Leukoencephalopathy with cerebral calcifications and cysts

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The patient is a 20-year-old woman with an unremarkable family history who began to have focal seizures at age 1 month. Progressive ataxia, generalized dystonia, spasticity, dysarthria, and cognitive decline started at age 14 months. At age 8 years she was wheelchair-bound, and at age 19 years she had severe dementia.

Leukoencephalopathy, brain calcifications, and parenchymal cysts as a particular disorder was first described in 1996. Clinical manifestations include dementia, ataxia, and pyramidal and extrapyramidal findings. Imaging is important in the diagnosis of leukoencephalopathy with cerebral calcifications and cysts, since the combination of these findings is unusual. Gradient echo imaging revealed the extent of the abnormalities not seen by conventional MRI and CT (figure, A, B).

Figure. (A) CT and MRI display calcification of the dentate nuclei, thalamus, basal ganglia, subcortical white matter, leukoencephalopathy, and cysts. (B) Gradient echo images show countless tiny, deep, hypointense, round lesions in the supratentorial white matter, in the periventricular region, and around the cystic lesions in the temporal lobes.

Disclosure: The authors report no conflicts of interest.

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Neurology 2006;66;E32
DOI 10.1212/01.wnl.0000204297.44936.27

This information is current as of May 8, 2006