Cerebellar amelanotic melanoma can mimic cerebellar abscess in a pediatric case of neurocutaneous melanosis

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Abstract

Neurocutaneous melanosis (NCM) is a rare phakomatosis that may be associated with intracerebral masses. The differential diagnosis of intracerebral masses in NCM is often challenging and should include pigmented and nonpigmented lesions.

KEYWORDS
adolescent medicine, neurology, paediatrics

INTRODUCTION

Neurocutaneous melanosis is a poor prognosis rare phakomatosis, characterized by congenital melanocytic nevi and leptomeningeal melanocytes proliferation. We report about a 8-year-old female affected by neurocutaneous melanosis with the recent discovery of a cerebellar mass; amelanotic melanoma and abscess were two of the differential diagnosis hypotheses.

Neurocutaneous melanosis (NCM) is a rare congenital syndrome characterized by the presence of large or multiple congenital melanocytic nevi and benign or malignant pigment cell tumors of the leptomeninges.¹

The pathogenesis is unclear, but it is considered as a consequence of an error in the embryonic neuroectoderm morphogenesis.²

NCM syndrome usually presents in the first two years of life with neurologic manifestations as a result of increased intracranial pressure, with mass forming lesions or spinal cord compression.¹

NCM is often associated with leptomeningeal melanoma, and symptomatic NCM had an extremely poor prognosis³ even if it occurs without any melanoma. Magnetic resonance (MR) imaging is the noninvasive gold standard method to perform NCM diagnosis of the central nervous system, which is invariably performed in association with cerebrospinal fluid (CSF) cytology and clinical evaluation.⁴,⁵ Nonetheless, it is difficult to differentiate benign from malignant leptomeningeal melanosis only with MRI as it is difficult, in some cases, to perform an exact differential diagnosis between NCM and other pathological entities, such as other types of mass forming lesions (e.g., cerebral abscess).

We want to describe how laboratory findings and MR images, in an 8-year-old female infant affected by NCM, can simulate the presence of a fungal cerebellar abscess instead of the histological confirmed amelanotic melanoma.

CASE DESCRIPTION

We present neuroradiological NCM findings of a symptomatic 8-year-old female infant with a 3q22.3 and 16p11.2 microduplication syndrome, intraparenchymal melanin infiltration, amelanotic melanoma, and also affected by epilepsy and neuromotor development delay.

At birth, on physical examination, she was noted to have multiple congenital pigmented nevi on the extremities, face, and trunk with variable sizes.

There was neither significant family history of neurologic disease nor NCM.
The patient was admitted for the first time to our department at 10-month-old, due to repeated seizures soon after birth and decreased consciousness level.

A brain computed tomography (CT) scan was performed and revealed tetraventricular hydrocephalus. She was further evaluated with 1.5T MR using multiplanar T1-weighted (T1-w) spin-echo and T2-weighted (T2-w) turbo spin-echo sequences with 2 to 4 mm thickness, with particular attention in the evaluation of T1-w images for their sensibility to melanin, before and after contrast medium administration.

MR scans confirmed CT hydrocephalus showing, besides, several findings suggestive for NCM: multiple T1-w hyperintense small lesions in the amygdala bilaterally, left thalamic region and left insula as well as cerebellum hemispheres, along with T2-w iso-hypointense signal on the left thalamic region (Figure 1A-E).

Hydrocephalus was treated with a ventriculoperitoneal shunt (Figure 1F) without any complications. Anticonvulsant therapy was then administered with good seizure control achievement. During follow-up, no neurological deficits were detected except for neurodevelopmental retardation; neuroradiological examinations were stable, and cutaneous melanocytic nevi histological analysis revealed no atypical cytological findings.

At 6-year-old, for recurrent headache and moderate increase of ventricular volume, she went to surgery for endoscopic third ventriculostomy. Few weeks after surgery, brain CT scan showed neither hydrocephalus nor other abnormalities except for hyperdensity of the left thalamic region and amygdala bilaterally consistent with her known MR diagnosis of NCM (Figure 2A-C).

