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Poster

Measuring encephalic atrophy with an automated method in a SCA3 patient using the SIENA package

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Abstract

Background: Spinocerebellar ataxia type 3 (SCA3) also known as Machado-Joseph disease (MJD) is an autosomal dominant progressive neurologic disorder. It´s caused by a CAG repeat expansion in protein-coding portions of a specific gene (14q24.3-32.1). That expanded allele displays intergenerational instability, particularly at male meiosis, that is associated with the clinical phenomenon of anticipation, whereby the symptoms become apparent at an earlier age and there´s a quicker progression of the disease. Several studies have been able to demonstrate that the brainstem and cerebellar atrophy are the most important features in magnetic resonance imaging (MRI) in SCA3. The SIENA package (part of FSL) is an automated and accurate method for quantifying the encephalic atrophy using two brain MRI studies taken at different points in time. It estimates the percentage of brain volume change (PBVC) between these two MRIs. Objective: The aim of this study was to quantify the atrophy progression in a patient with genetically confirmed SCA3. Methods: We compared two MRI studies of the same patient: a young individual with a high number of trinucleotids (CAG) repeats, therefore showing a rapid symptomatology progression. The sequences were acquired three years apart from each other, in a 3T scanner followed by pre-masking of the images to standard space brain and the two-time-points ("longitudinal") analysis of brain change with the SIENA package. Results: When comparing the 3D images with the SIENA routines it became clear the quick pace of both, the infratentorial and the supratentorial atrophy evolution, demonstrated by a very high PBVC value (-2.41%). Conclusions: Our data provided strong evidence that support the findings of previous studies, revealing a very fast atrophy progression in a young patient with high number of CAG repeats.

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