Neurocutaneous melanosisis: 
Criteria for diagnosis

Sir, 

We read with great interest the case report by Ahuja et al\(^1\) of a child, 6 weeks old, with multiple giant congenital melanocytic nevi and central nervous system melanosis. Neurocutaneous melanosis has been reported to manifest itself most commonly within the first two years of life\(^2\) and children with this entity have been born as still births or have been reported at as early as one month of age.\(^3\) However we agree that such a young age has not been probably reported before in Indian literature. We are very impressed at the fact that the authors suspected the entity of neurocutaneous melanosis in view of the fact that the child had multiple giant nevi located on the scalp, neck and posterior axilla (which are said to be risk factors for CNS melanosis)\(^2\) and proceeded to a MRI of the brain even though the baby was neurologically normal.

However, if one goes through the two landmark articles on this entity by Kadonaga et al\(^2\) and Fox et al\(^3\) one would come to the conclusion that this case reported would only qualify as a ‘provisional’ case of neurocutaneous melanosis. Both Fox and Kadonaga have laid down criteria for the diagnosis of neurocutaneous melanosis which are as follows:

1. Unduly large or unusually numerous pigmented cutaneous nevi in association with CNS melanosis or melanoma. This patient qualified this criteria.

2. No incidence of malignant change in any of the cutaneous lesions, except in patients in whom the examined areas of the CNS lesions are histologically benign. This criteria is very important because a significant percentage of patients with large congenital melanocytic nevi develop cutaneous melanomas\(^4\) and when cutaneous melanoma is present, the estimated incidence of CNS metastases is about 40%.\(^5\) Therefore if the CNS lesions have not been proved to be benign the cutaneous lesions have to be proved benign to rule out the possibility of the CNS lesions arising just out of metastases from a cutaneous melanoma.

Such histological confirmation would require at least a skin biopsy and/or autopsy on the death of the patient. Without this histological confirmation this case can best be labelled ‘provisional’ on the assumption that the skin lesions are benign and the CNS lesions are not metastases from a cutaneous melanoma. This concept of a provisional diagnosis of neurocutaneous melanosis has been advocated by Kadonaga.\(^2\)

The other strange about this case is the location of the CNS lesions in the amygdala and the thalamus. Fox\(^3\) and Kadonaga\(^2\) both have reported the leptomeninges to be the most commonly involved site of CNS melanosis. However, both these sites have been reported though rarely to be involved in CNS melanosis.

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References


Neurocutaneous melanosisis: 
Authors’ reply

Sir,

We thank the author(s) of the letter for taking interest in our article “Multiple giant congenital melanocytic nevi with central nervous system melanosis: a case report".\(^6\) We would like to reply to the three points raised as follows. Firstly, the diagnosis of neurocutaneous melanosis in our patient is ‘definite’ and is based on guidelines mentioned in ‘recent literature’.\(^7\) The diagnosis of multiple giant congenital melanocytic nevi (GCMN) is straightforward, a spot diagnosis, and doesn’t require a skin biopsy. On examining our patient, the same clinical diagnosis of “multiple GCMN” was made by a senior dermatologist and plastic surgeon in our institute. Although the neonate was neurologically normal, the presence of GCMN on scalp and dorsal spine prompted us to recommend an MRI brain study to screen for central nervous system (CNS) melanosis and operable presymptomatic non-melanocytic anomalies of the CNS.\(^2\) Eventually, MRI in our patient could be done at six weeks of age, as the parents took time to arrange money for this investigation. The diagnosis of CNS melanosis was