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Poster presentation

Olivopontocerebellar atrophy with dementia syndrome

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Background

Olivopontocerebellar atrophy belongs to a great category of neurodegenerative diseases, which characterized by cerebellar atrophy and mainly brain stem lesions. There are familiar and sporadic cases. The symptoms appear to be obvious at the age of 50 years old mostly. Patients often have limb ataxia, ataxia of brain stem muscles, parkinsonism, motor disorders. The degeneration is detected at cerebellum, septum off pons olive.

Materials and methods

A 65-year-old patient presented a 2-year history of walking instability with continual deterioration, drop attacks with the tendency to fall on his back, speech disturbances and memory loss. He also suffered from an acute psychotic syndrome (delirium type), which lasted for a month and was eased up with the appropriate drug therapy. Past medical history also revealed alcohol consumption until 4 years ago.

Results

The neurological examination showed: 1) A thorough neuropsychological evaluation revealed a remarkable cognitive reduction (MMSE: 14/30), 2) Paralysis of the upper oculogyration and debility of the eyes convergence without oculomotor muscle paresis, 3) Reflexes of the upper and lower extremities (+3), 4) Tremor of the upper extremities, 5) Bradykinesia, 6) Dystonia of the left upper extremity, 7) Dysarthria, 8) Trunk ataxia 9) Romberg test (+) with the tendency to fall on his back, 10) Positive corticomedullar reflexes (palm-chin, lactation) 11) Muscle atrophy of the upper and lower extremities without fasciculations, 12) Mild extra pyramidal hypertonia.

Discussion

Differential diagnosis included: 1) Steele -Richardson-Olszewski syndrome (P.S.P.), 2) Parkinsonic syndrome and 3) Olivopontocerebellar atrophy (O.P.C.A.), sporadic type.

Laboratory and biochemical investigations showed no signs of systematic disease. A brain MRI showed: 1) Extensive atrophy of vermis cerebelli, 2) Atrophy of the pons. Neuroimaging findings, in conjunction with his symptomatology, neuropsychological evaluation and neurological examination led to the diagnosis of "a sporadic type of O.P.C.A. with accompanying dementia".

He was treated with L-Dopa, anticholinergic medication and was asked to visit regularly the dementia outpatient's department of the Neurological clinic.