

# Neuroimaging Highlight

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## Widespread Symmetrical Subcortical Band Heterotopia

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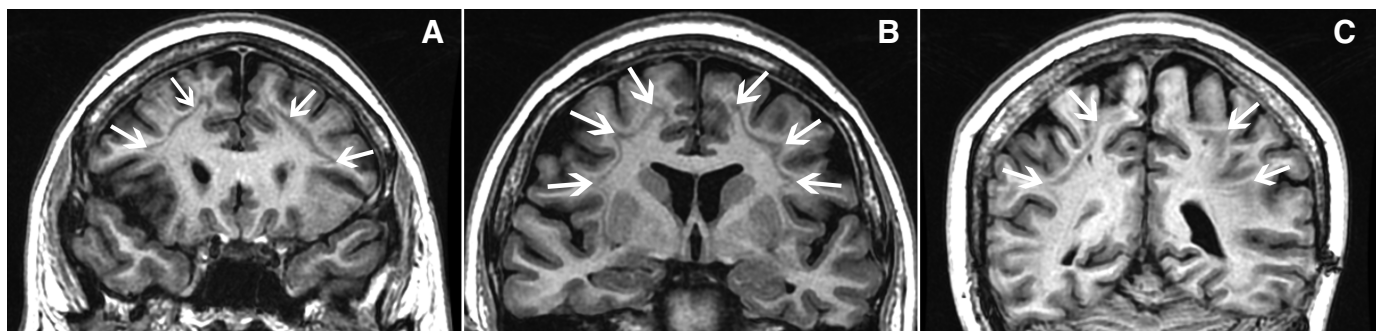
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A 47 year-old woman with normal neurological examination and normal intelligence developed medically refractory seizures at the age of 13 years. Over time, six distinct types of partial seizure were identified: left cheek myoclonus, forced tongue movements followed by lower extremity thrashing movements, left arm numbness and tingling, rising epigastric sensation, visual distortion, and graying of left superior quadrant vision. Each type could occur as a simple partial seizure, evolve to a complex partial seizure, or evolve to a secondarily generalized tonic-clonic seizure. Surface and subdural electroencephalographic studies confirmed independent seizure onsets from frontal, temporal and occipital lobes.

Magnetic resonance imaging (MRI) examinations at our own institution and at another epilepsy centre in the remote past were reported as normal (images no longer available for review). She underwent repeat MRI examination as part of a comprehensive re-evaluation (Figure 1). The MRI revealed a thin band of heterotopic grey matter paralleling the cortex closely on all sequences. This heterotopic grey matter was symmetrically distributed in the frontal, parietal, and occipital lobes. There was no other malformation.

Subcortical band heterotopia (SBH) is a disorder of neuronal migration that may be considered a forme fruste of classic X-linked lissencephaly. It is attributed to a defect in the production of doublecortin, a protein that stabilizes microtubules<sup>1</sup>. Doublecortin is encoded by the DCX gene on the X chromosome<sup>1</sup>. Females with a DCX mutation demonstrate functional mosaicism due to X-inactivation<sup>1</sup>. Consequently, neuronal precursors with normal DCX expression migrate to the cortex, while cells with mutant gene expression fail to migrate, giving rise to SBH<sup>1</sup>. Males with a DCX mutation can only produce abnormal doublecortin and so develop the X-linked form of classic (type 1) lissencephaly<sup>1</sup>. Different mutations of the DCX gene range in severity, with large deletions resulting in a more severe phenotype than single base pair substitutions<sup>2</sup>.

The imaging appearance of SBH was first described in 1989 by Barkovich et al<sup>3</sup>. In this series of five patients, the band of heterotopic grey matter was described as thick, and its margins did not parallel the infolding of the overlying cerebral cortex<sup>3</sup>. Even so, the abnormality had not been recognized at first reading in three of the five patients, because of its diffuse and

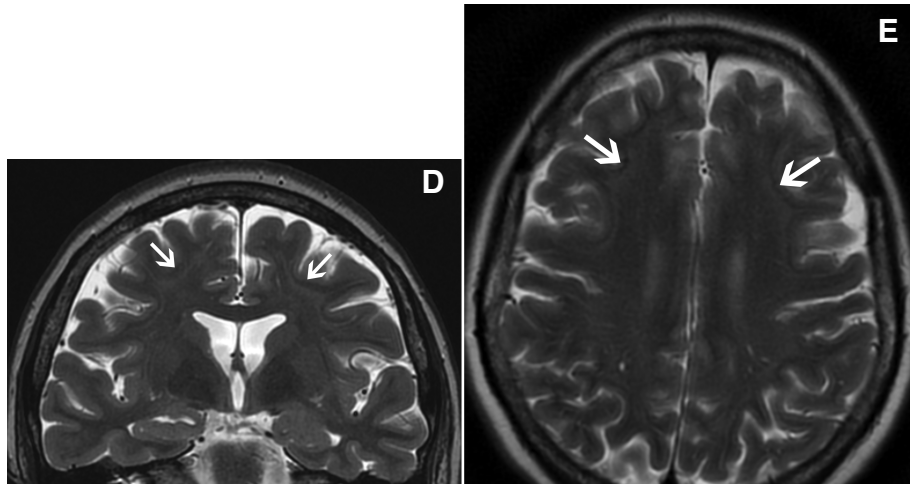


**Figure 1 A-C:** Coronal T1-weighted 3D volume gradient images (A-C) show a thin, symmetrical band of heterotopic grey matter that closely parallels the overlying cortex and extends from the frontal poles to the parietal/occipital lobes (white arrows).

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**Figure 1 D, E:** On coronal (D) and axial (E) T2-weighted images, the heterotopic grey matter follows the signal of the cortex. A thin, interposed layer of white matter can be appreciated (white arrows). The same findings were present on coronal and axial FLAIR images (not shown). The combination of high resolution coronal images and axial images in a dedicated epilepsy imaging protocol facilitates perception of the abnormality.

symmetrical nature<sup>3</sup>. In our case, the band of heterotopic grey matter is thin and parallels the cortex closely, making it even more difficult to perceive. Patients with this kind of SBH can be mislabeled as cases of “cryptogenic epilepsy”<sup>4</sup>. Such patients can benefit from re-imaging as higher resolution MRI sequences become available. Computer-assisted image analysis has also been suggested to aid further in the detection of subtle forms of SBH<sup>5</sup>.

Adult onset of epilepsy does not preclude an underlying malformation of cortical development<sup>6</sup>. Findings such as ventricular enlargement, pachygyria and band thickness appear to correlate with clinical severity in patients with SBH<sup>7</sup>. Our case demonstrates that widespread SBH need not be associated with neurological deficits or developmental delay. Semiology of seizures is often complex, with multiple types of seizures<sup>8</sup>. Surgical resections have been performed in a small number of patients, but the results have been largely disappointing<sup>8</sup>. Our patient continues to have 10-15 simple partial seizures and 1-3 complex partial seizures per month. Vagal nerve stimulation and medical treatment have allowed her to function as volunteer president of a nonprofit organization at the national level.

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