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### CLINICAL CASE OF PRIMARY PROGRESSIVE APHASIA

*In the offered article we discussed rare clinical syndrome – Primary progressive aphasia (PPA). PPA or Progressive motor aphasia is the one from three clinical syndromes which can be observed in frontotemporal lobar degeneration (frontotemporal dementia, PPA, semantic dementia). We observed one 76 years old male patient with PPA. Clinical symptoms and results of inspections coincide with PPA.*

**Keywords:** aphasia, dementia, primary motor aphasia, frontotemporal lobar degeneration.

Primary progressive aphasia (PPA) or progressive motor aphasia is one of the 3 clinical syndromes (progressive circumscribed cerebral atrophy (PCA), PPA, semantic dementia) occurring in Frontotemporal Lobar Degeneration (FTLD). Sometimes, it can develop as the initial symptoms of corticobasal degeneration, Parkinsonism and motor neuron disease. Description of the PPA as a new nosological entity for the first time was described by M. Mesulam in 1982 in his article named "Slowly progressive aphasia without generalized dementia" [1]. In 1998, Neary and et al. developed the diagnostic criteria for clinical syndromes encountered in FTLD [2].

**Epidemiology.** By population-epidemiological studies, the epidemiology of FTLD has been studied in several countries. In Netherlands (Zuid-Holland district), FTLD spread is 2.7 cases per 100,000, and 9.4 cases among people aged 60-69 [3]. In United Kingdom (Cambridge), this indicator has been 15.1/100,000 among the population over the age of 65 [4]. And this is in similar level to the prevalence of Alzheimer's disease among the elderly population. Among people aged 45-64 (3.5/100,000), Alzheimer's disease is less common (4.2/100,000) [5]. In the United States (Minnesota, Rochester), both FTLD and the prevalence of Alzheimer's disease has been the same among people aged 50-59: 3.3/100,000 [6].

**Clinical picture.** PPA has a stealth onset and continues with the next progression. Stammered spontaneous speech followed by at least one of the symptoms such as agrammatism, phonemic paraphasia and amnesic aphasia, remains isolated for at least 2 years, and the main symptom of the disease as it progressing. If, at the beginning of the disease, the speech pathology shows itself as stammering, falter speech, speech-offs, speech tempo changes, and literal paraphasias, later outrage motor or sensomotor aphasia develop. After the primary aphasia contraction, for a few years, other neurological symptoms are not observed, but later disorders of speech are added by behavioral, memory, executive functions and cognitive distortions. In PPA suffering patients, ideomotor apraxia (usually buccofacial), dyscalculia etc. (Joshi et al. 2003) can be found, in which it is recommended to use the term "PPA-plus" (Mesulam 2001). The differential diagnosis should be carried out on expected Alzheimer's disease and the AGD with relative memory preservation.

**Neurovisualization.** In the PPA, cortical atrophy with involvement of the frontal, temporal, and parietal components of speech net in the left hemisphere is observed.

**Treatment.** The only published study of PPA's treatment belongs to Reed et al. (2004). In the small randomized, placebo-controlled study, the effect of dopamine agonist bromocriptine has not been proven. According to some scholars, treatment with trazodone (Lebert et al. 2004), rivastigmine (Moretti et al. 2004), and selegiline (Moretti et al. 2002) can give positive results. Finocchiaro et al. (2006) have indicated the positive results of the efficiency of high frequency repetitive transcranial magnetic stimulation (HF-rTMS), but its exact mechanism is not clear.

**Clinical case.** A right-hander, Caucasian male patient under the age of 76 with the PPA diagnosis has been under our observations. When he consulted to doctor for the first time (2008), he had complaints of faltering, troubles to find words when speaking. The patient considered himself to be ill for more than 1 year. The patient had been diagnosed of cerebral atherosclerosis and treated accordingly. A year later, the patient re-consulted to a neurologist and worsened speech was found.

