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Cognitive impairments in patients with Spinocerebellar Ataxia Type 3 (SCA3) in China

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To date there is very few clinical studies published on the cognitive characteristics in patients with SCA3 in China, we sought to evaluate the cognitive function in a cohort of clinically diagnosed and molecularly confirmed patients with SCA3 in China. The neuropsychological tests that were used to evaluate the cognitive function consisted of the Mini-Mental State Examination (MMSE), Clock Drawing Test (CDT), Digit Cancellation Test (DC), Digit Symbol Substitution Test (DSST), Stroop Color-Word Test (SCWT), Trial-Making Test (TMT), Verbal Fluency Test (VFT), and Wechsler Intelligence Scale-Digit Span Test (WISC-DST). The psychiatric symptoms were assessed by the Hamilton Anxiety Scale (HAMA) and Hamilton Depression Scale (HAMD). The severity of motor symptoms was evaluated by the Scale for the Assessment and Rating of Ataxia (SARA). 15 patients with genetically confirmed SCA3 and 15 normal control subjects were enrolled in the study. There was no significant difference in age, gender or educational level among these 2 groups. CDT, DC, DSST, SCWT, TMT, VFT were significantly more impaired in patients with SCA3 than those in the control group. There was no significant difference in the MMSE, DST, HAMA and HAMD between SCA3 patients and controls. In conclusion, our study demonstrates that patients with SCA3 in China present cogintive impairments, manifesting mainly as executive and visuospatial dysfunction.

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