Teaching NeuroImages: Double cortex

Rare diagnosis in adulthood

Kristin M. Ikeda, MD, MSc, FRCPC, and Seyed M. Mirsattari, MD, PhD, FRCPC

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Correspondence

Dr. Ikeda
kmikeda@dal.ca

Figure 1 EEG

Anterior-posterior longitudinal bipolar montage demonstrates generalized epileptiform activity (arrows) superimposed on generalized background slowing, greater on the right (LFF: 1 Hz, HFF: 70 Hz, sensitivity: 200 µV, paper speed: 30 mm/s).

A 53-year-old, right-handed, developmentally delayed woman with presumed genetic generalized epilepsy was seen for evaluation. There was no relevant family history. EEG demonstrated generalized epileptiform abnormalities (figure 1). Neuroimaging demonstrated subcortical band heterotopia (SBH) and subcortical nodular heterotopia (figure 2). Genetic testing revealed heterozygous doublecortin (*DCX*) mutation (c.1150C>T p.Arg384*).

Despite the name heterotopia, SBH is a mild form of lissencephaly.¹ Lissencephaly is a spectrum of disorders resulting from insufficient neuronal migration.² The most common cause of SBH is *DCX* gene mutations, typically occurring in females.¹ Mutations of *DCX* in males result in lissencephaly.² It is typically diagnosed in childhood; rarely adulthood.

Author contributions

K. Ikeda contributed to study design, drafting, and revising manuscript. S. Mirsattari contributed to study design and revising manuscript.

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From the Division of Neurology (K.M.I.), Department of Medicine, Dalhousie University, Halifax Infirmary; and Department of Clinical Neurological Sciences (S.M.M.), Western University, London Health Sciences Centre, Canada.

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Figure 2 Brain MRI

Despite previous normal neuroimaging, T1-weighted sequences on repeat imaging clearly demonstrate bilateral subcortical band heterotopias (white arrows), slightly thicker on the right. Small periventricular nodular heterotopias (red arrows) are also present bilaterally.

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Disclosure

The authors report no disclosures relevant to the manuscript.

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References

1. Guerrini R, Dobyns WB. Malformations of cortical development: clinical features and genetic causes. *Lancet Neurol* 2014;13:710–726.
2. Di Donato N, Chiari S, Mirzaa GM, et al. Lissencephaly: expanded imaging and clinical classification. *Am J Med Genet A* 2017;173A:1473–1488.

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