Anterior temporal white matter lesions in adult-form myotonic dystrophy type 1

Lesões temporais anteriores da substância branca na forma adulta da distrofia miotônica tipo 1

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A 57-year-old woman presented with a 4-years history of progressive weakness, distal muscular atrophy and myotonia in left hand (Figure 1). Her electromyography had a myotonic pattern. Patient was first diagnosed as paraneoplastic limbic encephalitis based on her brain MRI (Figure 2). Diagnosis was genetically confirmed for myotonic dystrophy type 1 (DM1).

DM1 or Steinert’s disease is an autosomal-dominant disorder characterized by muscle weakness and unusual features, compared with other dystrophies, including myotonia, anticipation, and multiple organ involvement¹,². Anterior temporal lobe subcortical white matter lesions are described in DM1, but not in DM2 patients³. Limbic encephalitis and CADASIL are the most important imaging differential diagnosis.

References


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