**History.** For many years, he has worked as a policeman. He was treated with medicamentous therapy with a diagnosis of chronic cholecystitis. He has never had a surgical operation. According to his wife, his parents lived more than 90 years and did not suffer from a similar disease, and died from coronary heart disease. The patient has 3 brothers and 2 sisters. A sister died in 80-year-old of acute myocardial infarction. Any similar pathological process hasn't been observed in any of them. Any gross pathology by the circulatory system, urinary system, respiratory system hasn't been observed. When he was in the examination of neurologist for the first time, weak accommodation and convergence disorder, agrammatism and amnesic aphasia were observed.

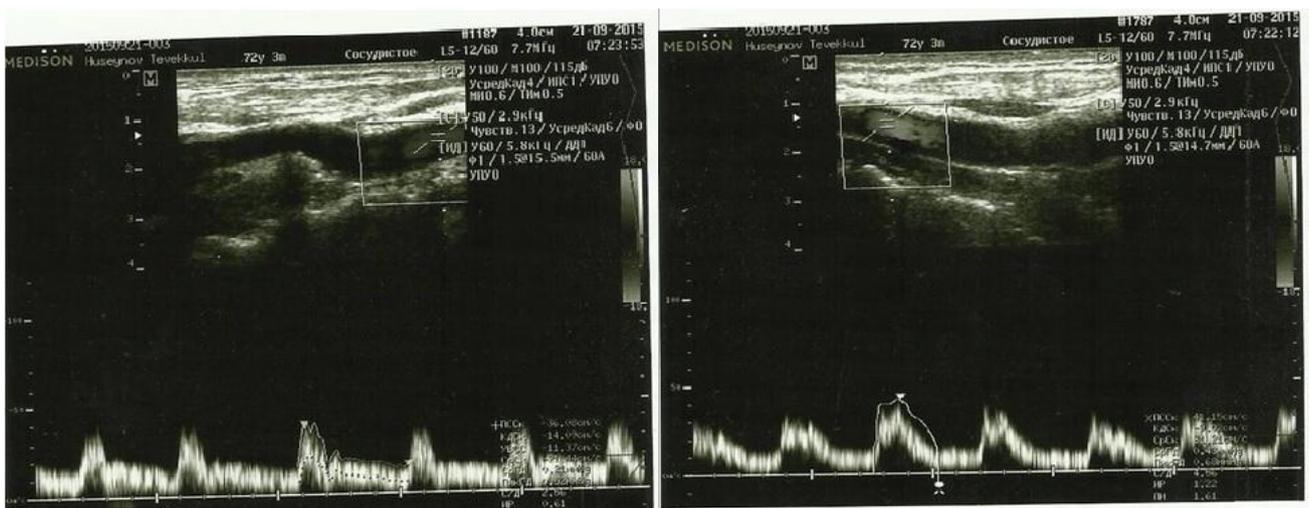
**Examinations.** General blood, urine examination has been normal. Gross deviations from the norm haven't been observed in biochemical blood examination. Through ECG, left ventricular hypertrophy has been found. Rentgenoscopy of the thorax has been with no pathology. LP has been with no pathology.

In native, daytime EEG recorded slowing the basic rhythm around Convex, in the left fronto-parietal-temporal region revealed high-amplitude  $\theta$ -potentials (Fig. 1).



Figure 1 - Electroencephalography of the patient.

In ultrasound examination of the extracranial cerebrovascular system moderate stenosis was found (Figure 2).



Hemodynamic characteristics of the carotid system	CCA		ICA		ECA	
	R	L	R	L	R	L
	Systolic velocity	34.9	41.5	34.4	36	31.7
Diastolic velocity	14.6	9	8	14	9.7	10.3
ICA/CCA	0.98	0.88				
RI	0.58	0.62	0.62	0.61	0.64	0.64

Hemodynamic characteristics of the vertebral-basilar system	R	L
	Systolic velocity	22.94
Diastolic velocity	9.16	13.5
Middle velocity	15.22	23
Volume	52.37	102.84
Overall volume		155.21
RI	0.64	0.67

Figure 2 - Ultrasound examination of extracranial arteries, moderate stenosis (<50%).

