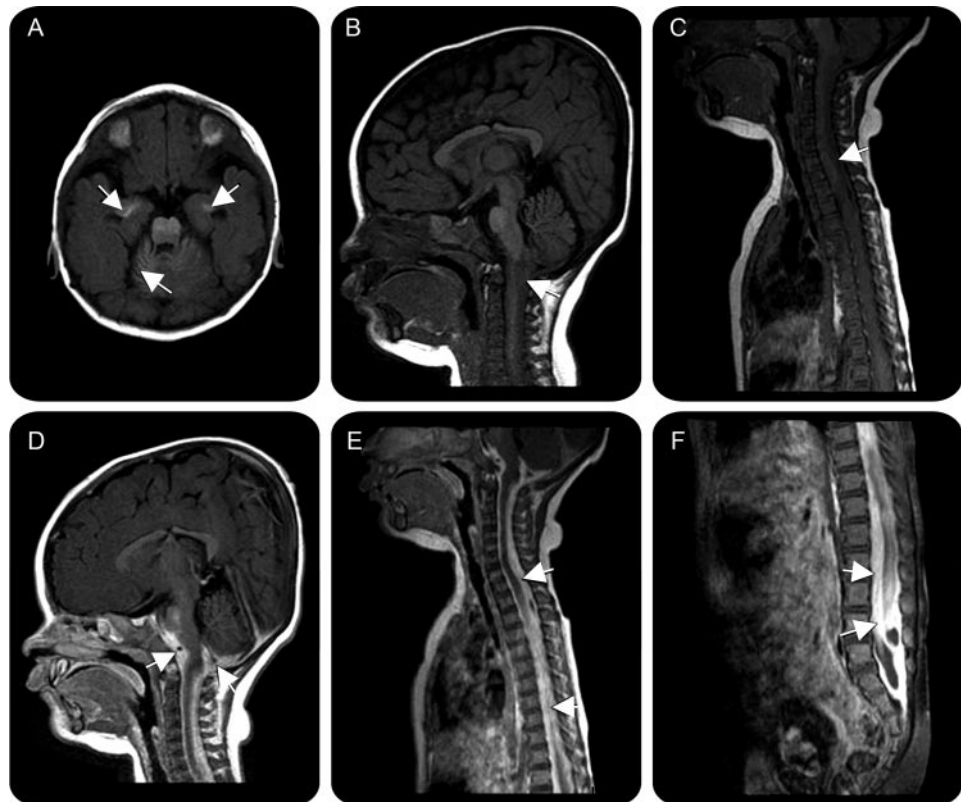


Teaching NeuroImages: Neurocutaneous melanosis

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Figure Unenhanced and contrast-enhanced MRIs



(A–C) Unenhanced T1-weighted MRI showing hyperintense areas consistent with parenchymal melanin in bilateral amygdala, pons, and cerebellar folia (arrows in A) and opacification of CSF spaces around the lower brainstem and cord (arrows in B, C). (D–F) Contrast-enhanced T1-weighted images showing florid enhancement (arrows) obliterating CSF cisterns at the base of brain and around the entire cord consistent with leptomeningeal melanocytic proliferation.

A 4-year-old boy presented with seizures, dysphagia, and weakness in all extremities. Examination revealed large nevi on the trunk which contained benign melanocytes on biopsy. A diagnosis of neurocutaneous melanosis was made on the basis of the MRI (figure) and CSF examination, which showed numerous melanocytes.

Neurocutaneous melanosis is a nonfamilial neurocutaneous syndrome characterized by large melanocytic nevi and excessive proliferation of melanocytes cells in the leptomeninges.^{1,2} CNS melanoma develops in about 50%. Patients present with hydrocephalus, seizure, cranial nerve

palsies, intracranial hemorrhage, and myelopathy.¹ Prognosis is poor. Treatment is shunting for the hydrocephalus and supportive.

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