



Verbal communication disorders in a patient with diagnosed progressive supranuclear palsy

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Progressive supranuclear palsy [Steele-Richardson-Olszewski] is a neurodegenerative disease of the central nervous system. It develops mainly in men over 40 years of age. The course of PSP is rather characteristic: motor, cognitive and non-cognitive impairments develop at a fairly rapid rate. The aim of the study is to assess inter-actio- n- al, communicative and linguistic competencies and skills, executive abilities (breathing, phonation and articulation) as well as primary functions of a patient diagnosed with PSP. The article confirms the thesis that dementia disorders overlap with motor disorders in the course of PSP.

KEYWORDS: progressive supranuclear palsy, dysarthria, verbal communication, non-verbal communication, dementia

1. Introduction

Progressive supranuclear ophthalmoplegia [Steele-Richardson-Olszewski] (ICD-10, G23.1) is a neurodegenerative extrapyramidal disease of the central nervous system, which develops mainly

in men over 40 years of age.¹ Most people affected by this disease experience cognitive impairment which increases at a fairly rapid rate. PSP is sometimes associated with subcortical dementia, fronto-subcortical dementia, corticobasal degeneration and frontotemporal dementia.² The course of PSP is rather characteristic, the first period is accompanied by hypokinesia and postural instability, slightly less frequently by pseudobulbar palsy, often by disorders of voluntary eye movements, axial parkinsonism, dysphagia and dystonia (commonly at neck); bilateral pyramidal and cerebellar symptoms may appear in the late phase.³ The picture of disorders is dominated by executive function disorders, slowed thinking as well as attention

¹ Conf. P. Liberski, T. Sobów, B. Sikorska, *Postępujące zwyrodnienie nadjądrowe (choroba Steele-Richardsona-Olszewskiego)*, [in:] *Neuropatologia Mossakowskiego*, ed. by P.P. Liberski, W. Papierz, Wydawnictwo CZELEJ, Lublin 2005, p. 539; J.C. Steele, J.C. Richardson, J. Olszewski, *Progressive supranuclear palsy; a heterogenous degeneration involving the brain stem, ganglia and cerebellum with vertical gaze and pseudobulbar palsy, nuchal dystonia and dementia*, „Archives of Neurology” 1963, no. 10, p. 333-359.

² I. Gałkowska, *Diagnoza dyzartrii u dorosłych w neurologii klinicznej*, Wydawnictwo UJ, Kraków 2012, p. 97.; K. Jendroska, M.N. Rossor, C.J. Mathias, S.E. Daniel, *Morphological overlap between corticobasal degeneration and Pick's disease: a clinicopathological report*, „Movement” 1995, 10(1), pp. 111-114; A. Kertesz, W. Davidson, D.G. Munoz, *Clinical and pathological overlap between frontotemporal dementia, primary progressive aphasia and corticobasal degeneration: The Pick complex*, „Neurology” 1999, no. 10(1), pp. 46-49; H. Olszewski, *Ołpienie czołowo-skroniowe. Ujęcie neuropsychologiczne*, Impuls, Kraków 2008, p. 179; M. Pąchalska, H. Kurzbauer, B. Grochmal-Bach, B.D. MacQueen, A. Urbanik, I. Herman-Sucharska, *Nietypowe zaburzenia języka i mowy u pacjentki z klinicznym rozpoznaniem zespołu Steele'a-Richardsona-Olszewskiego*, [in:] *Choroby ołpienne. Teoria i praktyka*, ed. by J. Leszek, Continuo, Wrocław 2011, p. 271.

³ Conf. P. Liberski, T. Sobów, B. Sikorska, *Postępujące...*, p. 539; B. Morales, A. Martinez, I. Gonzalo, L. Vidal, R. Ros, E. Gomez-Tortosa, A. Rabano, *Steele-Richardson-Olszewski syndrome in a patient with a single C212Y mutation in the parkin protein*, „Movement Disorders in Neurologic and Systemic Disease” 2002, no. 17(6), pp. 1374-1380; H.R. Morris, M. Baker, K. Yasojima, H. Houlden, M.N. Khan, N.W. Wood, J. Hardy, M. Grossman, J. Trojanowski, T. Revesz, E.H. Bigio, C. Bergeron, J.C. Janssen, P.L. McGeer, M.N. Rossor, A.J. Lees, P.L. Lantos, M. Hutton, *Analysis of tau haplotypes in Pick's disease*, „Neurology” 2002, no. 59(3), pp. 443-445; I. Litvan, Y. Agid, D. Calne, G. Campbell, B. Dubois, R.C. Duvoisin, C.G. Goetz, L.I. Golbe, J. Grafman, J.H. Growdon, M. Hallett, J. Jankovic, N.P. Quinn, E. Tolosa, D.S. Zee, *Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richard-*

and memory impairments. In addition, these disorders are often accompanied by non-cognitive changes, such as apathy, abulia, depressed mood, emotional disorders and hypochondria. The average survival of PSP patients is usually between 2 and 6 years after the onset of the disease.⁴

The aim of this study is to assess: 1) interactional, communicative and linguistic competencies and skills as well as the ability to read, write and count in the context of cognitive functioning, 2) executive abilities (breathing, phonation and articulation) 3) primary functions of a patient diagnosed with PSP.⁵

2. Methods

The article uses research material on a 62-year-old man.⁶ The research was conducted in a home environment during several meetings.