Through MRI of brain, progressive circumscribed cerebral atrophy was found (Figure 3, 4, 5.).

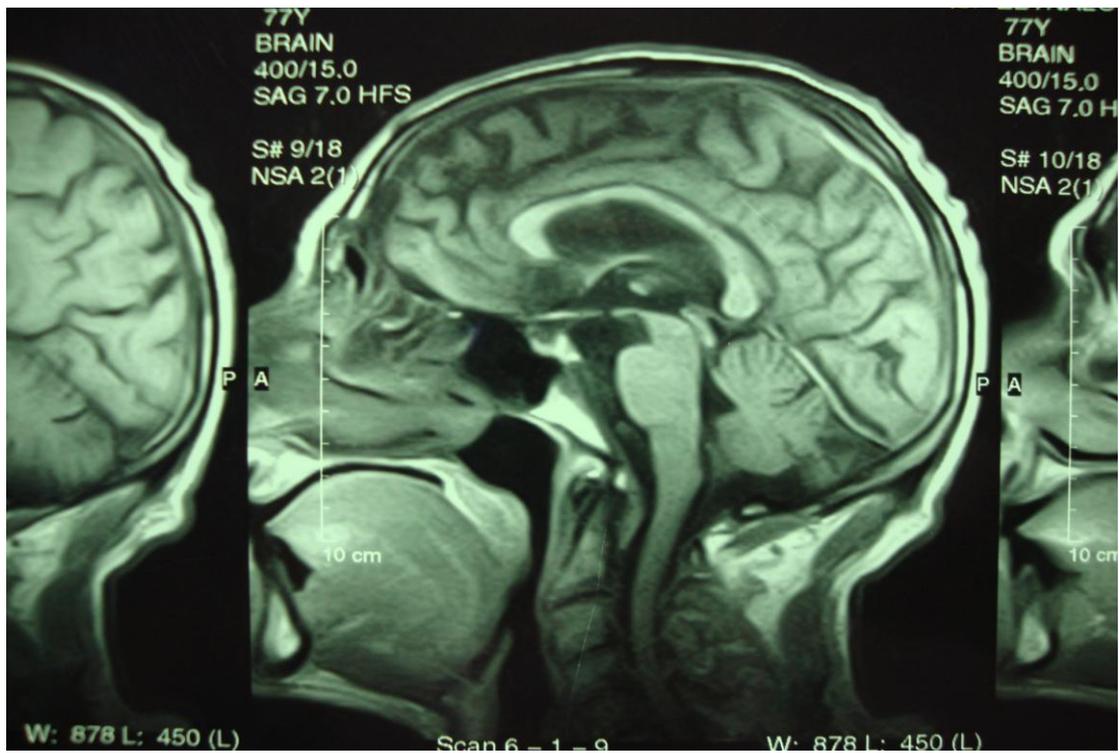


Figure 3 - MRI examination of the patient with primary progressive aphasia (PPA). Sagittal incision. T1 mode.

