Neuroimaging in myotonic dystrophy type 1

Figure 1 Axial fluid-attenuated inversion recovery image shows symmetric, confluent signal hyperintensity in the anterior temporal lobe white matter

Figure 2 Small basal angle (craniokypnosis), prominent frontal sinus, and diffuse thickening of the skull with ossification of the falx is noted on sagittal T1-weighted images

A 56-year-old woman with a 10-year history of myotonic dystrophy type 1 (MD) presented with progressive lower extremity weakness. MRI showed multiple discrete and confluent areas of abnormal signal intensity throughout the subcortical white matter with predominant involvement of the frontal and anterior temporal lobes (figure 1).

The confluent white matter lesions in the anterior temporal lobes seen in MD can be radiologically confused with cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL). Unlike CADASIL, MD also displays skull abnormalities (figure 2).^{1,2} Both syndromes should be considered in patients with confluent temporal lobe lesions on MRI.

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