2001; 17(1): 232–4.

Authors:
Vinokurov Mikhail Mikhajlovich, doctor of medical sciences, professor of department of faculty surgery of medical institute of NEFU. 677000, Street of Dzerjinskogo 26 flat 41. Office telephone (43-20-91), home telephone (45-25-30), Yakutsk, Russia. E - Mail: mmv mi@rambler.ru;
Savelev Vyacheslav Vasilievich, candidate of medical sciences, associate professor of department of faculty surgery of medical institute of NEFU. 677000 street of Krupskoj 37 flat 58. Mobile telephone 89241706330. Yakutsk, Russia. E - Mail: vvsaveliev@mail.ru;
Yalinskaya Tatyana Vadimovna, student of the 5th course of medical institute of NEFU. 677009 street of Dzerjinskogo 45/2 flat 70. Mobile telephone 89143033999. Yakutsk, Russia. barbuzyaka@mail.ru.

CLINICAL CASE

A.A. Tappakhov, T.Ya. Nikolaeva, A.D. Alexeeva, L.T. Okoneshnikova, L.A. Klarov, T.K.Davydova

DIFFERENTIAL DIAGNOSIS OF MULTIPLE SYSTEM ATROPHY AND ESSENTIAL TREMOR WITH PARKINSON'S DISEASE (CLINICAL CASES)

ABSTRACT
Differential diagnosis of Parkinson's disease is a difficult task especially at the level of primary medical care. It is one of the reasons for late diagnosis of some neurodegenerative diseases.
The article attracted the attention of doctors to clinical features of diseases similar with Parkinson's disease for early diagnosis and adequate treatment. We provide our own clinical cases of patients with neurodegenerative diseases (multiple system atrophy, essential tremor) who in the debut were diagnosed with Parkinson's disease.
Thus, there is hyperdiagnostics of Parkinson's disease, not only under diagnostics. Some neurodegenerative diseases such as multiple system atrophy, are accompanied by the development of parkinsonism, but they have a number of clinical features that contribute to choosing the right tactics and timely diagnosis. In the differential diagnosis of multiple system atrophy apart from typical clinical picture magnetic resonance imaging is important. Differential diagnosis of tremor form Parkinson's disease and essential tremor is often difficult, especially in the early stages of the disease, when there is no clinically severe rigidity. Timely clinical diagnosis involves the use of optimal methods of treatment based on evidence-based medicine, the identification of reliable epidemiological indicators and, consequently, appropriate use of health care resources.
Keywords: diagnostics, multiple system atrophy, essential tremor, parkinsonism, Parkinson's disease, MRI.

INTRODUCTION

Parkinson's disease (PD) is the most common cause of parkinsonism. Parkinsonism is a clinical syndrome characterized by hypokinesia with rest tremor, muscular rigidity and/or postural instability [1, 4, 5]. If at a later stage of PD patients have stereotypes clinical picture, then at an early stage even skilled experts have difficulty in diagnosis [1]. Therefore, PD should be differentiated with essential and dystonic tremor and other disorders that are accompanied by the development of Parkinsonism. For example, symptomatic parkinsonism may develop as a consequence of stroke or chronic vascular diseases of the brain, and traumatic brain injuries. Parkinsonism may accompany neurodegenerative diseases such as multiple system atrophy (MSA), progressive supranuclear palsy (PSP), and dementia with Lewy bodies (DLB) [1, 10].

Differential diagnosis of Parkinson's disease is a difficult task especially at the level of primary medical care. This is evidenced, for example, by the existing shortage of primary diagnosis of PD, which is associated with both underdiagnosed Parkinsonism and reducing the available symptoms to the natural aging, and insufficient information and late negotiability of the population for health care [3].

Each nosology accompanied by the development of parkinsonism has several distinctive clinical features. Vascular parkinsonism is characterized by a temporary connection with cerebrovascular disease, lesions mainly the lower half of the body, early onset of gait disturbances, symmetrically symptoms and the low efficiency of levodopa [2]. The diagnosis of PSP is considered in cases of early postural instability with falls, early cognitive dysfunction, slowing of vertical saccades, and supranuclear vertical gaze palsy [7]. Various combination of progressive autonomic failure, parkinsonism with the low efficiency of levodopa, cerebellar ataxia, urinary urgency and pyramidal syndrome most often occurs in MSA [8].