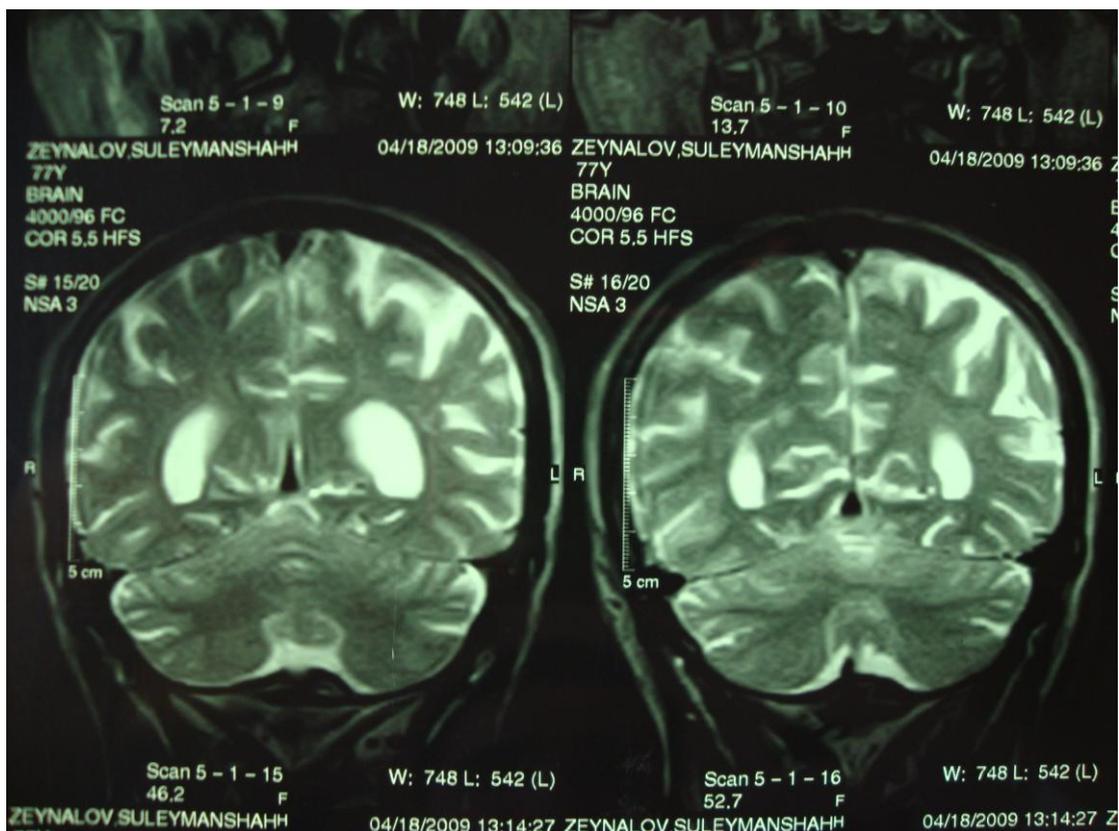


Figure 4 - MRI examination of the patient with primary progressive aphasia (PPA). Frontal incision. T2 mode.

