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MR Imaging of Incomplete Band Heterotopia

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In 1989, Barkovich et al. [1] described a new neuronal migration anomaly referred to thereafter as band heterotopia. Similar findings were also reported by Livingston and Aicardi [2] in describing two children with mild epilepsy and learning behavior problems. In band heterotopia, a continuous band of heterotopic neurons is interposed between the lateral ventricles and the cerebral cortex. The layers of ectopic gray matter are described as bilateral and symmetric. This distribution is one of the main reasons why CT and MR imaging often fail to identify it.

A case of atypical band heterotopia, characterized by an asymmetric distribution, is described in the present report.

Case Report

A 28-year-old woman was referred for assessment of drug-resistant jacksonian seizures. She was the second child of nonconsanguineous parents. There was no family history of note. Gestation period and birth were uncomplicated, and early development was normal. At the age of 11 years she had a generalized tonic-clonic seizure during sleep. An EEG recorded at that time showed a slow background with sharp waves located mainly on the left. Clinical examination was normal.

At the age of 14 years she had several tonic seizures with loss of consciousness lasting for a few seconds. An EEG showed epileptiform activity arising from the left frontotemporal region. Treatment with sodium valproate was started with a good response.

The patient was referred at the age of 27 years for assessment of seizures starting in the right upper extremity and occasionally extending to the lower limb, lasting a few seconds without impairment of consciousness. Neurologic examination and mental development were normal. She continued to have one or two seizures per week despite treatment with carbamazepine.

MR imaging showed the presence of layers of gray matter located under the hemispheric cortex and following the cortical convolutions (Figs. 1A and 1B). The anomaly was bilateral but asymmetric, being more marked on the left. A mild cortical abnormality could be seen in the left frontal area (pachygyria?) (Fig. 1C).

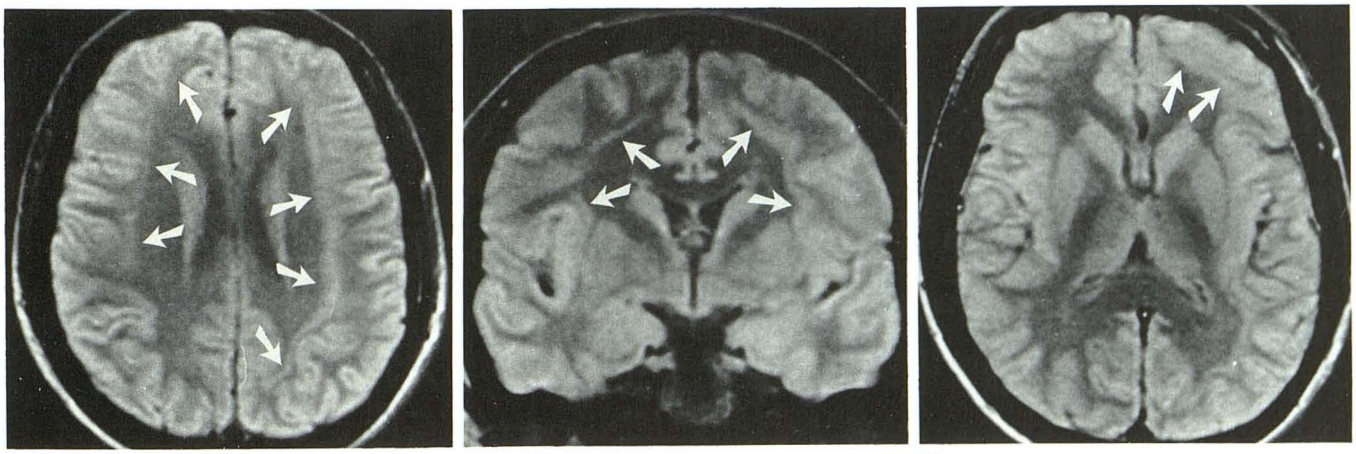

Fig. 1.—28-year-old woman admitted for drug-resistant seizures.
***A–C*, Axial (*A*) and coronal (*B*) proton density-weighted (2500/25/2) MR images show the presence of gray matter layers located in the white matter between the ventricles and the cortex. Note that the heterotopic layers are strongly asymmetric (*arrows*). Scan of frontal area (*C*) shows abnormal gyral pattern (pachygyria?) (*arrow*).**

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Discussion

Heterotopias comprise a complex and wide spectrum of abnormalities, characterized by the presence of islands of gray matter located outside the cortex or the basal ganglia. Different classifications have been proposed since the first descriptions of isolated cases appeared in the second half of the last century.