Essential tremor (ET) is characterized by slowly progressive isolated tremor without muscular rigidity and hypokinesia, in the most cases patients have family history of disease and the positive effect of alcohol [6, 9].

Aim of study: To focus doctors on clinical features of diseases similar with Parkinson's disease for early diagnosis and adequate treatment. In this article, we present our own clinical cases of patients with neurodegenerative diseases (multiple system atrophy, essential tremor), who in the debut were diagnosed with Parkinson's disease.

RESULTS OF STUDY

Clinical case 1. A 66-year-old man admitted to the neurological department of the Republican Hospital №2 – The Center emergency medical care (Yakutsk city) in August 2015 with

complaints of rigidity, slowness and depletion movements; weakness in the right extremities; intermittent tremor of the hands, especially in the performance of any purposeful movements; unsteadiness of gait; slurring of speech; frequent urination in small portions; fluctuations of blood pressure from 90/... to 160/... mm Hg with syncope.

Medical history: the first symptoms of the disease appeared in 2009 as frequent urination and recurrent syncope when changing position of the body and physical activity. In 2010, the patient noticed a weakness in the right extremities, changes in speech, and slowness of movement. The diagnosis of Parkinson's disease was made. Appointment of levodopa/carbidopa 750/75 mg per day was characterized by positive dynamics in the form of reduced stiffness and increased amplitude of movements. In 2012, the patient reported a progressive unsteadiness of gait and periodic tremors of the hands. Then he began to walk on a walker. Since 2014 patient is taking levopoda/carbidopa 750/75 mg per day and piribedil 150 mg per day.

The patient denies head injury and acute cerebrovascular accident. Heredity on the nervous system disease is not burdened, brother and sister are healthy. Bad habits: smoking for about 50 years.

Neurological status. Clear consciousness. Hypoosmia, lack of response to convergence, hypomimia, easy right-sided central prosoparesis, elements of pseudobulbar syndrome (dysarthria, oral automatism reflexes). Moderate right-sided hemiparesis. Muscle tone is increased in the right extremities on mixed type (the phenomenon of «jackknife» and «cogwheel»), in the left - «lead tube» type. Anisoreflexia, D>S. Babinski symptom positive on both sides. Severe hypokinesia. Left-side hemiataxia. Small-amplitude postural and kinetic tremor of the hands. Walking on the walker on a broad basis. Autonomic failure: arterial hypotension (blood pressure = 100/60 mm Hg) with episodes of syncope, frequent urination, and constipation. MMSE – 27/30.

Blood and urine tests are not significantly disturbed.

MRI of the brain: atrophy of the cerebellum and all of its legs with expansion of the 4th ventricle; atrophy of the pons with expansion tank front axle and the brain cerebellopontine cisterns. Determined pathological signal cross the bridge fibers and fibers forming the seam region of the nuclei, which together form a figure «cross», cross the bridge, in the form of increased signal. Conclusion: MR-signs of multisystem atrophy of the brain (Fig. 1-3).

[Fig. 1]

MRI study of patient B., 67 years old, in the axial projection in T1-weighted images (A), T2* (hemo) (B) and TIRM (C). White arrows - pathological signal of transversal fibers of pons and fibers forming region of nuclei raphes, which forms together cross-figure. Black arrows - atrophy of the cerebellum with the expansion of subarachnoid spaces between its leaves, and the expansion of the 4th ventricle.

[Fig. 2]

MRI study of patient B., 67 years in the median sagittal projection in T1-weighed images. White arrow - flattening the front surface of the pons.

[Fig. 3]

MRI study of patient B., 67 years old in the axial projection in T2-weighted images.

The overall size of the lateral ventricles at 4.4cm, width of 3rd ventricle - 1.3 cm. 4th ventricle has a typical shape and dimensions. Convexital subarachnoid space to the frontal brain and basal cisterns expanded.

Routine EEG: moderate diffuse changes of cortical rhythms with abnormal activation of stem-diencephalic structures.

Daily monitoring of blood pressure: according to the monitoring of blood pressure recorded systolic-diastolic hypertension 1 degree. Maximum blood pressure 160/93 mm Hg, minimum BP – 90/63 mm Hg.

