Malignant Transformation In Neurocutaneous Melanosis Masquerading As Intracerebral Hematoma

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Citation

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Abstract

A 15 year-old boy was admitted with history of headache, deteriorating vision and recent onset of seizures and was brought in emergency in an unconscious state. A CT scan was done which showed hyperdense variegated bifrontal mass lesion. A bifrontal craniotomy was done suspecting it to be a hematoma and the mass evacuated. The histopathology of the evacuated mass showed feature of melanocytic proliferation, nuclear atypia and brain invasion. The boy had multiple congenital hairy nevi but no demonstrable evidence of cutaneous melanoma. The diagnosis of malignant transformation in neurocutaneous melanosis was given. The case highlights the importance of keeping the differential diagnosis of NCM in mind in children with congenital nevi and also of a thorough CNS examination including MRI.

INTRODUCTION

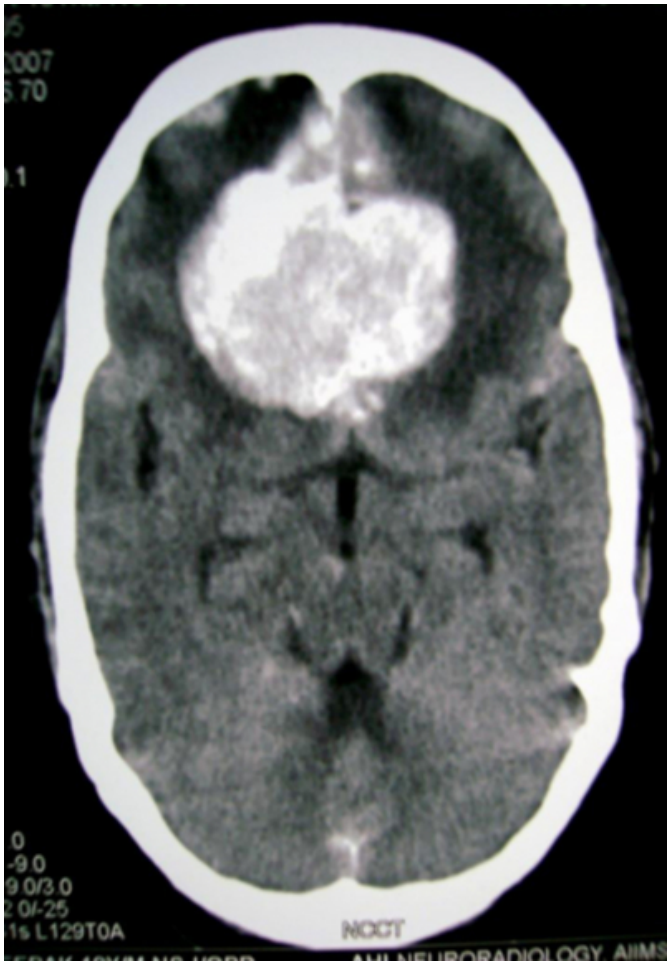
Neurocutaneous melanosis (NCM) is a rare congenital non-hereditary syndrome characterized by large congenital melanocytic nevi and leptomeningeal melanosis. The majority of patients are asymptomatic until the time of presentation that is usually heralded by an acute intracerebral event.

CASE REPORT

A 15-year-old boy with history of headache and gradually deteriorating vision was brought in neurosurgery emergency in an unconscious state. He had two episodes of generalized tonic-clonic seizures in a gap of 12 hours prior to admission. There was no history of trauma. On arrival he had a Glasgow Coma Score of 7(E1V1M5) and was stabilized with intubation, ventilation and epileptic therapy. Physical examination revealed 3 large congenital hairy nevi on buttock, nape of neck and one half of face. Additional variable sized 15 satellite nevi were present throughout the body. These lesions were present since birth and no new lesions had developed since then. Neurological examination was unremarkable except bilateral papilledema. CT scan of brain showed a bifrontal variegated hyperdense mass lesion with hypodensities within the mass (Fig 1).

Figure 1

Figure 1: Non-contrast CT scan showing a bifrontal variegated hyperdense mass lesion



He was advised an MRI scan with contrast that he refused due to financial reasons. A working diagnosis of intracerebral hematoma consequent to ruptured arteriovenous malformation was entertained.

