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Case Report

Hemimegalencephalic appearance of normal hemisphere in unilateral heterotopia and absent corpus callosum

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Abstract

We report two patients with medically refractory epilepsy who had MRI evidence of unilateral subcortical nodular heterotopia and agenesis of corpus callosum. The abnormal hemisphere was small, whereas the contralateral normal hemisphere appeared large and crossed the midline. Although the normal hemisphere was initially mistaken for hemimegalencephaly, there were no typical radiological features. Moreover, the electroencephalographic abnormalities lateralized to the hemisphere showing heterotopia. Because contralateral hemispheric abnormalities like heterotopia, hemimicrencephaly, and hemimegalencephaly can occur in patients with hemispheric heterotopias, we emphasize the importance of careful scrutiny of the contralateral hemisphere in patients with unilateral heterotopia. Absence of typical radiological features and appropriately lateralized electroencephalographic abnormalities will help differentiate the two. This is crucial when planning epilepsy surgery.

Keywords: Heterotopia; Hemimegalencephaly; Agenesis of corpus callosum; Hemimicrencephaly

1. Introduction

Gray matter heterotopia is a common malformation of neuronal migration associated with epilepsy and neurodevelopmental abnormalities. It is characterized by the presence of gray matter in the subcortical white matter [1,2]. Heterotopia may affect both hemispheres or can be confined to one hemisphere [2,3]. Advances in imaging technology have led to a better understanding of this interesting malformation of cortical development. We report the cases of two patients with medically refractory seizures caused by unihemispheric heterotopia and absent corpus callosum, in whom the normal contralateral hemisphere appeared hemimegalencephalic. Although abnormalities in the contralateral hemisphere have been described in patients with hemimegalencephaly [4,5], our cases are unique in that the normal hemisphere appeared unusually large and was initially mistakenly thought to be hemimegalencephalic, whereas the abnormal hemisphere was small.

2. Case 1

A 6-year-old right-handed boy with normal birth and mild language developmental delay presented with medically refractory seizures from the age of 3 years. Clinical seizure semiology was suggestive of right hemispheric, extratemporal complex partial seizures with infrequent secondary generalization, with a seizure frequency of about four per month. No benefit was derived from optimal doses of oxcarbazepine. Examination revealed normal head

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circumference and no focal neurological deficits. The EEG showed frequent right hemispheric spike-and-wave discharges with normal background activity. There were no epileptiform discharges over the left hemisphere.

MRI of the brain revealed irregular lobulated masses of gray matter located subcortically in the right hemisphere, consistent with subcortical heterotopia along with dysplastic basal ganglia and thalamus (Fig. 1). The right temporal lobe showed poor gray–white differentiation. The cortical mantle was thin, with poorly formed sulci. The sylvian fissure was ill-defined, and the corpus callosum was absent. The right hemisphere, along with the right half of the brainstem, was smaller than the contralateral side. Cerebellar hemispheres were normal. The normal left hemisphere appeared unusually large, crossing the midline and bulging into the right side. The cortex, however, showed normal gray–white distinction with no sulcal or gyral abnormalities. The signal intensities from the

