Teaching NeuroImages: Myotonic dystrophy type 1

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Figure MRI findings in myotonic dystrophy type 1

(A) MRI brain axial T2 fluid-attenuated inversion recovery: symmetric, confluent hyperintensity in the anterior temporal lobes white matter (arrow). (B) MRI brain axial T2 fluid-attenuated inversion recovery: bilateral hyperintensity in the external capsules (arrow). (C and D) MRI brain axial T1+ gadolinium: no contrast enhancement.

A 51-year-old woman presented with 2 years' history of progressive distal weakness and muscle atrophy. Clinical and electrophysiological myotonia were found. MRI showed bilateral discrete and confluent areas of abnormal hyperintensity throughout the white matter with predominant involvement of the anterior temporal lobes and external capsules (figure). Genetic testing of DMPK revealed more than 50 CTG repeats in 1 allele, confirming the diagnosis of myotonic dystrophy type 1 (DM1). DM1 is the most common muscular dystrophy observed in adults.¹ Though MRI in the late stage of DM1, CADASIL and mitochondrial myopathy may be similar, the clinical history for each condition is very different.²

Author contributions

Bingjun Zhang: study concept and design, acquisition and analysis of data, preparation of manuscript including figures. Yu Yang and Haotian Wu: data analysis and interpretation. Siyuan Liao:

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acquisition and preparation of data. Zhengqi Lu: study concept and design, data acquisition and interpretation, critical revision.

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