CNS ABNORMALITY IN NEVUS DEPIGMENTOSUS

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A 5 - month old female child with seizures, motor defect, and EEG abnormalities with a hypopigmented skin lesion suggestive of nevus depigmentosus is presented.

Key words : Nevus depigmentosus, CNS involvement

Introduction

Nevus depigmentosus (ND) first described by Lesser in 1884 [1] is an uncommon, congenital, stable, quasidermatomal hypomelanosis which occurs sporadically. Although patients with ND are generally considered to be healthy, neurological abnormalities such as seizures have been rarely reported[1]

Case Report

A 5 month - old female child born at full term with normal vertex delivery of a non- consanguineous primigravida was referred from the Pediatrics department with history of tonic- clonic convulsions associated with decreased motor activity and high grade fever since 2 days for the evaluation of a hypopigmented lesion over the left shin which was stable and present since birth. The child had convulsions 18 days back which were associated with altered sensorium and loss of consciousness .

Examination revealed a mildly febrile child with epicanthal folds lying in frog's position. There was hypotonia of all the four limbs (lower limbs> upper limbs) with grade 4 muscle power. There was complete loss of head control which the child had attained at 3.5 months. All deep tendon reflexes were reduced bilaterally and the plantars were upgoing. Sensory examination, skull and spine were normal.

A single hypopigmented macule with normal sensation measuring 4 x 3 cm with irregular margins was seen on lower 1/3 of the left shin. Diascopy was negative.

Blood picture, blood sugar, Na^+ ,K^+ , Ca^{++} , P^+ levels were normal. Blood culture was sterile. No evidence of storage disorder was seen. CSF examination, CT scan, neurosonogram were normal. EEG studies revealed temporoparietal epileptiform transients. Urine examination showed proteins in traces with 30-40- WBCs, urine culture grew citrobacter species 75000 col \ ml. Urine metabolic screening was negative. ELISA for HIV was negative. Biopsies taken from the lesional and normal skin for H & E revealed normal number of melanocytes in both the specimens. Child was treated with phenobarbitone 30 mg. diazepam, ampicillin, chloromycetin, dilantin, frusemide, paracetamol with which the seizures and fever were controlled.

Discussion

Nevus depigmentosus has to be differentiated from nevus anemicus, incontinentia pigmenti achromians of ITO and vitiligo. Incontinentia pigmenti achromians

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of ITO[2] usually presents in early childhood, evolves, stabilizes and often reverses. It appears as swirls and parallel streaks of hypomelanosis and almost 75% of the patients may have CNS, eye, hair, musculoskeletal or internal organ involvement. Mental retardation in 60% and seizures in 50% cases have been reported. Nevus anemicus a pharmacological nevus is usually present since birth as a hypopigmented macule with a well-defined serrated margin. The lesion becomes nearly indistinguishable from the surrounding coloured skin on diascopy. In congenital vitiligo[1] there is complete loss of pigment with absence of melanocytes.

In contrast ND is usually present at birth, is chronic, stable and the shape is quasidermatomal as was seen in our patient. Patients with ND are generally considered to be healthy, neurological abnormalities being rare.[1]

Our Patient of ND had seizures, motor weakness, epicanthal folds but no evidence of musculoskeletal or hair anomalies.

References

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