

Multiple System Atrophy with Primary Progressive Aphasia: A Case Report

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Abstract

Background

Multiple system atrophy (MSA) is a rare neurodegenerative disorder. It is characterized by a combination of parkinsonian syndrome, autonomic failure, rapidly increasing cognitive, cerebellar and pyramidal disorders.

Case Presentation

We describe a patient, who initially complained of a slight difficulty in speech, then rapidly progressing parkinsonian syndrome and memory impairment. We also highlight that she didn't have any symptomatic improvement on levodopa, which do typically provide some relief for patients. The case also highlights the clinical variability and diagnostic difficulties of MSA.

Introduction

MSA is characterized by a predominant lesion of the basal ganglia, nuclei of the brainstem, cerebellar systems, autonomic neurons of the brainstem and spinal cord. The disease most often occurs at 50-60 years of age and is characterized by relatively rapid progression with a fatal outcome within 9 years. MSA accounts for 10-12% of cases of Parkinson's syndrome. The incidence rate is 3 cases per 100 thousand population, i.e. about 20 times less common than Parkinson's disease.

Currently, two types of multisite atrophies are distinguished, depending on the dominance of the cerebellar or akinetic-rigid syndrome in the clinical picture. With a predominance of cerebellar lesions, we have olivopontocerebellar atrophy. In cases where the symptoms of parkinsonism predominate, a diagnosis of striatonigral degeneration is made. In both cases, there

are manifestations of progressive autonomic failure. The disease progresses steadily, postural instability appears very early. In addition, cerebellar and autonomic disorders are observed. The initial symptoms may be subtle, and often the exact time of the onset of the process cannot be suspected. Sometimes even the patient himself and his relatives cannot indicate the cause of the symptoms.

Keywords

Multiple System Atrophy, Olivopontocerebellar Atrophy, Parkinsonism. Neurodegenerative Diseases.

Case Report

A case of multisystem atrophy in a 68-year-old patient T. is presented. Her daughter noticed a violation of speech by the type of trembling, stammering, then gradually stiffness and trembling appeared in her right hand with rapid progression. The daughter provided a video from May 2019, where the patient has free movements during the "jogging", there was no tremor. Only the trembling of speech was disturbing, but it was understandable. At this time, they went to the neurologist, where she was prescribed levodopa 250 mg / day, the condition did not improve.

Also constant headaches appeared, stiffness was progressing, speech became incomprehensible and a memory impairment appeared. She constantly falls mainly to the right, cannot take care of herself, does not control the function of the pelvic organs, wears diapers, does not understand spoken language. The last week the temperature fluctuation in the morning up to 38.5 C
In June 2020, she fell ill with Covid-19 of moderate severity, after which the condition worsens sharply: stiffness progresses, the gait "trampling" when walking has changed, tremor has appeared in the right hand, speech has become incomprehensible, memory impairment is rapidly progressing.

Neurological Status

Not oriented, unresponsive to questions, anxious. She does not follow the hammer, does not follow instructions. The face is symmetrical, the tongue trembles, it cannot stick out the tongue. Choking when swallowing, incomprehensible speech, voice trembling, pronounced proboscis reflex. Tendon reflexes predominate on the right. Muscle tone is sharply increased in a plastic type more on the right.

Pronounced general stiffness and discoordination of movements. Needs help when trying to get up, stomping on the spot when trying to walk, tendency to fall to the right. Periodic increasing tremor in the right hand, like "counting pills". Fresh bruise in the parietal-temporal region on the right, after a fall a week ago.

MRI of the brain 03.10.19 - The subarachnoid space above the convexital surface of the brain and the basal cisternal spaces are markedly enlarged. At the level of the anterior pole of the left temporal lobe, an expansion of the cerebrospinal fluid space is determined, hyperintense on T2 VI with dimensions of 17 * 26 * 23 with clear, even, contours and a homogeneous structure. The topography of the median structures was not changed. The brain stem structures were without signs of pathological changes.

Later MRI of the brain wasn't able to repeat, due to severe cognitive impairment.

Based on complaints, medical history, life, results of neurological examination - cerebellar ataxia, pyramidal syndrome, akinetic-rigid-tremor syndrome, progressive cognitive disorders, the results of paraclinical examination methods, the patient was diagnosed with: MSA with primary progressive aphasia, severe subcortical insufficiency akinetic-rigid-trembling syndrome, pseudobulbar syndrome, severe cognitive impairment to the level of dementia, dysphoric reactions and impaired control of the function of the pelvic organs.

The patient was examined and treated on an outpatient basis.

Conclusion

Thus, in order to establish the diagnosis of multisystem atrophy, it is necessary to strictly take into account the features of the onset and course of the disease, carefully conduct a neurological examination with the advice of related specialists, and analyze the data of neuroimaging examinations.

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