Adult-onset leukoencephalopathy with intracranial calcifications and cysts (Labrune syndrome)

A 42-year-old woman was admitted for a complex partial seizure. At age 18 years, she had transient diplopia, which was considered a simple strabismus. Current examination revealed a left VI cranial nerve palsy and right arm hyperreflexia. Neuroimaging showed multiple intraparenchymal cystic lesions in white matter (figure), multiple calcifications in basal ganglia, and extensive bilateral leukoencephalopathy. Lack of extraneurologic involvement and genetic testing (CTCI gene) ruled out other disorders (mainly Coats plus syndrome). Leukoencephalopathy with intracranial calcifications and cysts is considered a diffuse microangiopathy affecting children. Adult-onset patients may present a heterogeneous and milder clinical picture, with slow progression.

AUTHOR CONTRIBUTIONS
Rocío-Nur Villar-Quiles: acquisition of data, analysis and interpretation of data, literature review, writing of the manuscript. Natividad Gómez-Ruiz: acquisition of data, literature review, critical revision of manuscript for important intellectual content. Manuela Jorquera-Moya: acquisition of data, analysis and interpretation of data, critical revision of manuscript for important intellectual content. Jorge Matías-Guiu: interpretation of data, critical revision of manuscript for important intellectual content. Jordi A. Matias-Guiu: acquisition of data, analysis and interpretation of data.
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REFERENCES
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