

Contrast-Enhanced MR Imaging of Neurocutaneous Melanosis

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Neurocutaneous melanosis (NCM) is a rare neuroectodermal dysplasia characterized by multiple congenital pigmented cutaneous nevi and melanotic thickening of the leptomeninges. We present a case of NCM displaying diffuse meningeal enhancement involving the brain and spinal cord. The role of contrast-enhanced MR imaging is discussed.

Case Report

A previously healthy 10-year-old girl presented with a 3-week history of headaches, vomiting, and diplopia. Physical examination revealed multiple pigmented cutaneous nevi, bilateral sixth nerve palsies, and papilledema. No pigmented intraocular lesions were found. Unenhanced CT and MR scans were unremarkable. An opening pressure of 38 cm H₂O was found at lumbar puncture with no abnormalities on CSF analysis. Treatment with corticosteroids for a presumed diagnosis of pseudotumor cerebri did not result in clinical improvement.

A lumboperitoneal shunt was placed 4 months later without improvement in the patient's symptoms. Repeat CSF analysis revealed a pleocytosis with a mononuclear cell predominance, hypoglycorrhachia, and an elevated protein. CSF cultures were negative. Cranial (Figs. 1A and 1B) and spinal (Fig. 2) MR imaging performed with a 1.5-T superconducting GE Signa imaging system after the administration of gadopentetate dimeglumine revealed marked diffuse leptomeningeal enhancement. A T2-weighted MR image was unremarkable (Fig. 1C). No focal mass or parenchymal invasion was demonstrated.

Primary CNS melanoma was suspected. Left frontal cerebral and meningeal biopsies were performed, which displayed a proliferation of cells, some containing melanin, in thickened leptomeninges, consistent with melanosis. There was no evidence of gross atypia or cerebral invasion. Multiple pigmented cutaneous nevi were excised, all displaying benign histology.

Following the placement of an Ommaya reservoir, the patient received 10 courses of intrathecal interleukin-2 (IL-2) and lymphokine activated killer (LAK) cells. An MR scan 10 months after initial presentation displayed increased ventricular size, correlating with symptoms of increasing nausea, vomiting, and mental confusion. A ventriculoperitoneal shunt was placed. A follow-up cranial MR scan

(Fig. 3) after gadopentetate dimeglumine administration revealed persistent diffuse leptomeningeal enhancement unchanged from the examination depicted in Figures 1 and 2. Ventricular size had returned to normal.

Discussion

NCM is a rare neuroectodermal dysplasia first described by Rokitsky in 1861. It is characterized by either multiple congenital pigmented or giant hairy cutaneous nevi in conjunction with a proliferation of melanin-producing cells in the leptomeninges. It is postulated that NCM is a congenital disorder involving cells of the neural crest, the progenitor of melanoblasts that embryologically migrate to the skin and pia mater [1].

NCM has been classified as one of the phakomatoses displaying hamartomatous malformations with a high rate of malignant transformation. Most cases occur sporadically and affect Caucasians in the first decade of life. No sex predilection has been demonstrated.

Those afflicted usually have multiple large pigmented or giant hairy cutaneous nevi in a truncal distribution. Patients with nevi involving the upper cervical dermatome or scalp are thought to be more likely to harbor leptomeningeal melanosis [2]. Malignant degeneration of cutaneous lesions in NCM is unusual.

The CNS is involved in all cases of NCM. There is diffuse pigmented thickening of the leptomeninges, classically described as being most marked at the base of the brain and surrounding the brainstem. Most display diffuse leptomeningeal involvement over the cerebral convexities while the spinal cord and its meninges are affected in up to 20% of cases. Intracerebral and intracerebellar pigmentation may also be present, which is a result of the migration of pigment-laden macrophages into the brain substance [3].

Hydrocephalus, psychiatric disturbances, and seizures oc-

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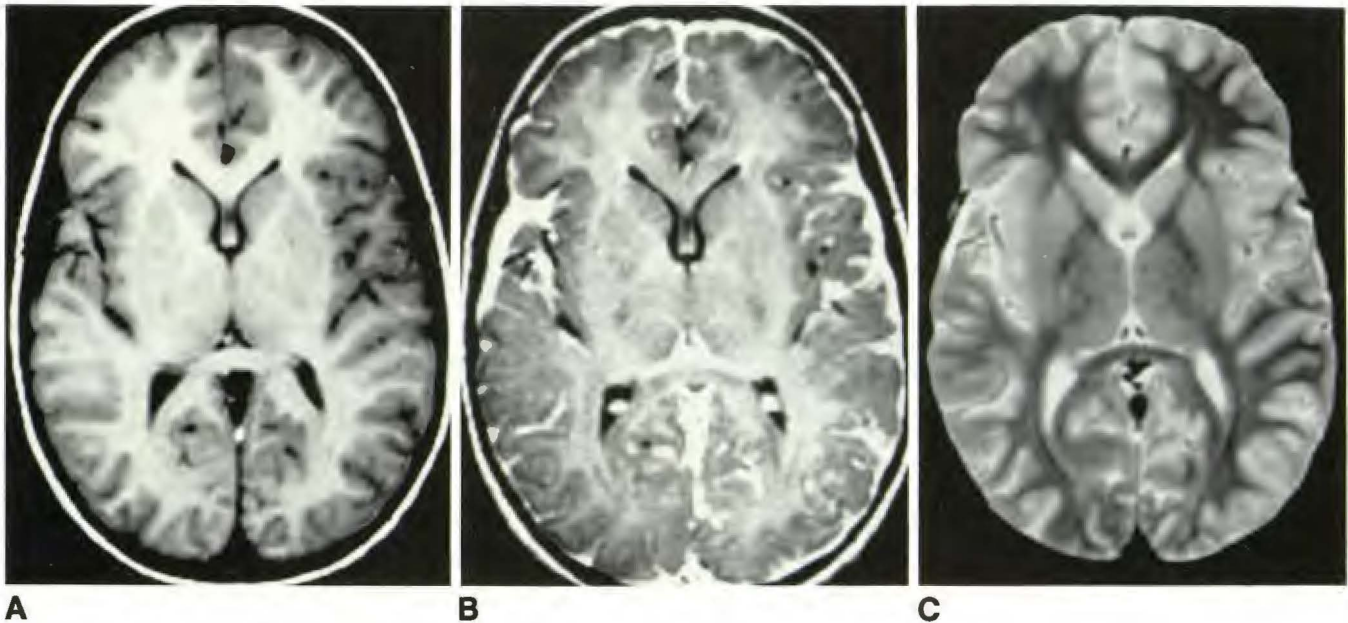


