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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 paraneoplasic 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 involvement[1,2]. Anterior temporal lobe subcortical white matter lesions are described in DM1, but not in DM2 patients[3]. Limbic encephalitis and CADASIL are the most important imaging differential diagnosis.

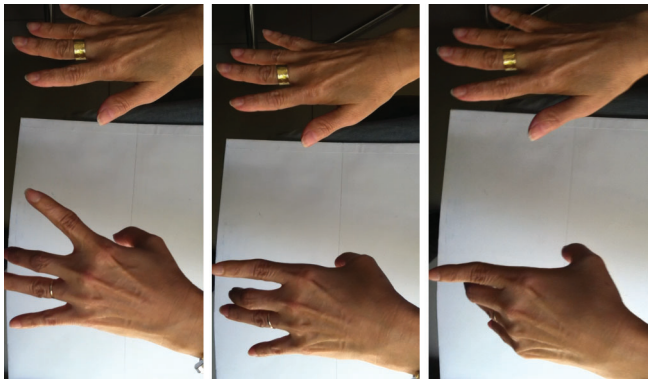

Figure 1. Dystrophy and sequence of myotonia in left hand.

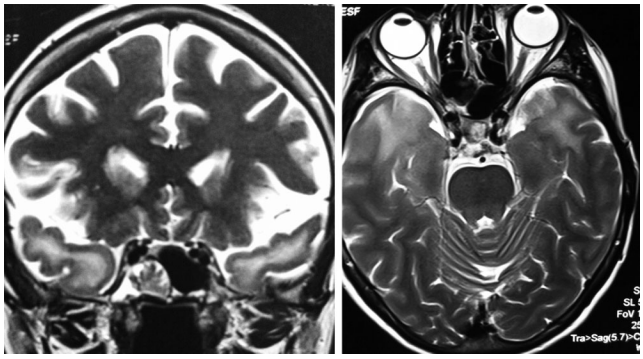

Figure 2. MRI (T2 weight coronal and axial slices) showing bilateral anterior temporal hypersignal in our case. Notice that mesial temporo-limbic structures are not affected.

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