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

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Teaching NeuroImages: Hemimegalencephaly in linear nevus sebaceous syndrome
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