Fig. 1.—*A* and *B*, Axial spin-echo T1-weighted MR images (600/20/2) before (*A*) and after (*B*) contrast administration show marked leptomeningeal enhancement in cortical sulci, supracerebellar and sylvian cisterns, and the region of the velum interpositum. *C*, T2-weighted image (3000/80) is unremarkable.

Fig. 2.—Sagittal postcontrast spin-echo T1-weighted MR image (600/20) through thoracic spine shows enhancement of leptomeninges covering entire thoracic cord.

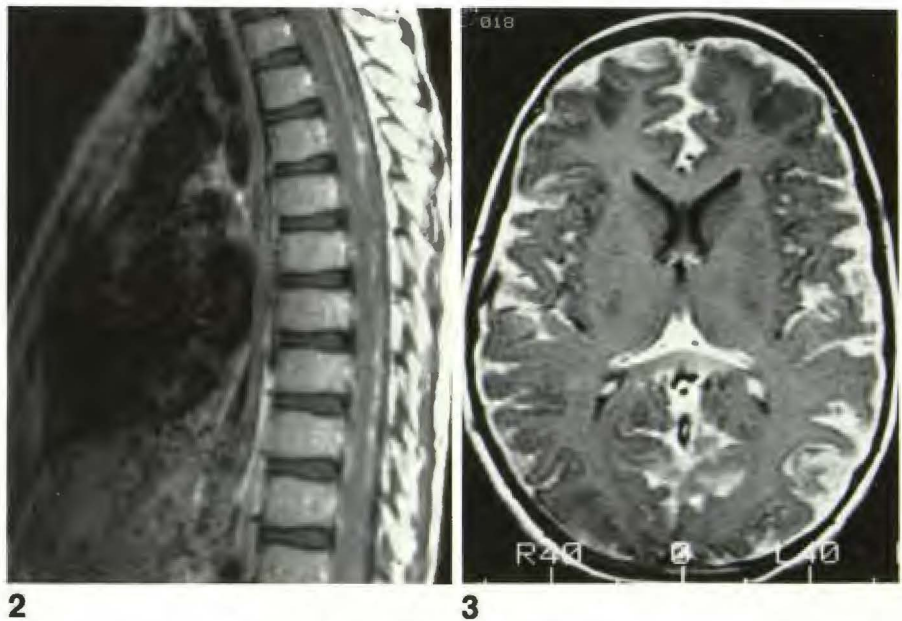


Fig. 3.—Axial postcontrast spin-echo T1-weighted MR image (500/20) after intrathecal IL-2 and LAK cell therapy. Diffuse leptomeningeal enhancement is unchanged from pretherapy images.

cur commonly. Spinal arachnoiditis, cranial nerve palsies, and syringomyelia have been reported as complications [4]. Hydrocephalus, occurring in almost all cases, is likely caused by melanotic deposits obstructing CSF outflow or by decreasing its absorption. Primary malignant melanoma of the CNS develops in approximately 40–50% of patients with NCM [3]. The diagnosis of leptomeningeal melanosis can only be made when no evidence of primary melanoma is found elsewhere. Malignant transformation is heralded by the appearance of

one or more intracranial or intraspinal masses or by direct parenchymal invasion [2].

An imaging method sensitive to the detection of leptomeningeal involvement in patients with multiple large pigmented cutaneous nevi would be clinically useful in identifying those with NCM. Cross-sectional imaging of NCM has been limited to CT in a few of the reported cases [1, 4, 5]. Meningeal enhancement by CT has been shown to involve the basilar cisterns, brainstem, cerebellum, and cerebrum. While en-

hanced MR imaging of the spine in NCM has been reported [6], a review of the literature reveals no reported cases of intracranial leptomeningeal melanosis imaged by MR. In our case, diffuse leptomeningeal enhancement involving both the brain and cord was present. The absence of focal intracranial masses or parenchymal invasion was consistent with the subsequently performed biopsy, which displayed benign proliferation of lightly pigmented cells throughout diffusely thickened leptomeninges.

Melanin has been shown to elicit paramagnetic effects by MR imaging, shortening both T1 and T2 relaxation times. The melanin-containing leptomeninges in our patient did not display these characteristics. One possible explanation for this may be the fact that the pia/arachnoid were only lightly pigmented in this patient. The lesser the melanin content, the less pronounced is the decrease in both T1 and T2 relaxation times [7]. Several metastatic melanotic melanoma lesions have also failed to display T1 and T2 shortening at 1.5 T, arguing against the universality of this finding in melanin-containing lesions [8].

Contrast-enhanced MR imaging will be useful in demonstrating leptomeningeal involvement in patients thought to

have NCM on the basis of cutaneous findings with or without CNS symptoms.

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