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An Unusual Presentation of Focal Cortical Dysplasia

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Summary: In a case of histologically proved focal cortical dysplasia, there was an absence of cortex-white matter delineation in the right parietooccipital area only on the T2-weighted images. This pattern correlated with the gross and histologic findings obtained on the resected cerebral tissue.

Index terms: Brain, abnormalities and anomalies; Brain, magnetic resonance; Seizures

Focal cortical dysplasia is a rare cause of intractable partial epilepsy. Abnormalities such as alterations of gyral pattern, thickening of the cortical ribbon, and abnormal magnetic resonance (MR) signal have been described in such entities (1, 2). We report a case of focal cortical dysplasia in which the MR features were correlated with pathologic findings.

Case Report

A 7-year-old boy presented with intractable partial epilepsy since birth. The obstetric history was normal. There were no cutaneous or systemic abnormalities. The first seizure occurred in the first days of life consisting of partial unilateral-clonic fits with left ocular jerks, blinking eyelid, right head deviation, and clonus of the upper limbs. Some of these seizures were associated with transient left hemiplegia. In addition to right occipital lobe spikes, electroencephalography showed multifocal foci. At 16 months of age, an enhanced cerebral computed tomography (CT) scan was normal. MR was performed twice, at 3 years and 4.5 years of age (Fig 1A and B). The findings were unchanged between the first and the second MR examinations and suggested a focal cortical dysplasia. The worsening of the clinical status led to surgery, and resection of the right occipital cortex was performed after intraoperative corticography. The macroscopical and histologic examination revealed a typical focal cortical dysplasia; specifically, the specimen was firm to palpation, and multiple small sulci were present giving a pseudopachygyric appearance of the cortex on pathologic examination. There was an absence of gray-white matter delineation on the cut sections (Fig 1C and D). Histologically, the thickened cortex was disorganized. Numerous binucleated or multinucleated large (up to 40 μm in diameter) heterotopic neurons were visible in the subcortical white matter (Fig 1C). Gemistocytic astroglial cells and microglial cells were located in the molecular layer. No "balloon cells" (very large glial cells) could be recognized. Myelin stain by Luxol fast blue method showed the limit between gray and white matters to be blurred, with myelinic pallor in the subcortical area. With antiepileptic treatment (phenytoin), no seizure occurred after surgery during 2 years' follow-up.

Discussion

Focal cortical dysplasia is a common cause of intractable partial epilepsy. Electroencephalographic abnormalities may not strictly correlate with the macroscopical data (3). Electroencephalography can disclose additional homolateral or contralateral foci without MR abnormality (4). Focal cortical dysplasia consists of congregation of large neurons littered through all but the first layer. In most, but not in all, cases, abnormal large glial cells (balloon cells) also are present in the depth of the affected cortex (1).

The MR findings in focal cortical dysplasia have been previously reported (2, 5, 6). Short-repetition-time sequences can disclose a localized increase of cortical thickness with abnormalities of sulci and gyri. Their internal limit can be irregular. The signal of the abnormal cortex increases as repetition time became longer. On long-repetition-time sequences, the underlying white matter may have an increased signal. The

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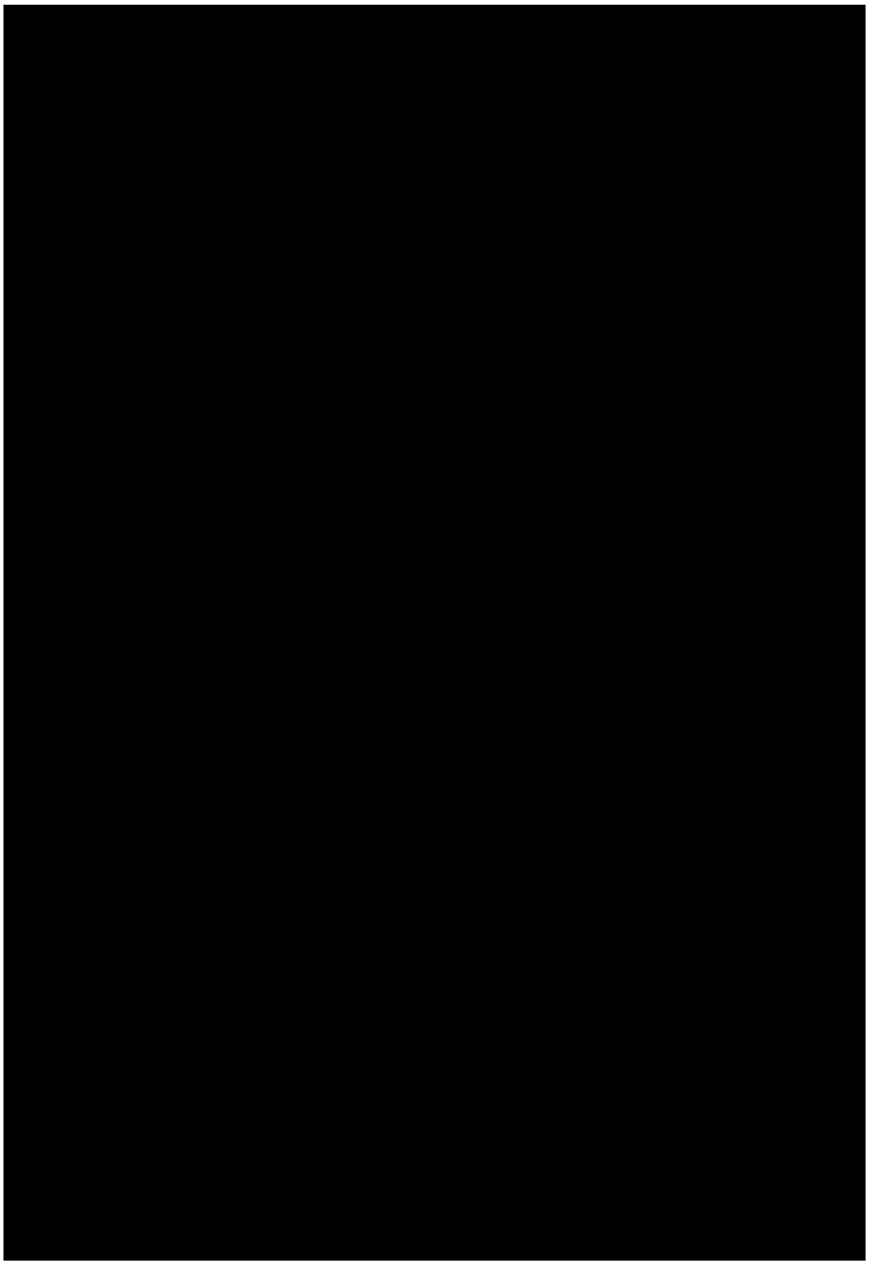

Fig 1. *A,* Axial T1-weighted (480/12/2 [repetition time/echo time/excitations]) MR image. Right parietooccipital cortex is well delineated. The white matter is symetric and normal. There is a mild deformity of the right ventricle.

B, Axial T2-weighted (2000/120/1) MR image. Abnormal signal with a blurring of the cortex–white matter junction throughout the right parietooccipital area. Mild ventricular enlargement on the right.

C, Nissl stain; magnification, ×20; myelin stain section. Disappearance of the normal horizontal layering of the cortex. There is a loss of gray-white delineation because of the numerous heterotopic neurons scattered in the subcortical areas. All subcortical dark points are heterotopic neurons (*arrowheads*). *C* indicates cortex; *WM*, white matter. The arrows indicate approximately the blurred limit between gray and white matter.

D, Luxol fast blue stain; magnification, ×20. Blurred limit between cortex (gray) and white matter (dark gray). Focal myelin pallor (*arrows*).

ventricles are normal or sometimes moderately enlarged. The lesion can be centered around a deep sulcus.

Our case is interesting because the morphology of the right parietooccipital area is normal on T1-weighted sequences. The main abnormality is the focal absence of delineation between gray and white matter, with mild hyperintensity of the white matter on the T2-weighted sequences. This pattern is well correlated with the histologic blurring of the cortex–white matter junction observed throughout the sections of focal cortical dysplasia (7). The origin of the MR signal is complex, involving water, protein and lipid contents, and the relative membrane volume (8). The mild elevation of the right parietooccipital white matter signal on long-repetition-time sequences may be explained by the different constituents of these elements throughout the focal cortical dysplasia area. The large number of neurons scattered in the subcortical white matter may help explain the

myelin pallor seen on histologic examination and mild hypersignal of the white matter on T2-weighted sequences. Other differential diagnoses such as leukodystrophy can be ruled out, because the white matter abnormalities are localized.

References

1. Taylor DC, Falconer Ma, Bruton CJ, Corsellis JAN. Focal dysplasia of the cerebral cortex in epilepsy. *J Neurol Neurosurg Psychiatry* 1971;34:369–387
2. Barkovich AJ, Kjos BO. Nonlissencephalic cortical dysplasias: correlation of imaging findings with clinical deficits. *AJNR Am J Neuroradiol* 1992;13:95–103
3. Palmini A, Andermann F, Olivier A, Tampieri D, Robitaille Y. Focal neuronal migration disorders and intractable partial epilepsy: results of surgical treatment. *Ann Neurol* 1991;30:750–757
4. Guerrini R, Dravet C, Raybaud C, et al. Epilepsy and focal gyral abnormalities detected by MRI: electroclinico-morphological correlations and follow-up. *Dev Med Child Neurol* 1992;34:706–718
5. Sellier N, Kalifa G, Lalande G, et al. Focal cortical dysplasia: a rare cause of epilepsy. *Ann Radiol* 1987;30:439–445
6. Titelbaum DS, Hayward JC, Zimmermann RA. Pachygyric like changes: topographic appearance at MR imaging and CT and correlation with neurologic status. *Radiology* 1989;173:663–667
7. Vinters HV, De Rosa MJ, Farell MA. Neuropathologic study of resected cerebral tissue from patients with infantile spasms. *Epilepsia* 1993;34:772–779
8. Curnes JT, Burger PC, Djang WT, Boyko OB. MR imaging of compact white matter pathways. *AJNR Am J Neuroradiol* 1988;9:1061–1068