Congenital Melanocytic Nevi and Leptomeningeal Melanos: A Case Report

Dr. Manish Bhagat¹, Dr. Ekjot Gurudatta², Dr. Rimpi Rana³, Dr. Alisha Yadav⁴, Dr. Harjot Gurudatta⁵

¹ Assistant Professor, DNB Radiology, Department of Radiodiagnosis, Sri Aurobindo Medical College and Post Graduate Institute.
², ³ ³rd year Resident (M.D Radiodiagnosis), Department of Radiodiagnosis, Sri Aurobindo Medical College and Post Graduate Institute.
³ ³rd year Resident (M.S Ophthalmology), Department of Ophthalmology, Sri Aurobindo Medical College and Post Graduate Institute.
⁵ Assistant Professor, M.S Orthopaedics, Department of Orthopaedics, JNMC Raipur

ABSTRACT:

Introduction: Neurocutaneous melanosis is a rare congenital disorder characterized by the presence of large or multiple congenital melanocytic cutaneous nevi associated with intracranial leptomeningeal melanocytosis. Case Report: We notify the MR imaging findings of brain and spine of a 14 year male child with congenital giant hairy nevi who presented atypically with progressive neck rigidity, irritability, symptoms of meningitis and whose neurologic condition rapidly deteriorated. Method and Material: MRI was carried out on a 1.5-T SIEMENS symphony with Tim Technology (18 channel) the sequences which were evaluated: STIR –coronal and axial, T2 WI sagittal, axial and coronal, T1WI sagittal, axial and coronal, Gradient /medic –axial T2 space coronal (3D sequence). Results: MRI revealed diffuse moderate asymmetrical leptomeningeal pachymeningeal enhancement along cerebral convexities (LT>RT), sylvian fissures and basal cisterns with moderate communicating hydrocephalus. Abnormal T1 hyper intense foci in the ventral pons, right cerebral peduncle, right medial temporal lobe involving right amygdala, and over the left frontal convexities and in a given clinical setting may represent melanotic deposits.

1. Introduction

Neurocutaneous melanosis is a rare congenital disorder characterized by the presence of large or multiple congenital melanocytic cutaneous nevi associated with intracranial leptomeningeal melanocytosis[1]. Approximately 60% to 70% of all individuals with NCM develop symptoms, which usually appear before five years of age [2]. In 1861, Rokitansky, reported a case of a 14-year-old girl with mental retardation and congenital nevus who developed hydrocephalus. [3] Since that, around 100 cases of neurocutaneous melanosis have been stated in the literature. [3] It is associated with, melanomas of the central nervous system with melanocytic nevi present since birth. These are either solitary or huge (>20cm in greatest dimension) or multiple [3][4]. Maximum number of the patients reported are sporadic and characteristically occur in Caucasians.

We report the MR imaging findings of brain and spine of a child with congenital giant hairy nevi who developed progressive neck rigidity, irritability, symptoms of meningitis and whose neurologic condition rapidly deteriorated and was put on ATT for suspected TB meningitis. He died 7 months after the initial presentation.

2. Case Report

A 14-year-old boy presented with fever, poor diet, abdominal pain and vomiting, progressive neck
rigidity, irritability, symptoms of meningitis and whose neurologic condition rapidly deteriorated and was put on
Anti tubercular treatment (ATT) for suspected TB meningitis.
During the general physical examination the presence of a multiple nevus over the trunk thigh and arm, which he had since birth (Figure: 1). Though all of them were irregular, yet none showed any clinical signs of malignancy. On examination, the vitals were stable. He was fully conscious and oriented, pupils were 2mm dilated & both equally reacting to light, no cranial nerve palsies, motor or sensory deficit noted. His past medical history was unremarkable.

MRI was done which revealed asymmetrical leptomeningeal and pachymeningeal enhancement along cerebral convexities, sylvian fissures and basal cisterns with moderate communicating hydrocephalous [Figure 2 (a to c)]. These findings were suggestive of meningitis. CSF study revealed only slight increase in proteins without any increase in lymphocytes contradicting clinical diagnosis of tubercular meningitis.

Abnormal areas of T1 shortening [Figure 2(c)] were also seen in the ventral pons, right cerebral peduncle, right medial temporal lobe involving right amygdala, and over the left frontal convexities.

On clinical examination, he also had scattered melanocytic naevi over the back since birth without any itching or ulceration. None of the skin lesions were larger than 1cm. He would fit into Fitzpatrick skin photo type IV. The patient had no family history of melanoma or atypical melanocytic nevus.

Considering scattered melanocytic naevi over the back, above described T1 hyperintense foci could represent melanotic deposits.
Constellation of these clinical and radiological findings, were highly suggestive of leptomeningeal melanosis (NCM) rather than infectious or tubercular meningitis.

### 3. Discussion

NCM is a rare congenital syndrome characterized by the proliferation of melanin-producing cells both the skin and the leptomeninges [1]. Melanocytes originate from the neural crest cells and migrate to eyes, oral cavity, skin and leptomeninges. NCM develops from an abnormality in the development of these neural crest–derived melanocyte precursors, or melanoblasts of the skin and pia mater [8]. The criteria for diagnosis of NCM, as proposed by Fox [1], include the following:

1. Unduly large or unusually numerous pigmented nevi in association with leptomeningeal melanosis or melanoma,
2. No evidence of malignant change in any of the cutaneous lesions, and
3. No evidence of malignant melanoma in any organ apart from the Meninges.

Patients with leptomeningeal melanosis have a possibility for malignant degeneration, with the likely prevalence of 40–60% as per many other reports [4, 9]. Even in the absence of melanoma, symptomatic NCM has a poor prognosis [4]. GCMN is, generally defined as a congenital melanocytic lesion that will reach, at least, 20 cm in the skin in adult life [10].

Previously, the diagnosis of leptomeningeal melanosis was difficult to establish because malignant cells or melanin-containing cells were hardly found [4]. However, with development in techniques, the diagnosis of leptomeningeal melanosis can be made radiologically in patients with suspected NCM, especially when they become symptomatic or when they have neurologic signs such as epilepsy, hydrocephalus, cranial palsies [9–11] and myelopathy [11].

Involvement of spinal meninges in NCM has been reported in as many as 20% of cases [1] and it was also not present in our case. Van Heuzen et al has documented a focal intradural mass in the thoracic region in a case with NCM [12]. Rhodes et al described the diffuse spinal involvement in one patient [11].

The most common MR finding in asymptomatic children with NCM was T1 shortening in the infratentorial structures rather than leptomeningeal thickening as reported by Frieden et al [13]. Their findings correlates to the findings seen in our case. It was proposed by Faillace et al that malignant change was indicated by the presence of intracranial or intraspinal masses or from direct parenchymal invasion [14].

### 4. Conclusion

Our patient is a rare case as he presented with the most bizarre clinical presentation of fever, meningeal irritation, overlapping clinical features of TB meningitis pointing towards the diagnosis of infectious or tubercular meningitis. However, on MRI the presence of areas of T1 shortening and pachymeningeal enhancement along with the melanotic naevi on the chest and arm and other regions in our case highly suggested of leptomeningeal melanosis. Hence, it changed the diagnosis along with the approach to the management and the prognosis of the disease.

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### 7. Footnotes

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### 8. References