Clinoorthostatic test: blood pressure (lying) = 140/90 mm Hg, heart rate (lying) = 66 minutes; blood pressure (standing)

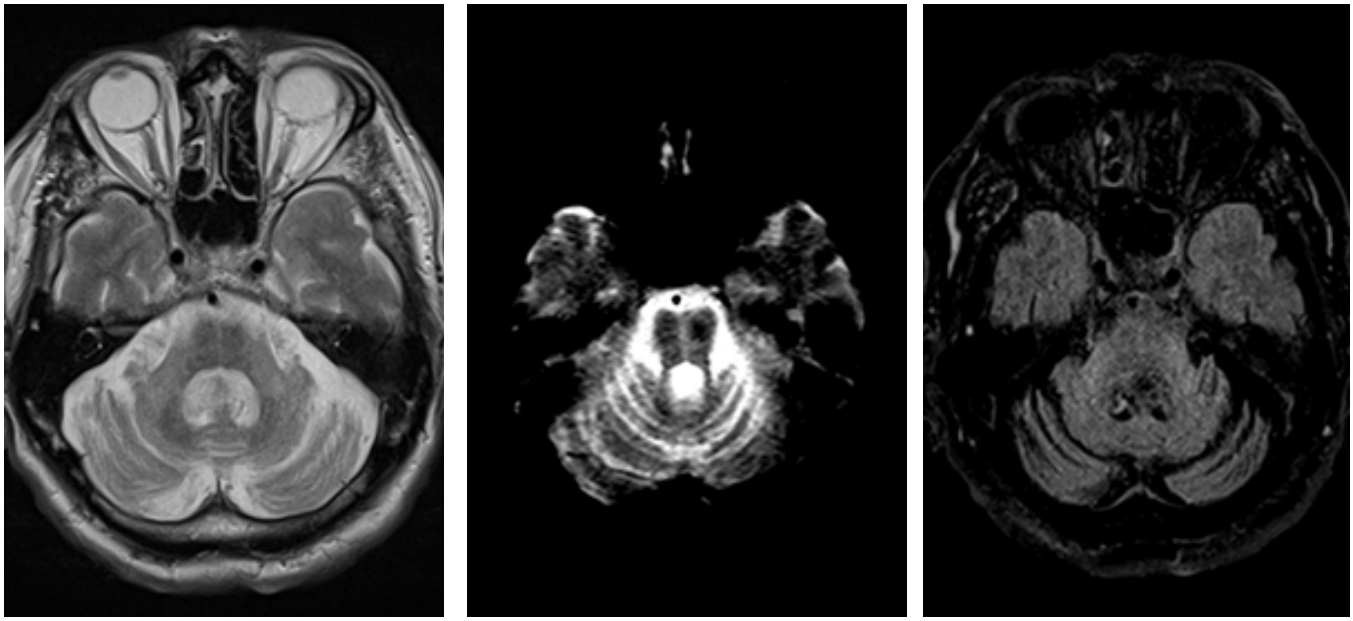

Fig. 1

MRI study of patient B., 67 years old, in the axial projection in T1-weighted images (A), T2* (hemo) (B) and TIRM (C). White arrows - pathological signal of transversal fibers of pons and fibers forming region of nuclei raphes, which forms together cross-figure. Black arrows - atrophy of the cerebellum with the expansion of subarachnoid spaces between its leaves, and the expansion of the 4th ventricle.

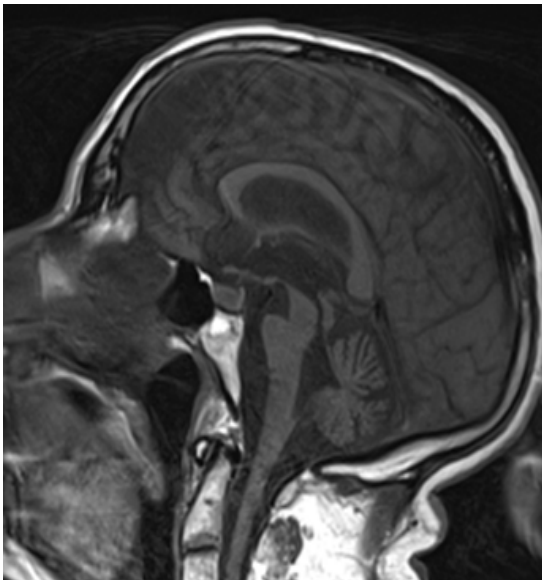

Fig. 2
MRI study of patient B., 67 years in the median sagittal projection in T1-weighed images. White arrow - flattening the front surface of the pons.

