Letter to Editor

Unusual cause of recurrent flaccid paralysis in a child

Sir,

I read with interest the recent article by Jain et al. They report a child with two episodes of acute flaccid paralysis occurring at ages two and four years respectively, as a manifestation of serum sickness (related to cefadroxil administration). However, I would like to make certain observations.

Firstly, Jain et al have not presented the details of the nerve conduction studies and therefore, the electrophysiological pattern (axonal or demyelinating) is not apparent in this report. However, the child fulfills the diagnostic criteria (flaccid quadriaparesis, universal areflexia and albuminocytological dissociation in the cerebrospinal fluid analysis) for the Guillain-Barre syndrome (GBS). A detailed report of nerve conduction studies would help in further characterization of the disease.

Secondly, we need to exclude certain other conditions in a child presenting with arthritis and flaccid quadriaparesis, as GBS is a rare manifestation of serum sickness. These include Lyme disease, Campylobacter jejuni infection and common variable immunodeficiency with autoimmune disease. Exclusion of these possibilities would have further supported a diagnosis of GBS.

Finally, I have concerns regarding the use of steroids in treating this child. The current consensus is to use either plasmapheresis or immunoglobulins for treating GBS in children as steroids have not been found to be beneficial. Moreover, children treated with steroids have been found to have a higher rate of relapse of GBS.

S. Kumar

Neurology Unit, Department of Neurological Sciences, Christian Medical College Hospital, Vellore - 632004, India.
E-mail: sk@cmcvellore.ac.in

References

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 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. 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.

S. K. Sanyal, A. Gupta
Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi, India. E-mail: adtyagupta71@hotmail.com

References