

Hemimegalencephaly Presenting As Microcephaly

Hemimegalencephaly is a rare congenital brain malformation involving enlargement of only one cerebral hemisphere(1). Hemimegalencephaly is a type of hamartomatous lesion(2) or migration disorder, but its etiology remains unknown. Patients with the syndromic form of the disease have underlying disorders, most of which are neurocutaneous syndromes. Those with the isolated form of hemimegalencephaly have no such disorders. Usually hemimegalencephaly is suspected due to a large head, early onset of intractable seizures, hemiparesis, and/or skin lesions. We report a case of infant with incidentally detected hemimegalencephaly having global developmental delay and microcephaly.

A 6 month old girl infant first born of non consanguineous marriage, having global developmental delay (developmental age 2-3 months), presented for the first time with atypical generalized febrile convulsion (multiple). Perinatal history was uneventful. Clinically she was well nourished on breast feeds. Head was dolicocephalic with some sloping of forehead (head circumference 36 cm, microcephaly) all sutures were fused, weight 6.5 kg and length 65 cm. She had no dysmorphism or asymmetry of face and body. There were no neurocutaneous markers. In the absence of perinatal adverse events and fused cranial sutures, primary craniostenosis *versus* cerebral dysgenesis were considered as differential diagnosis.

EEG showed multifocal epileptical abnormalities and CT brain showed right sided hemimegalencephaly (*Fig 1*). She was put on sodium valproate and stimulation therapy. On follow up till ten months of age she had developmental age of 8-9 months. There was no recurrence of seizures.

In hemimegalencephaly, heterotopic neurons are immature and are of both neuronal and glial cell lineage(3). Glial proliferation has also been reported to explain the increased volume of white matter(4). It



FIG. 1 Right hemimegalencephaly with dilatation of ventricle on same side.

is generally suspected in patient with large head, intractable seizures and hemiparesis, or its association with neurocutaneous syndromes. Our patient had presented with microcephaly and atypical febrile convulsion with global developmental delay but had no hemiparesis even on follow up. Diagnosis is straightforward with neuroimaging, preferably MRI which can grade and pick up migration disorder(5).

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