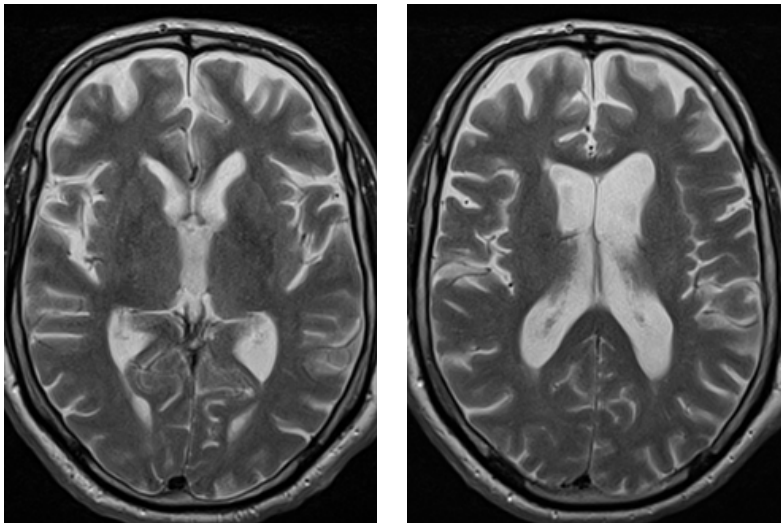

Fig. 3
MRI study of patient B., 67 years old in the axial projection in T2-weighted images.
The overall size of the lateral ventricles at 4.4cm, width of 3rd ventricle - 1.3 cm. 4th ventricle has a typical shape and dimensions. Convexital subarachnoid space to the frontal brain and basal cisterns expanded.

= 115/80 mm Hg, heart rate (standing) = 68 min.

Duplex ultrasound examination of the brachiocephalic artery: ultrasonographic signs of brachiocephalic atherosclerosis without stenosis.

On the basis of medical history and clinical data (onset after 50 years, progressive duration, the lack of family history, a combination of ataxia, pyramidal and pseudobulbar syndromes, parkinsonism, signs of autonomic failure) clinical diagnosis was established: Multiple system atrophy, nigrostrial form, parkinsonism-plus, mild right-sided hemiparesis, left-sided hemiataxia, autonomic failure in the form of arterial hypotension, bladder dysfunction and constipation.

Symptomatic treatment of the patient includes continue receiving levodopa with possible daily dose titrated up to 1000 mg in 4-6 doses. To prevent attacks of syncope is recommended to increase the intake of salt, frequent small feedings, tight bandaging of the lower extremities.

Clinical case 2 demonstrates essential tremor. Patient C., 45 - yrs old woman, asked for a consultation at the Clinic of Medical Institute NEFU in September 2015 with complaints of constant tremor of the head, voice, both hands and the right leg, with increasing excitement, stress. Alcohol leads to a significant reduction in symptoms.

Medical history: The first symptoms appeared in 2007 in the form of periodic small tremor of the head and right hand, especially when the excitement, experience. Since 2009, the tremor was virtually constant spread on the right foot, in 2013 - all the limbs, greatly increased in amplitude. MRI brain without pathology. Parkinsonism was established. The patient took Pronoran without clinical effect.

The patient noted that the father and aunt have Parkinson's disease. Father 76 year-old-man, was diagnosed in 50-year-old, currently he has tremor of the head and limbs, but maintains motor activity. Siblings do not have movement disorders.

Neurological status: Muscle hypotonia. Anisoreflexia, S>D. No paresis. No hypokinesia. Low-amplitude tremor of the head of the type «no-no», «yes-yes», the vocal cords. Mid-amplitude postural and kinetic tremor of both hands, D>S. Mid-amplitude tremor of right leg. Gait is normal.

On the basis of typical complaints, positive reactions to alcohol, slowly progressive duration, no change at neuroimaging, positive family history, neurological status diagnosed was made: Essential tremor, family form, with mixed tremor of the head of the type «no-no», «yes-yes» , vocal cords, and postural and kinetic tremor of both hands, right leg.

Titration of beta-blocker propranolol up to 120 mg / day is recommended with blood pressure and pulse control.

We have also examined the patient's father, who was diagnosed essential tremor too.

Both patients were observed for a long time with Parkinson's disease. Diagnosis upon further examination has not received confirmation, and thorough analysis of complaints, medical history and neurological status allowed exposing other neurodegenerative diseases.

In re-examination in 1.5 months while taking propranolol 60 mg / day is noted positive effect of the disappearance of the tremor of the right leg and the vocal cords, reducing the severity of head tremor.