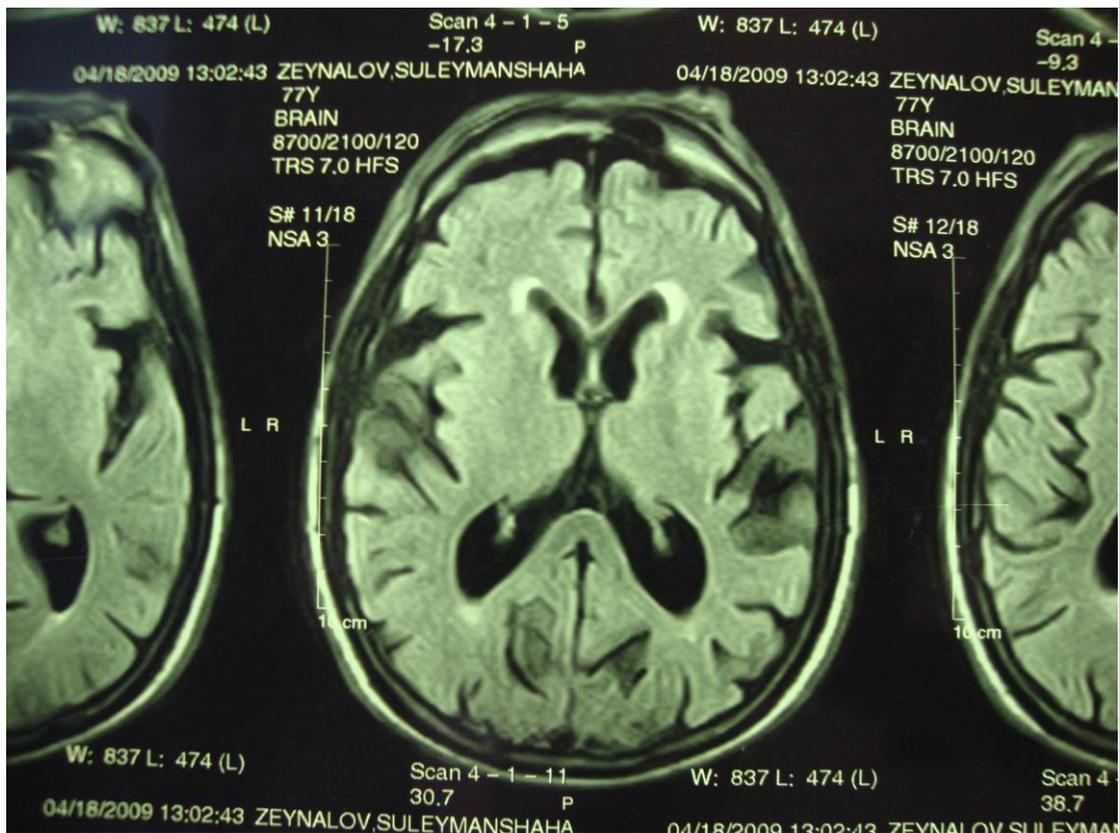


Figure 5 - MRI examination of the patient with primary progressive aphasia (PPA). Axial incisions. T2 FLAIR.

**Treatment.** The patient has been prescribed aspirin (100mg:0:0), gliatilin (400mg:400mg:400mg) and bromocriptine (0:0:1.25). As the drugs gave no effects, the patient spontaneously stopped taking it. About a year later (2009), the patient was re-consulted to neurologist with deepened amnesic aphasia and agrammatism. Gnosis and praxis were kept. The patient was re-prescribed of the previous treatment and galantamine (5mg:0:5mg) was added. However, the patient did not follow the treatment for long-term. Nearly 2 years after the start of symptoms of speech disorders for the first time (2010), through repeated examination, rough sensomotor aphasia, agraphia was observed in the patient. Course of the disease is compatible with the course of PPA. However, after two years of onset first symptom (speech problem), other cognitive problems appear – agrammatism, agraphia.

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#### **БІРІНШІЛІК ҮДЕМЕЛІ АФАЗИЯНЫҢ КЛИНИКАЛЫҚ ЖАҒДАЙЫ**

**Түйін:** Мақалада сирек кездесетін синдромдардың бірі берілген – біріншілік үдемелі афазия синдромы (БҰА). БҰА немесе үдемелі моторлы афазия маңдай-самай бөлімдерінің дегенерациясында кездесетін 3 клиникалық синдромның (маңдай-самайлық деменция, БҰА, семантикалық деменция) бірі болып табылады. Біздің бақылауымызда БҰА диагнозымен 76 жасар ер адам болды. Клиникалық ағымы және зерттеу нәтижелері БҰА ағымына сәйкес келеді.

**Түйінді сөздер:** афазия, деменция, біріншілік моторлы афазия, маңдай-самай бөлімдерінің дегенерациясы.

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#### **КЛИНИЧЕСКИЙ СЛУЧАЙ ПЕРВИЧНОЙ ПРОГРЕССИРУЮЩЕЙ АФАЗИИ**

**Резюме:** В данной статье представлен один из наиболее редко встречаемых синдромов – синдром первичной прогрессирующей афазии (ППА). ППА или прогрессирующая моторная афазия является одним из 3 клинических синдромов (лобно-височная деменция, ППА, семантическая деменция), встречаемых при дегенерации лобно-височных долей. Под нашим наблюдением находился больной мужчина 76 лет, с диагнозом ППА. Клиническое течение и результаты обследований соответствуют течению ППА.

**Ключевые слова:** афазия, деменция, первичная моторная афазия, дегенерация лобно-височных долей.