Neurocutaneous melanosis: Criteria for diagnosis

Sir,

We read with great interest the case report by Ahuja et al1 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 life2 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 al2 and Fox et al3 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 melanoma4 and when cutaneous melanoma is present, the estimated incidence of CNS metastasis 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.6

The other strange about this case is the location of the CNS lesions in the amygdala and the thalamus. Fox3 and Kadonaga2 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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made in our patient because of the presence of highly characteristic melanin deposits in the MRI study. This is the only method to diagnose CNS melanosis in a living patient. Secondly, in our patient there were no clinical indicators suggestive of a developing cutaneous melanoma, viz. changes in colour, size, shape, rapid growth rate, proliferative nodules or ulceration, in any of the GCMN. Hence doing a skin biopsy to rule out a developing cutaneous melanoma was not indicated. We would like to mention that malignant transformation within GCMNs is exceptionally rare in the neonate and has been reported in only three neonates. This recent review has stated that even if clinical indicators of cutaneous melanoma are present, careful histological evaluation may eventually reveal the lesion to be benign. Also, the melanin deposits detected in our patient’s MRI did not reveal any perilesional edema or necrosis or enhancement with gadolinium contrast; ruling out the lesion to be benign.

Thirdly, the presence of melanin deposits in our patient’s amygdala and thalami is not ‘strange’. A recent report has stated that the most common area of involvement in their case series was the amygdala; detected in eight out of 10 MRIs. Also presence of melanin deposits in thalami in cases of neurocutaneous melanosis is common.

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References

Clinical characteristics of organophosphate-induced delayed polyneuropathy
Sir,
I read with interest the recent article by Chatterjee et al. They report a patient with organophosphorus (OP) poisoning who developed intermediate syndrome (IS) and organophosphate-induced delayed polyneuropathy (OPIDP) in succession. However, I would like to make certain comments regarding the diagnosis of OPIDP in this case.

OPIDP as a clinical syndrome is well known and was described even before the association of IS with OP poisoning became known. The prevalence of OPIDP is variable; however, it occurred in 22% of patients with OP poisoning in a recent study. OPIDP occurs within a period of one week to five-six months of the ingestion of an OP compound, almost exclusively in patients with preceding acute cholinergic toxicity related to severe acute exposure (to an OP compound). However, there are occasional reports of OPIDP developing in a patient without prior cholinergic toxicity following a low-dose chronic exposure to OP.

The neuropathy in OPIDP is typically a symmetrical sensorimotor neuropathy, with a distal predominance. Initial symptoms are paraesthesia in the lower limbs and pain in the calves, followed by motor involvement of the lower limbs, manifested by leg weakness, foot drop and muscle hypotonia. The neuropathy in OPIDP is motor-predominant, and pure sensory neuropathy does not occur. If sensory symptoms are present, they are always milder than the motor component. Upper limb symptoms are always preceded by lower limb involvement. OPIDP is typically a “dying-back” neuropathy as revealed by clinical, electrophysiological and nerve biopsy data. The neuropathy has a typical “subacute” course of progression over a two-week period. In addition, features of pyramidal tract and posterior column involvement may be noted later in the course of illness. The patient reported by Chatterjee et al presented with a wrist drop and had features of right radial nerve palsy alone (even after electrophysiological studies) without any involvement of the lower limbs. This does not fit with the description of OPIDP as mentioned above and an alternative etiology for the same should be considered.

References