Progressive supranuclear palsy (PSP) is a neurodegenerative disease classified among the atypical forms of parkinsonism. PSP is characterized by great variability in the involvement of different areas of the central nervous system (CNS). The clinical picture is associated with impaired gait and balance, generalized bradykinesia, visual impairment, dysarthria, dysphagia, pelvic incontinence, dementia and others. We present a clinical case of a 50-year-old woman who, at the end of 2021, was admitted to the Rheumatology department, UMBAL „Sv. Ivan Rilski“ – Sofia, due to weakness in the hands, dropping objects, pain in small joints of the hands, disorder in coordination, difficulty walking and frequent stumbling, slurred speech, tremors involving both hands (more pronounced on the right), memory impairment, pelvic incontinence and hair loss.

The clinical case is very indicative of the long journey that a patient with progressive supranuclear palsy takes before being correctly diagnosed.

**Key words:** progressive supranuclear palsy, atypical forms of parkinsonism, MRI
noticed a worsening of the tremor, which involved both hands. In February 2021, an MRI of the brain was performed, from which old gliotic changes of a vascular nature were established. Suspicious myogenic changes in the upper and lower limbs were suspected from the performed ENMG. After hospitalization in a neurology department, the patient was diagnosed with Multiple Sclerosis (MS), treated with CS – with an unsatisfactory effect. Due to worsening in her condition, the patient was hospitalized in another neurological department, where the diagnosis of MS was rejected and she was discharged with a diagnosis of Parkinson’s disease. Therapy with Madopar and Prampexole was started for 1 month – initially with minimal improvement – later – complete absence of effect. SPECT was performed – with the conclusion that there was no evidence of loss of dopaminergic neurons in the striatum. The diagnosis of Parkinson’s disease was rejected and the patient was referred for consultation with a rheumatologist. Due to the progressive muscle weakness, in the clinic of Rheumatology, immunological tests were carried out in the direction of Systemic connective tissue disease – ANA titer 1:160 was established, with a negative ANA immunoblot and Myositis immunoblot, ANCA – negative, Antiphospholipid package – negative. The patient is diagnosed with congenital thrombophilia – Heterozygous carrier for MTHF and heterozygous carrier for PAI, 4G/5G mutation. Abdominal ultrasound showed pronounced liver fibrosis, which is why Wilson’s disease was suspected and was recommended testing of the serum levels of Cu. There was made a consultation with an imaging specialist, who expressed the opinion that the changes in the brainstem from the MRI of the brain were most consistent with progressive supranuclear palsy. The patient was referred to the Clinic for Nervous Diseases, where the diagnosis was confirmed and treatment was started.


Immunological results: ANA – titer 1:160; ANA immunoblot – negative; Myositis immunoblot – negative; ANCA – negative; Antiphospholipid package – negative.


Brain MRI – Irregularly shaped or punctate areas of altered signal intensity are visualized supratentorially bilaterally in the subcortical white matter. The findings are without mass effect and without restriction of the diffusion of water molecules. The ventricular system is symmetrically dilated as in internal non-occlusive hydrocephalus. The subarachnoid spaces and basal cisterns are dilated. A reduction in the volume of the mesencephalon is visualized, which has convex upper contours, forming an image of the „Humming bird sign” type. A thinning of the superior cerebellar pedicules is visualized.

Conclusion: MR data for old gliotic changes of a vascular nature. MR evidence of cerebral atrophy. The described changes in the brain stem are suspicious for Progressive supranuclear palsy.
**Discussion**

The present clinical case is indicative of the long journey that a patient with progressive supranuclear palsy takes before being correctly diagnosed. In clinical practice, the only way to make the correct diagnosis of these patients is on the basis of history, neurological status and correct reading of MRI of the brain [1]. MRI examination is the best diagnostic method for PSP, the most characteristic being the „Hummingbird sign“ and „Mickey Mouse sign“ [3, 4]. „Hummingbird sign“ represents mid-brain atrophy. It is also known as the „penguin sign“ and is used to distinguish PSP from Parkinson’s disease and multisystem atrophy (MSA) [5, 6]. The magnetic resonance parkinsonism index, defined as the ratio of the area of the midbrain to the area of the pons, is another suitable method for the diagnosis of PSP [7, 8]. „Mickey Mouse sign“, also called „Morning glory sign“ describes the image of increased lateral concavity of the midbrain, as a result of atrophy [9, 10, 11].

PSP is a rare neurodegenerative disease affecting patients over 40 years of age, in about 55% of cases male. The differential diagnostic process is an important and difficult stage in this group of patients, due to the heterogeneity of symptoms. It is necessary to rule out other diseases with neurological deficits, including MS, cerebrovasculitis (isolated or in the context of SVT), Wilson’s disease, recent encephalitis, and others. Life expectancy in these patients is about 7 years, with a delay in diagnosis ranging from about 3-4 years [1].

**Библиография / References**