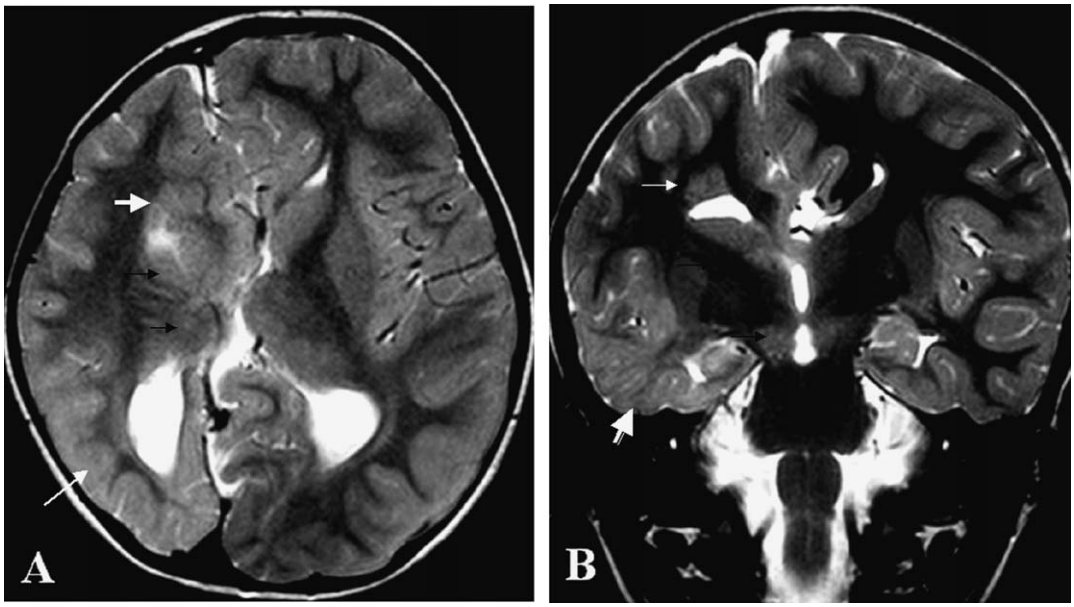

Fig. 1. T2-weighted axial (A) and coronal (B) magnetic resonance images of the brain of case 1. Subcortical heterotopia in the right frontoparietal white matter with overlying thin cortex (thin white arrow) and dysplastic basal ganglia and thalamus (black arrows) is evident. The right hemisphere appears small. Note the absence of corpus callosum and poorly formed sylvian fissure. Gray–white differentiation in the right temporal lobe is poor (arrowhead). Left hemisphere appears larger and crosses the midline. However, the gray–white differentiation is normal, with no evidence of increased signals in the white matter.

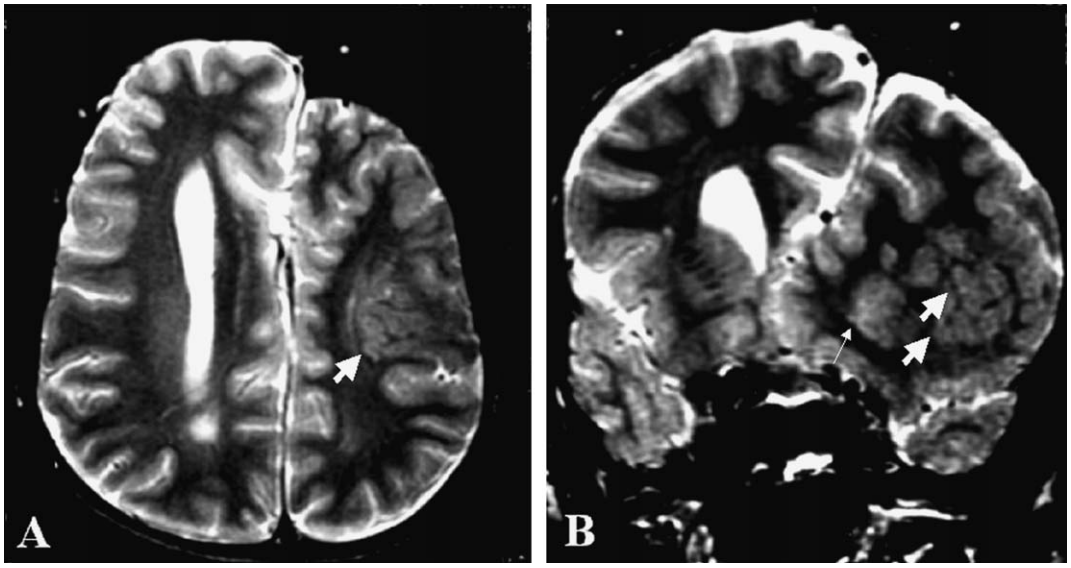

Fig. 2. T2-weighted axial (A) and coronal (B) magnetic resonance images of the brain of case 2. Heterotopia of the left frontoparietal region (arrowheads) along with smaller hemisphere is clearly seen. The basal ganglia are dysplastic on the affected side (thin arrow). Note the agenesis of corpus callosum. Right hemisphere appears enlarged, but with normal gray–white differentiation and signal intensities.