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

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Figure Neuroimaging findings in leukoencephalopathy with intracranial calcifications and cysts (Labrune syndrome)

(A) CT head without contrast axial: scattered calcifications in basal ganglia (1,541 HU, lentiform nucleus), left thalamus, and white matter, frontal cyst, and diffuse low-density areas in white matter. (B) MRI brain axial T2 fluid-attenuated inversion recovery: extensive leukoencephalopathy. (C) MRI brain sagittal reformatted fast spoiled gradient echo T1 3D with IV contrast: left pons cyst without contrast enhancement. (D) Cystic lesions with mild mass effect without vasogenic cerebral edema or restricted diffusion on diffusion-weighted imaging (D) and apparent diffusion coefficient map (E). (F) MRI brain T2 coronal: large cystic lesion in the left pons and leukoencephalopathy.

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.¹ Adult-onset patients may present a heterogeneous and milder clinical picture, with slow progression.²

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, literature review, critical revision of manuscript for important intellectual content. Manuela Jorquera-Moya: acquisition of data, literature review, critical revision of manuscript for important intellectual content. Jorge Matías-Guiu: interpretation of data, critical revision of manuscript for important intellectual content. Jordi A. Matías-Guiu: acquisition of data, analysis and interpretation of

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