CONCLUSION

Differential diagnosis of Parkinson's disease with other neurodegenerative diseases, especially with multiple system atrophy and essential tremor, is a difficult and important task. Essential in the differential diagnosis is a neurovisualization's picture, particularly, magnetic resonance imaging, which can explain to some extent symptoms. Timely clinical diagnosis involves the use of optimal methods of treatment based on evidence-based medicine, the identification of reliable epidemiological indicators and, consequently, appropriate use of health care resources.

References

1. Levin O.S. Fedorova N.V. Bolezn Parkinsona [Parkinson's disease] Moscow: MEDPress-inform, 2012. – 352 p.

2. Sayutina S.B. Shpakh V.V., Blokhina I.A. Sosudistyy parkinsonizm [Vascular parkinsonism] Sibirskoye meditsinskoye obozreniye [Siberian medical review]. –

2011. – Vol. 6 – P. 15-18.

3. Problemy diagnostiki i lechenija bolezni Parkinsona v Irkutskoj oblasti [Problems of diagnosis and treatment of Parkinson's disease in Irkutsk region] S.B. Sayutina V.V. Shprakh M.A. Valiulin N.L. Burdukovskaya I.A. Blokhina Sibirskoye meditsinskoye obozreniye [Siberian medical review]. – 2009. – Vol. 7 – P. 173-175.

4. Shnayder N.A. Sapronova M.R. Genetika bolezni Parkinsons [Genetic of Parkinson's disease] Krasnoyarsk: "Kopyrka", 2013. – 108 p.

5. Shtok V.N. Ivanova-Smolenskaya I.A. Levin O.S. Ekstrapiramidnyye rasstroystva: rukovodstvo po diagnostike i lecheniyu [Extrapyramidal disorders: guidelines for diagnosis and treatment] Moscow: MEDPress-inform, 2002. – 608 p.

6. Dalvi A. Tremor: Etiology, Phenomenology, and Clinical Features / A. Dalvi, A. Premkumar // Disease-a-Month. – 2011. – Vol. 57 – P. 109–126.

7. Executive dysfunction is the primary cognitive impairment in progressive supranuclear palsy / A. Gerstenecker, B. Mast, K. Duff, T. J. Ferman, I. Litvan, E.-P. S. Group // Arch Clin Neuropsychol. – 2013. – Vol. 28 – P. 104–113.

8. Second consensus statement on the diagnosis of multiple system atrophy / S. Gilman, G. K. Wenning, P. A. Low et al. // Neurology. – 2008. – Vol. 71 – P. 670–676.

9. Quinn N.P. Tremor – some controversial aspects / N. P. Quinn, S. A. Schneider, P. Schwingenschuh, K. P. Bhatia // Mov. Disord. – 2011. – Vol. 26 – P. 18–23.

10. Williams D.R. Parkinsonian Syndromes / D. R. Williams, I. Litvan // Contin. Lifelong Learn. Neurol. – 2013. – Vol. 19 – P. 1189–1212.

Information about authors

Tappahov Alexei Alexeevich- intramural graduate student of department of neurology and psychiatry Medical Institute NEFU; neurologist MI Clinic NEFU and Neurology Dep. RBN№2 TSEMP

Address: 677000, Yakutsk, ul. Kulakovskogo, 36; tel. 8 (4112) 496700

E-mail: tappakhov@gmail.com; aa.tappakhov@s-vfu.ru

Nikolaeva Tatiana Yakovlevna - PhD, Head of Department of Neurology and Psychiatry Institute of Medicine NEFU

Address: 677000, Yakutsk, ul. Oiunsky 27; tel. 8 (4112) 363046;

E-mail: tyanic@mail.ru

Alekseeva Alena Danilovna - neurologist Neurology Dep. RBN№2-TSEMP

Address: 677005, Yakutsk, ul. P. Alekseeva, 83A; tel. 8 (4112) 432294

E-mail: alyona83@list.ru

Okoneshnikova Lyudmila Timofeevna – Head of neurological department RBN№2-TSEMP

Address: 677005, Yakutsk, ul. P. Alekseeva, 83A; tel. 8 (4112) 432294

E-mail: okoneshnikovalt@rambler.ru

Clarov Leonid Alexandrovich – head of the MRI radiodiagnostics department RBN№2-TSEMP

Address: 677005, Yakutsk, ul. P. Alekseeva, 83A; tel. 8 (4112) 432953

E-mail: pdygyn@mail.ru.