At 8-year-old, she presented increased intracranial pressure neurological symptoms, such as intractable headache, vomiting, and generalized seizures. CT scan documented a 1 cm round faintly hyperdense intraparenchymal area in the left cerebellar hemisphere (Figure 3A-C).

MRI confirmed this finding as a 1 cm focal area of low signal intensity on T1-w and T2-w images, surrounded by edema and characterized by inhomogeneous contrast after Gadolinium administration. DWI showed restricted diffusion of the lesion (Figure 3D-Q).

No spine alteration was found at the MR scan. Lumbar puncture was performed, and cerebrospinal fluid was positive for Cryptococcus Neoformans var. Gattii.

This laboratory finding, in association with neuroradiological imaging, raised the suspicion of a cerebellar abscess, although the occurrence of other entities was not excluded, such as a neoplasm.

First, the patient was treated with antibiotics iv and antifungal, then only with antifungal and corticosteroids iv.

Two months later, a new brain MRI revealed a slight volume increase of the left hemisphere cerebellar lesion and the patient underwent total left cerebellar lesion removal.

**FIGURE 1** MR and CT scan of our 10-month-old patient with Neurocutaneous Melanosis and hydrocephalus. Axial T1-weighted images (A-D) MRI show a 10-month-old girl patient with hydrocephalus (white asterisk) and accumulation of melanin that presents with hyperintense signal (white arrowheads) in the insula (A), in the thalamus (B), in the retro-thalamic cistern (C), and in the amygdala bilaterally (D). Axial T2-weighted image of the same MRI acquisition (E) shows hypointense signal of melanin accumulation in the thalamic region (black arrow head) and hydrocephalus (black asterisk). After ventriculoperitoneal shunt placement (white arrow), axial CT scan (F) shows decrement of hydrocephalus (white asterisk), hyperdensity of the retro-thalamic cistern (white arrow head), and bilaterally frontal hygroma (black arrow head).
A new lumbar puncture and the intraoperatory CSF evaluation excluded the presence of Cryptococcus Neoformans var. Gattii and showed normal opening pressure, lower glucose levels (71 mg/dL), elevated proteins (maximum 7 g/dL).

The surgical specimen from the left cerebellar lesion was examined, and the result was a malignant proliferation with a solid growth pattern characterized by bulky and markedly pleomorphic epithelioid cells, eosinophilic cytoplasm with prominent nucleoli. Vascular invasion and atypical mitotic figures have also been identified.

The tumor cells immunohistochemistry revealed a positive reaction to anti-HMB-45, S-100, and Melan-A antibodies but a negative reaction to GFAP, OLIG-2, synaptophysin, CK AE1/AE3, CD 99, CD 56, beta-catenin, and GAB-1 and Ki67 = 40%.

The final diagnosis was secondary localization of epithelioid variant amelanotic melanoma.

Several months later, the patient died at the age of 8-year-old.

### 3 | DIFFERENTIAL DIAGNOSIS

Differential diagnosis of a cerebellar mass in NCM should include any type of mass forming lesion of the posterior cranial fossa (ie, abscess) and primary melanocytic lesions of the CNS. These latter, less common in patients without NCM, should be carefully considered in NCM cases, as they occur as discrete masses or diffuse proliferation. Primary melanocytic lesions of the CNS are meningeal melanocytoma, primary leptomeningeal melanomatosis, metastatic melanoma, primary cerebral malignant melanoma, and melanotic schwannoma.

Other more common mass forming lesions of posterior cranial fossa were not included in this list, since beyond the aim of this manuscript. Nonetheless, a fungal abscess was included in the differential diagnosis since Cryptococcus Neoformans was detected in the CSF of our patient, after a lumbar puncture.

#### 3.1 | Cryptococcosis

The clinical course of CNS cryptococcosis simulates NCM syndrome, with similar symptoms like headache, nausea, and vomiting. There could be also altered mental status, personality changes, confusion, lethargy, obtundation, or less frequently coma. Fever and nuchal rigidity are not generally noted. Hydrocephalus can be the consequence of meningeal scarring with dementia as a possible late complication.

