Teaching NeuroImages:

Hemimegalencephaly in linear nevus sebaceous syndrome

Parayil Sankaran Bindu, DM
Sanjib Sinha, DM
Arun B. Taly, DM
Rose Dawn Bharath, DM

Address correspondence and reprint requests to Dr. Sanjib Sinha, Department of Neurology, National Institute of Mental Health and Neurosciences, Bangalore 560 029, Karnataka, India
sanjib_sinha2004@yahoo.co.in

Figure 1 Patient with linear nevus sebaceous syndrome

Facial nevus on the left side of the face with facial asymmetry.

A 15-month-old girl presented with developmental delay, intractable right focal motor seizures, hyperpigmented patch on the left side of the face, facial asymmetry, and right hemiparesis (figure 1). A diagnosis of linear nevus sebaceous syndrome (LNSS) was considered. Brain MRI revealed hemimegalencephaly (figure 2). EEG showed spike/wave discharges in left occipital leads. Ophthalmologic and skeletal evaluation were normal.

LNSS is a rare, sporadic neurocutaneous syndrome characterized by linear sebaceous nevus of Jadassohn,

mental retardation, and seizures.¹ Hemimegalencephaly is a major feature of this syndrome.² Skeletal and ocular involvement occur in up to 68% of the patients. Intractable seizures may necessitate epilepsy surgery.

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Figure 2 MRI of the patient

Brain MRI demonstrates the presence of hemimegalencephaly with an enlarged left hemisphere, colpocephaly (A, B), midline shift of a dysplastic occipital lobe (occipital sign, A, B), white matter signal intensity changes (B, C), and straightened frontal horn (arrow, C).

From the Departments of Neurology (P.S.B., S.S., A.B.T.), Neuroimaging and Interventional Neuroradiology (R.D.B.), National Institute of Mental Health and Neurosciences (NIMHANS), Bangalore, India.

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