A first differentiation between nodular and laminar forms was made by Jacob [3]. He described the more frequently observed *nodular* forms as subependymal masses of gray matter, which form clusters of rounded nodules well separated from the cortex by normally myelinated white matter. They are most often localized at the corners of the lateral ventricles. *Laminar* forms are separated from both the cortex and ventricle walls by thick layers of white matter. Since 1989 a third form was described by Barkovich et al. [1], who labeled it *band heterotopia*. This newly recognized neuronal migration anomaly is characterized by the presence of bilateral and symmetric layers of heterotopic neurons between the lateral ventricles and the cerebral cortex. The heterotopia spans the whole cortex of both hemispheres with smooth margins at the interfaces with the adjacent white matter. Mild abnormalities of the cortical pattern, mostly represented by pachygyria, may coexist.

The symmetric distribution and the almost normal cortical pattern are considered the main reasons for the difficult identification of band heterotopia on CT and MR imaging. In all cases described by Barkovich et al. [1], intractable seizures and severe developmental delay led them to the conclusion that "the apparent poor outlook of these patients makes band heterotopias one of the more severe forms of migration anomaly." Others described a better prognosis in patients with band heterotopia [2, 4].

The pathogenesis of heterotopias is poorly understood; fever, radiation, and toxins have been implicated in animals and humans [5–7] if encountered between the fourth and 14th week of gestation. Nodular heterotopia is supposedly ascribed to the incapacity of the neurons in the laminar zones to regularly migrate after proliferation. In the other two forms, the migration starts but is never completed. In such cases some neurons lie inside the white matter between the ventricles and the cortex, giving rise to the heterotopic cortex. An incomplete involution during the back-migration of the superficial granular layer could be another explanation for the origin of heterotopias. The heterotopic cortex, therefore, might derive from the persistence of the granular layer inside the white matter.

Hypothetically, the main factor responsible for the neuronal migration may be a chemical factor whose defect would explain the arrest of migration. In band heterotopias, the diffuse and symmetric abnormality makes other hypotheses (vascular or infectious diseases) less tenable. The uniform distribution of the heterotopia could easily be attributed to an incomplete migration factor [1].

In our case, the heterotopic layers, although bilateral, presented a marked asymmetry. This finding is quite useful in evaluating the pathogenesis of heterotopias. The asymmetry observed in our study suggests that the hypothesis of a chemical factor defect, even if possible, cannot be generalized. Therefore, other causes must be considered. Our patient had no history of infectious or vascular disease or exposure to radiation or other chemical agents during gestation. Therefore, we have no feasible alternative pathogenetic proposal.

Concerning the clinical manifestation of band heterotopias, no developmental delay was found in the subject of our report. Because she was 28 years old at the time of MR examination, a worsening of the disease seems unlikely. These data are not in accordance with the findings of Barkovich et al. [1], in which all patients were severely delayed, nor with those of Livingston and Aicardi [2], who described two cases of mild epilepsy with learning and behavior problems. The concept that neuronal migration disorders have a wide spectrum of severity is therefore confirmed.

Concerning the MR imaging findings, the size of the ventricular system in our patient was normal. Since in earlier reports [1, 2] a dilatation of the ventricles was evident, we think that ventricular size could be an important prognostic factor in evaluating these patients.

In conclusion, we describe an incomplete form of band heterotopia in which the asymmetry of the anomaly and the normal mental and physical development of the patient suggest a milder form of band heterotopia than previously reported. In such cases, the outlook for the patient seems to be more favorable than that in more severe cases. Owing to the asymmetric distribution of the anomaly, a pathogenesis other than a chemical factor defect seem possible.

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