An emergency bifrontal craniotomy was performed and hematoma evacuated. Intraoperatively, the surgeon was slightly suspicious of the unduly blackish discoloration of the evacuated friable appearing ‘blood clots' and histopathological opinion was requested. (Fig 2)

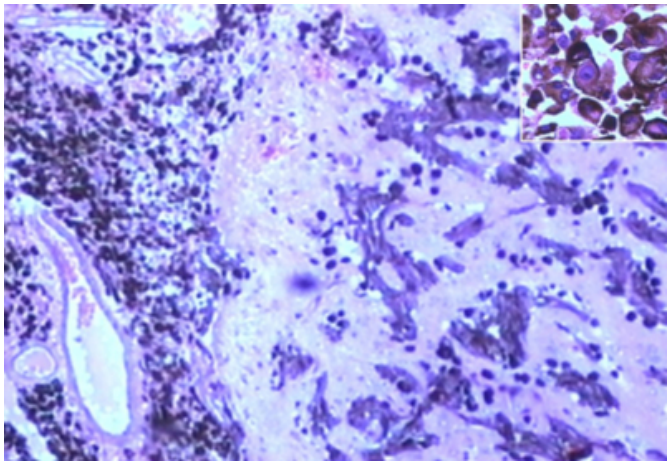
Figure 2

Figure 2: Macroscopic appearance of the evacuated mass

Microscopic examination of the mass showed proliferation and infiltration by pigmented cells in the meninges around blood vessels cerebral parenchyma and within Virchow-Robin spaces (Fig 3). Also there were focal necrosis and cells with nuclear pleomorphism with prominent nucleoli. (Fig 3inset)

Figure 3

Figure 3: Photomicrograph showing pigmented cells around blood vessels, within cerebral parenchyma and Virchow-Robin spaces (H&E, x100) with prominent nucleoli (inset)(H&E, x400).



Histochemistry for Masson-Fontana and immunohistochemistry for HMB-45 and MIB-1 confirmed it to be a malignant melanoma with proliferation index of 6%. A further dermatological opinion was sought to exclude cutaneous melanoma that was not found after three skin biopsies. Ultrasound abdomen and colposcopy were done to exclude a malignant melanoma elsewhere but none was found. In view of co-existence of congenital nevi and no evidence of malignant melanoma at any other site, a

diagnosis of primary intracranial malignant melanoma in neurocutaneous melanosis was given.

NCM is postulated to represent a congenital error in morphogenesis of the embryonal ectoderm.[1] The diagnostic criteria of NCM were defined by Kadonaga and Frieden [2] where the patients have a benign or malignant melanocytic tumor of the leptomeninges in conjunction with large (> 20 cm) and multiple (> 3) congenital melanocytic nevi without evidence of cutaneous melanoma in skin or elsewhere. Patients with NCM usually present in early childhood before the age of two years. [3] In a review of 39 cases of NCM, 62% had developed a intracranial melanoma over course of time.[2] Pathological proliferation of leptomeningeal melanocytes may commonly produce hydrocephalus internus or rarely a mass lesion representing a primary meningeal melanoma. Though a histological examination is essential for a confident diagnosis of NCM, a leptomeningeal melanoma can be visualized on MRI typically as hyperintense on T1-W and hypointense on T2-W due to unusual paramagnetic properties of melanin. [4] As in our case an MRI could not be done resulting in a misdiagnosis of hematoma on CT. The prognosis of NCM is extremely poor when patients become symptomatic or when neurological manifestations appear. In case of CNS involvement with or without malignant transformation the therapeutic options are limited and radiotherapy or chemotherapy do not improve the outcome and rapid deterioration in course is the rule. [5] Our patient was discharged after 2 weeks in conscious state and on antiepileptics and oral steroids but was lost to follow-up

In conclusion, NCM though rare presents in early childhood and should always be considered in patients presenting with congenital nevi and a thorough CNS examination including MRI should be performed for early and correct diagnosis.

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References

1. Cramer SF.The melanocytic differentiation pathway in congenital melanocytic nevi: theoretical considerations. Pediatric Pathol 1988; 8:253-65
2. Kadonaga JN, Frieden IJ. Neurocutaneous melanosis: Definition and review of literature. J Am Acad Dermatol 1991; 24:747-55
3. De David M, Orlow SJ, Provost N, et al. A study of large congenital melanocytic nevi and associated malignant melanomas: review of cases in New York University Registry and the world literature. J Am Acad Dermatol 1997; 36: 409-13
4. Woodruff WW Jr, Djang WT, McLendon RE, Heinz ER, Voorhees DR. Intracerebral malignant melanoma: High-field strength MR imaging. Radiology 1987; 165:209-13
5. Chu WCW, Lee V, Chan Yu-leung, et al. Neurocutaneous melanosis with a rapidly deteriorating course. Am J Neuroradiol 2003; 24:287-90

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