

Teaching NeuroImages: Hemimegalencephaly in linear nevus sebaceous syndrome

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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.

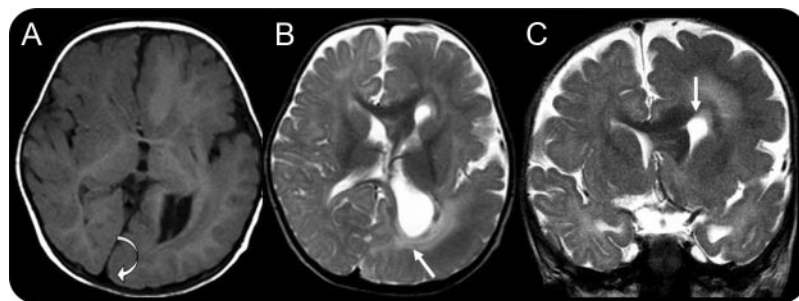
LNSS is a rare, sporadic neurocutaneous syndrome characterized by linear sebaceous nevus of Jadassohn,

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).

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