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Tremor syndromes in the elderly: three cases

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Introduction and objective: We report three patients who presented a late onset, slowly progressive tremor syndrome associate to mild parkinsonian, cerebellar and psychiatric features.

PATIENT 1: 82 years old man who presented with gait and balance disturbances associated with bilateral hand tremor, with onset 2 years ago. On examination, we can appreciate mild cerebellar ataxia, dysmetria of left arm and intentional tremor of both hands.

PATIENT 2: 63 years old man who reports tremor of both hands, mild depressive symptoms and behavioral issues which started ten years ago. On examination, he showed bilateral rest and action tremor, mild clumsiness on finger tapping and difficulty in tandem gait walking.

PATIENT 3: 60 years old woman generalized tremor syndrome, which started 8 years ago. On examination, she showed mild bradykinesia in finger tapping bilaterally and rest and action tremor on the four limbs, head and chin. She also reported anxiety and mild depression.

Methods: The patients underwent Brain MRI, routine laboratory testing, neuropsychological assessment and FMR1 gene PCR analysis.

Results: Laboratory workout showed normal findings. Neuropsychological and behavioral assessment of patient 2 reported a control disorder and decrease of motivation without cognitive issues. Brain imaging of patient 1 and 2 showed diffuse cerebellar atrophy and hyperintensity of middle cerebellar peduncles. MRI of patient 3 showed diffuse supratentorial atrophy associated with white matter hyperintensity. Therefore, they underwent genetic testing for Fragile X-associated tremor/ataxia syndrome (FXTAS), that revealed a CGG expansion in the permutation range in FMR1 gene (respectively 88, 106 and 100 CGG repeats).

Conclusion: Fragile X-associated tremor/ataxia syndrome (FXTAS) is a late onset neurodegenerative disorder characterized by progressive ataxia, tremor, cognitive involvement, neuropathy, and autonomic dysfunction. The diagnosis should be considered in elderly patient who present these clinical features with or without family history for Fragile X syndrome (FXS). Brain MRI can provide an important support for diagnosis that must be confirmed by genetic test.