Other possible symptoms include hearing defects, seizures, ataxia, aphasia, choreoathetoid movements, blurred vision, photophobia, and diplopia, generally secondary to arachnoiditis, papilledema, optic nerve neuritis, and chorioretinitis. It is worth nothing that immunocompetent hosts may present with either meningitis or focal cryptococcomas.

MR imaging of CNS cryptococcosis is generally a focal round, oval, or punctate imaging lesion, hypointense on T1-w and hyperintense on T2-w images, seen bilaterally in the basal ganglia, brainstem, and white matter of the cerebellar peduncles because cryptococcal organisms spread from the basal cisterns through the dilated Virchow-Robin spaces. FLAIR and postcontrast images are the most sensitive sequences used to demonstrate leptomeningeal infiltration.

Sometimes, an abscess can present as a single parenchymal mass formed by inflammatory cells and gelatinous mucoid material that shows hypointense on T1-w and hyperintense on T2-w images and isointense to CSF. Depending on the host’s immunological condition, there may be present a subtle rim of enhancement and edema.

It can be also associated with choroid plexitis of lateral and 4th ventricles or enhancing intraventricular mass and leptomeningitis.
FIGURE 3  MR and CT scan of our 8-year-old patient with Neurocutaneous Melanosis and a new onset left hemispheric cerebellar lesion. Multiplanar CT scans (A-C) show a hyperdense focal round lesion (white arrowhead) in the left cerebellar hemisphere. T1-weighted images (F-H) confirmed a hypointense round lesion (white arrowhead), mildly hyperintense in T2 (i,m,q) and in FLAIR images (J-L), surrounded by edema (white arrowhead). EPI-DWI (b = 1000s/mm²) (D) and ADC map (E) demonstrate diffusion restriction of the lesion (white arrowhead). In (H) leptomeningeal melanin infiltration is well depicted as leptomeningeal hyperintensity (empty arrowheads) in the left temporal lobe. Postcontrast multiplanar TFE-3D T1-weighted images (N-P) show the lesion in the left cerebellar hemisphere with inhomogeneous enhancement (white arrowhead)
Another imaging pattern can be represented by multiple, miliary and hypointense T1-w and hyperintense T2-w images, with enhancing parenchymal or leptomeningeal granulomas.  

3.2 | Meningeal melanocytoma

Meningeal melanocytomas are rare and generally benign neoplasms, found in the posterior cranial fossa, Meckel cave, or cervical and thoracic spinal canal. Some of these cases can have a malignant transformation.  

CT scans demonstrate extra-axial iso- to hyperattenuating lesion that enhances after administration of contrast material, like a meningioma but meningioma shows also calcifications and hyperostosis of the adjacent bone.  

MRI shows isointensity to hyperintensity lesions with T1-w pulse sequences and hypointensity or isointensity with T2-w pulse sequences, with homogeneous and intense enhancement. The signal intensity varies for different content of melanin and intratumoral hemorrhages.

3.3 | Primary leptomeningeal melanomatosis

Primary leptomeningeal melanomatosis is considered a meningeal variant of primary malignant melanoma, that involve the leptomeninges, without extracranial evidence of metastasis. It is a rare and aggressive neoplasm with a poor prognosis, and it presents with symptoms and signs related to increased intracranial pressure.  

Melanocytosis shows iso-hyperintensity of the signal on T1-w images and hypointensity on T2-w images, with intense enhancement and diffuse thickening of the leptomeninges after gadolinium administration.

3.4 | Metastatic melanoma

Metastatic melanoma can be diagnosed when there is a known primary malignant melanoma or when we cannot exclude a hidden primary neoplasm outside the CNS.  

It presents with multiple lesions, situated in the gray-white matter junction and surrounded by vasogenic edema, especially in the cerebellum, considered as the most common site of metastasis.

3.5 | Primary cerebral malignant melanoma

Primary malignant melanoma of the CNS is very rare and occurs most frequently in adult men. It develops most often within the leptomeninges and rarely within the ventricles. It has been reported that melanoma has a 2.6-fold higher incidence in patients with immunodeficiency than in immunocompetent ones.  

