

Image of the month

Double cortex syndrome – A rare cause of refractory epilepsy



Saurabh Bansal, Manoj Goyal*, Manish Modi, Chirag Ahuja

Department of Neurology, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

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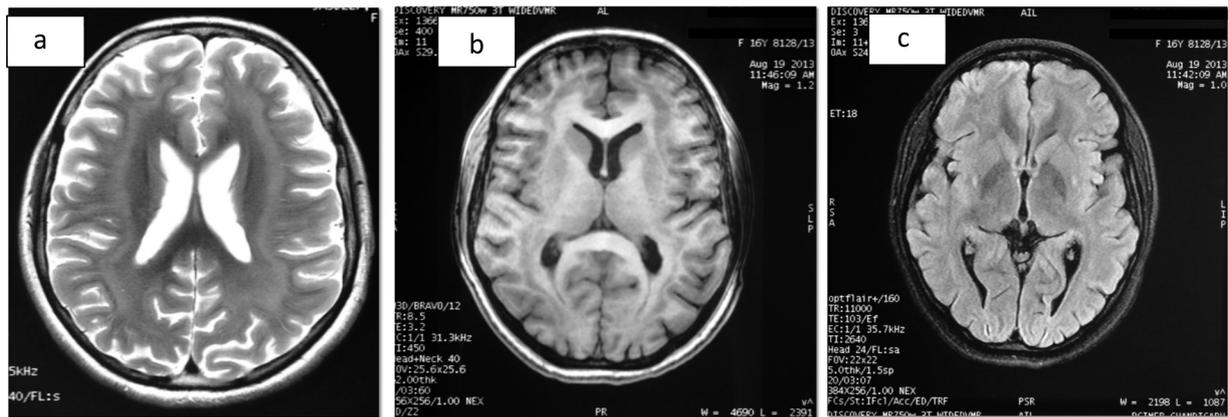


Fig. 1. Axial T2 weighted (a), T1 weighted (b) and FLAIR (c) magnetic resonance imaging sequences showing smooth laminar tissue with the intensity of grey matter interspersed between cortical grey matter and subcortical white matter, producing the classic picture of double cortex.

Miss A, a 16-year-old girl presented with dyscognitive focal (daily) as well as generalized seizures (once or twice a month) for past 12 years with poor scholastic performance. Magnetic resonance imaging (MRI) of brain revealed diffuse subcortical band heterotopia or double cortex syndrome (DCS) (Fig. 1). She was treated with carbamazepine (30 mg/kg) and levetiracetam (40 mg/kg) with decrease in frequency of focal seizure to once every two to three months and amelioration of generalized seizures. DCS usually occurs due to mutations involving doublecortin (DCX) gene located on X chromosome and almost exclusively occurs in females.¹ This gene encodes a microtubule associated protein essential for neuronal migration during embryogenesis. Most common presentation includes cognitive maldevelopment and seizures which usually start in first decade.

The severity of symptoms usually correlates with size of the band.² Recognition of the classic MRI pattern is important for prognostication, proper treatment as well as rehabilitation of these patients.

Conflicts of interest

The authors have none to declare.

References

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* Corresponding author.

E-mail address: goyal_mk@yahoo.com (M. Goyal).