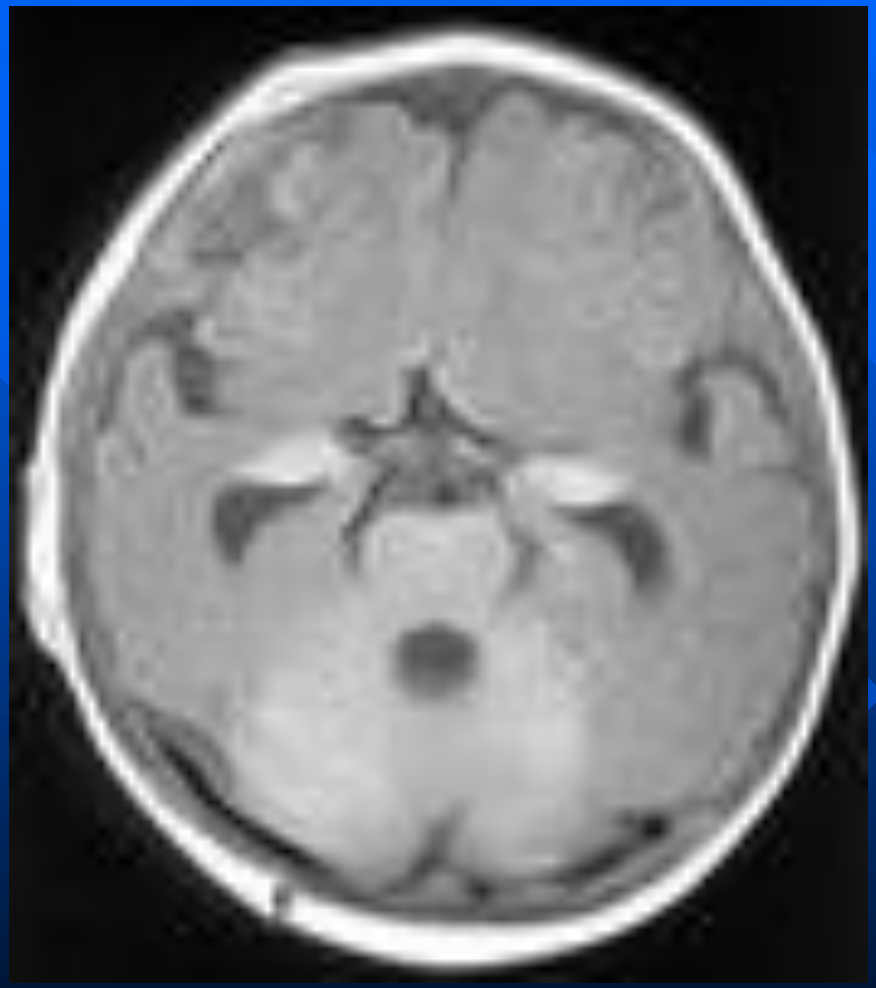
Neurocutaneous Melanosis

- Complex neuroectodermal dysplasia
  - Leptomeningeal and brain tissue proliferation of melanin-producing cells, primary malignant melanomas of the CNS and giant cutaneous naevi.
- Strong association: Cerebellar hypoplasia (10%)
- Normal MR does not exclude diagnosis NCM
- Sx NCM manifests by 2-3 years of age

Imaging

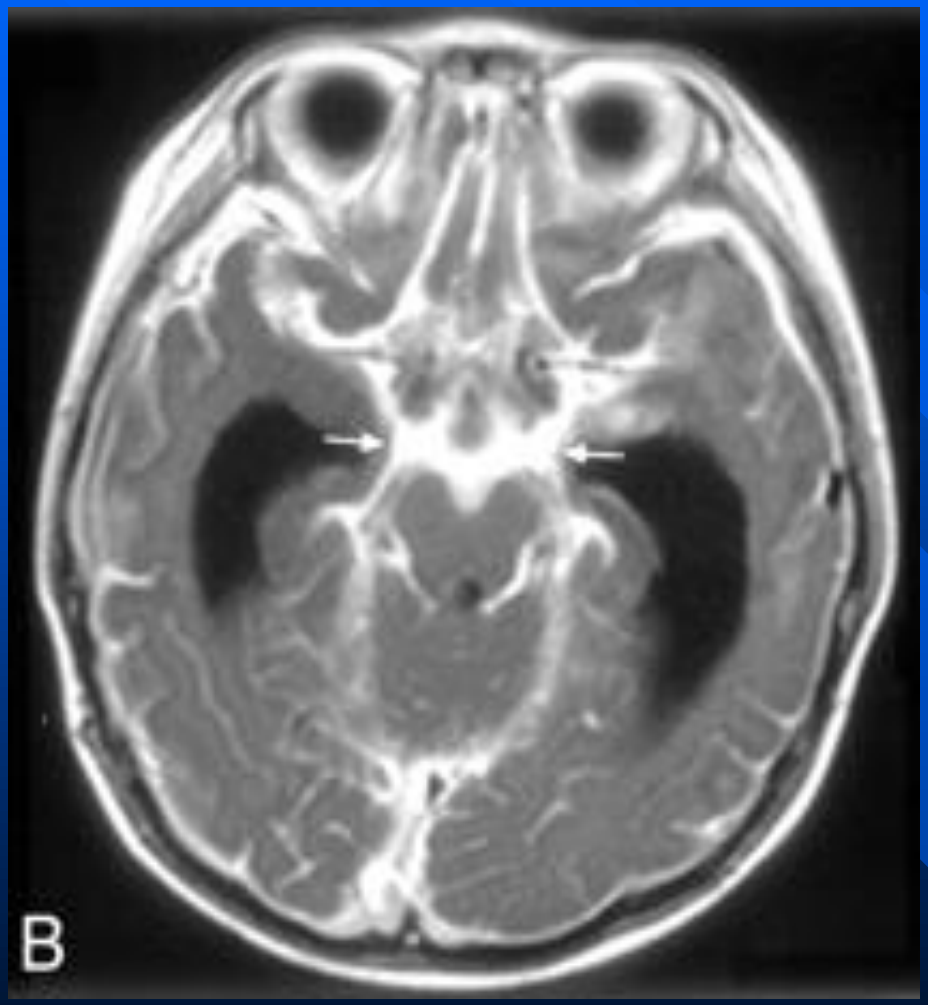
- Increased T1 signal in the amgdala and cerebellum, may enhance and bloom
- Diffuse leptomengial enhancement., increased signal on flair in sulci.
- May also see posterior fossa cyst and syrinx

Neurocutaneous Melanosis



T1

Neurocutaneous Melanosis (Post T1)



In child when see leptomengial enhancement, you need to consider Primary PNET and Leukemia should be the 1st thing you think about

Graphic shows localized dark (melanotic) pigmentation of the leptomeninges. Inset demonstrates extension of melanosis into the brain substance along the Virchow-Robin spaces (black solid arrow).