Only a few cases of spinal melanoma are reported in the literature, and they can have both intra- and extradural locations.  

A primary CNS melanoma can be diagnosed if there are not found other melanoma in another any sites and the median survival for patients with primary melanoma can be over 12 years.

MRI of a melanotic melanoma shows hyperintense signal on T1-w images and hypointense signal on T2-w images. Instead, an amelanotic melanoma shows isointense-hypointensity signal on T1-w images and moderately hyperintense signal on T2-w images. The enhancement pattern is generally homogeneous and sometimes inhomogeneous, peripheral, or nodular.

3.6 | Melanotic Schwannoma

Only about 80 cases are described in the literature, with similar features of both schwannoma and malignant melanoma. They are generally benign, with a 10% of malignant transformation possibility, and they present on CT as hyperattenuating lesions, associated with calcifications.  

MRI shows hypointense to isointense lesions on T1-w images and isointense to slightly hyperintense on T2-w images, with a variable enhancement pattern.

4 | DISCUSSION

NCM is a rare phakomatosis, characterized by the presence of giant or multiple congenital melanotic naevi and diffuse leptomeningeal infiltration of melanin-producing cells.  

The pathogenesis is unclear, but it is considered as a consequence of an error in the embryonic neuroectoderm morphogenesis.  

The first case of NCM was described in 1861 by Rokitansky in a 14-year-old girl with a giant congenital pigmented nevus who presented with hydrocephalus and mental retardation.  

The autopsy demonstrated diffuse infiltration of the leptomeninges with benign melanin-producing cells.  

However, the term was used before by van Bogaert in 1948 to describe another clinical syndrome, the heredofamilial melanosis.  

Criteria for NCM were proposed for the first time by Fox in 1972 and then revised by Kadonaga JN et al as follows:

1. Giant congenital melanocytic naevus
2. Diffuse leptomeningeal melanocytosis
3. Dermal melanocytosis
4. Evidence of melanoma in other areas

The median survival for patients with primary melanoma can be over 12 years. It has been reported that melanoma has a 2.6-fold higher incidence in patients with immunodeficiency than in immunocompetent ones.  

Only a few cases of spinal melanoma are reported in the literature, and they can have both intra- and extradural locations.  

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Criteria for NCM were proposed for the first time by Fox in 1972 and then revised by Kadonaga JN et al as follows:
Leptomeningeal melanoma has been reported in 40% to 64% of cases of symptomatic NCM,4,27,28 without gender prevalence.1

NCM syndrome usually presents in the first two years of life with neurologic manifestations as a result of increased intracranial pressure, with mass forming lesions or spinal cord compression.1

Our patient presented in her clinical history different degrees of hydrocephalus, seizure, consciousness disturbance. In most cases of NCM, symptoms are a consequence of intracranial pressure increase due to diffuse infiltration of leptomeningeal melanocytes or to development of primary central nervous system melanoma arising from these cells.5,28,29

Patients can present also with seizures, cranial nerve palsies, hemiparesis, aphasia, myelopathy due to spinal cord compression, and psychiatric disturbances.4,30,31

Hydrocephalus is the most common neurological finding due to flow obstruction and decreased absorption of CSF by infiltrated arachnoid villi, and it is treated with a ventriculoperitoneal shunt when symptoms occur.

Symptomatic patients with NCM commonly have a very poor prognosis,1 as they die within 2 or 3 years from the diagnosis.30,33,34

However, our patient died within several years after neurological symptoms onset.

In our case, imaging findings alone were not sufficient to differentiate the benign from malignant nature of the cerebellar lesion and quick neurological deterioration with cryptococcus presence in CSF supported the suspect of a cerebellar abscess. This CSF finding was of unclear nature since it is uncommon especially in an immunocompetent patient.6

Nonetheless, we were not able to exclude a leptomeningeal melanoma; thus, en-bloc lesion resection, specimen immunohistochemistry, CSF re-analysis, and follow-up MRI were necessary to guide a proper diagnosis.

