Neurocutaneous melanosis
Published on 29.04.2005

DOI: 10.1594/EURORAD/CASE.3062
ISSN: 1563-4086
Section: Paediatric radiology
Imaging Technique: MR
Case Type: Clinical Cases
Authors: Çevikol C, Karaali K, Åženol U, Lüleci E
Patient: 3 years, female

Clinical History:
A three-year-old girl presented to our hospital, with seizure.

Imaging Findings:
A three-year-old girl was admitted to our hospital with seizure. A physical examination revealed congenital nevi on the dorsal spine area. Subsequently, an MR examination was performed.

Discussion:
Neurocutaneous melanosis was first described by Rokitansky. He reported a 14-year-old girl with a giant congenital melanocytic nevus and mental retardation who developed late-onset hydrocephalus. Since that initial report, nearly a 100 cases have been reported. Giant congenital nevi are themselves a relatively rare birthmark, occurring in approximately 1/20,000–50,000 live births. With few exceptions, affected individuals have congenital pigmented nevi on the head and neck or dorsal spine area. A symptomatic CNS disease typically develops at less than two years of age with seizures or signs and symptoms of increased intracranial pressure. The prognosis of symptomatic neurocutaneous melanosis is poor; the majority of patients die within three years of onset of their initial neurological symptoms, either from CNS malignant melanoma or from the progressive growth of "benign" melanocytic cells. Computed tomography scans in children with neurocutaneous melanosis are found to be normal unless a collection of melanocytes has converted into a melanoma. An MR imaging procedure shows foci of T1, and sometimes T2, shortening in the brain parenchyma or meninges, signal characteristics that are compatible with deposits of melanin. These deposits of melanin are more easily detected in the unmyelinated brain, as the T1 and T2 shortening caused by the white matter myelination make the melanin less conspicuous. T1-weighted images are usually more effective in the detection of melanin deposits. The foci of abnormal signal are typically 3 cm or less in diameter; they are most commonly seen in the anterior temporal lobe, cerebellar nuclei, cerebellar white matter, and the brainstem. The T1 and T2 shortening is presumably the result of the presence of stable free radicals in melanin (identified by electron spin resonance studies). T1 hyperintensity is also seen in subacute hemorrhage (methemoglobin), proteinaceous material and presence of paramagnetic contrast material (gadolinium). The other imaging findings in neurocutaneous melanosis include cerebellar (primarily vermian) hypoplasia, the Dandy–Walker malformation, and arachnoid cysts.

Differential Diagnosis List: Neurocutaneous melanosis.

Final Diagnosis: Neurocutaneous melanosis.
References:


Description: A sagittal T1-weighted image showing hyperintense foci on the anterior temporal lobe.

Origin:
Description: A transverse T1-weighted image showing bilateral T1 shortening due to melanin deposition on the temporal lobes. Origin: