A young man with symptomatic epilepsy and right hemianopia

Family affair

Figure 1 Cranial MRI scan

Cranial MRI revealed an irregular, T2-hyperintense, partly gadolinium (Gd)-enhancing left parieto-occipital lesion extending into the corpus callosum.

A 32-year-old man presented with generalized epileptic seizures and right homonymous hemianopia. MRI detected a left parieto-occipital lesion suspicious for lymphoma (figure 1); biopsy showed inflammatory demyelinating white matter lesions suggestive of multiple sclerosis (figure 2, A–C). Hemianopia and neuro-

Figure 2 Histologic analysis of the brain biopsy

Brain biopsy showed inflammatory white matter lesions with increased cellularity and reactive gliosis (A), (confluent) demyelination (B), and macrophages that showed LFB-positive myelin degradation products within their cytoplasm (C).

imaging remained unchanged 12 weeks after high-dose IV steroids. Seizures responded to levetiracetam. A maternal uncle had neurologic impairment leading to infant death; blood screening for very-long-chain fatty acids established the diagnosis of X-adrenoleukodystrophy. Because histologic findings are nonspecific and asymmetric focal brain lesions are a rare manifestation, family history was the key to the diagnosis, considering that severity and age at onset of X-adrenoleukodystrophy may vary.¹

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