MR is the best noninvasive method to study and evaluate NCM, but differential diagnosis between amelanotic melanoma and a fungal abscess may be challenging for their marked imaging similarities.

The first case of NCM associated with intracranial amelanotic malignant melanoma is described by Vanzieleghem BD et al and parenchymal melanin deposits are a consequence of melanocytes contained in the perivascular spaces,35 showing the importance of correlation of cutaneous lesions with intracerebral lesions.

When T1 shortening is absent, leptomeningeal enhancement is indicative of leptomeningeal melanosis.3

It is not possible to differentiate benign from malignant leptomeningeal melanosis with MR imaging,4 but the presence of hemorrhage, necrosis, vasogenic edema, lesion enlargement, and nodular enhancement is suggestive of malignant transformation.36,37

Different MRI pattern could be present in NCM patients, isolated or combined:

- in symptomatic patients, some authors noted an evident and spread enhancement of thickened leptomeninges surrounding the brain and spinal cord;4
- a second pattern of meningeal involvement was reported by other authors as increased signal intensity on unenhanced T1-w images and decreased signal intensity on T2-w sequences, due to the paramagnetic proprieties of the melanin.38 These findings are shown most in the anterior temporal lobes, especially in the amygdala;
- a third pattern was recognized in leptomeningeal melanomatosis, with the presence of amelanotic (unpigmented) tumor cells,39 resulting in a more difficult diagnosis that is based mainly on immunohistochemistry. S-100, Melan-A, and HMB-45 are important tumor markers for the diagnosis of melanoma. In particular, S-100 has higher sensitivity and HMB-45 has high specificity.40 In this setting, amelanotic melanocytosis, amelanotic melanocytomas, and amelanotic melanomas—as amelanotic infiltration of leptomeninges—could be characterized by the absence of T1-w hyperintense signal abnormality, as reported by Byrd SE et al4. Moreover, MRI can show different amelanotic patterns, ranging from iso-hypointense on T1-w images to hyperintense on T2-w images19,38;
- a fourth pattern, nonetheless, could present an exaggerated T2 effect due to a paradoxical decrease of T1 signal intensity, due to the presence of paramagnetic materials (melanin or contrast agent), before or following administration of Gadolinium-based contrast media.4 Several studies reported melanin deposits without signal abnormality on unenhanced MRI.35,41,42

5 | CONCLUSIONS

This case report illustrates the importance of melanotic cutaneous lesions correlated with MR imaging findings for the diagnosis of NCM.

Early MR, when available, is helpful if NCM suspect is raised because it is the best noninvasive method to evaluate the disease evolution.
Careful and repeated cytology screening of CSF with immunostaining is mandatory in the case of MR findings that can simulate different diseases. Sometimes, as in our case, brain biopsy or lesion excision at the level of abnormal radiological findings can be necessary to improve the chance for a good diagnosis.

Different MR patterns are described in the literature, although lesions characterized by short T1-w signal and decreased signal intensity on T2-w, due to the melanin paramagnetic effect, are the most frequent.

However, a differential diagnosis should be made between other paramagnetic substances and between other similar diseases.

Intracranial amelanotic malignant melanoma is a rare entity and it can develop from melanotic cerebral lesions.

Cryptococcoma presents with similar imaging findings of NCM lesions and the suspect can be raised when CFS analysis confirms the presence of Cryptococcus. Nonetheless, the final diagnosis requires histological analysis or meningeal biopsy.

In conclusion, a multidisciplinary approach is necessary for the management of NCM patients that require neurological, neurodevelopmental, and dermatological evaluations.

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CONFLICT OF INTEREST
Authors declare no conflict of interest.

AUTHOR CONTRIBUTIONS
ME, VSL, and CA: involved in study design and manuscript crafting. TA, CM, and GK: involved in images preparation and manuscript editing. VC and GF involved in study quality assessment, clinical data, and MR scan acquisition. GF: involved in ethical standard ad study guarantor.

ETHICAL APPROVAL
This study is following the Helsinki Declaration of Ethical standards.

DATA AVAILABILITY STATEMENT
The data that support the findings of this study are available on qualified request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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