The Mini Mental State Examination (MMSE)⁷ and the Clock Completion Test were used for the screening assessment of cognitive functioning. In order to assess interactional, communicative and linguistic competencies and skills the author used observation, data from medical records, psychometric testing, clinical and experimental tests. They include a test elaborated by J. Szumska included in the publication titled *Metody badania afazji*,⁸ selected tests from the

son-Olszewski syndrome): report of the NINDS-SPSP international workshop, „Neurology” 1996, no. 47(1), pp. 616–117.

⁴ Conf. M. Pączalska, H. Kurzbauer., B. Grochmal-Bach, B.D. MacQueen, A. Urbanik, I. Herman-Sucharska, *Nietypowe...*, p. 271; P. Liberski, T. Sobów, B. Sikorska, *Postępujące...*, op. cit., p. 539.

⁵ J. Panasiuk, *Postępowanie logopedyczne w przypadkach chorób neurodegeneracyjnych*, [in:] *Logopedia. Standardy postępowania logopedycznego*, ed. by S. Grabias, J. Panasiuk, T. Woźniak, Wydawnictwo UMCS, Lublin 2015, pp. 955–995.

⁶ The author of this article has obtained permission to conduct the study and to publish it.

⁷ M.F. Folstein, S.E. Folstein, G. Fanjiang, *Mini Mental State Examination. Polska normalizacja*, Warszawa, 1975.

⁸ J. Szumska, *Metody badania afazji*, Wydawnictwo PZWL, Warszawa 1980.

neuropsychological study by E. M. Szepietowska,⁹ a test from the Boston Diagnostic Aphasia Examination (BDAE) (assessment of comprehension: complex language material, tests: 4-8, 11-12);¹⁰ the verbal fluency test (VFT).¹¹ For the assessment of executive disorders, the article relies on the author's tests developed on the basis of the works by O. Jauer-Niworowska and J. Kwasiborska¹², O. Jauer-Niworowska¹³, I. Gatkowska¹⁴ and Z. Tarkowski.¹⁵

3. Case study

The patient has a secondary education and he worked as a clerk before being diagnosed with the disease. He is right-handed. He had a myocardial infarction in 2000. The first alarming symptoms of the disease concerned difficulties in maintaining balance, the subject's gait gradually became clumsy and slow, and for a short period the patient was suspected of having alcohol problems. No other disorders were observed throughout that period. The breakthrough took

⁹ E.M. Szepietowska, *Badanie neuropsychologiczne. Procedura i ocena*, Wydawnictwo UMCS, Lublin 2000, p. 23.

¹⁰ H. Goodglass, E. Kaplan, *Boston Diagnostic Aphasia Examination*, Lea & Febiger, Philadelphia 1972.

¹¹ Conf. E.M. Szepietowska, B. Gawda, *Ścieszkami fluencji werbalnej*, Wydawnictwo UMCS, Lublin 2011; E.M. Szepietowska, T. Hasiąg, A. Jańczyk-Mikoś, *Fluencja słowna werbalna i niewerbalna w różnych stadiach i formach choroby Parkinsona*, „Psychogeriatrya Polska” 2012, no. 9(4), pp. 137-148; M. Piskunowicz, M. Bieliński, A. Zgliński, A. Borkowska, *Testy fluencji słownej – zastosowanie w diagnostyce neuropsychologicznej*, „Psychiatria Polska” 2013, vol. XLVII, no. 3, pp. 475-485; R. Gliwa, *Fluencja słowna w otępieniu naczyniopochodnym – ujęcie kliniczne*, [in:] *Teoria i praktyka logopedyczna. Wybrane zagadnienia*, ed. by E. Gacka, M. Kaźmierczak, Wydawnictwo UŁ, Łódź 2018, pp. 83-102.

¹² O. Jauer-Niworowska, J. Kwasiborska, *Dyzartria. Wskazówki do diagnozy różnicowej poszczególnych typów dyzartrii*, Komlogo, Gliwice 2009.

¹³ O. Jauer-Niworowska, *Dyzartria nabyta. Diagnoza logopedyczna i terapia osób dorosłych*, Wydawnictwo APS, Warszawa 2009.

¹⁴ I. Gatkowska, *Diagnoza....*

¹⁵ Z. Tarkowski, *Dyzartria*, Wydawnictwo Fundacji Orator, Lublin 1999.

place when the patient caused a car accident and was taken to hospital due to his injuries. MR examination of the brain showed no post-traumatic changes, and the diagnosis revealed a small vascular lesion in the frontal region, a slight widening of the left lateral ventricle as well as cerebral cortical and subcortical atrophy (mainly in the frontal and temporal lobes). The clinical picture indicated motor disturbances and clear difficulties in producing speech manifested by a significant slowing down of its pace.

The patient underwent rehabilitation yet the neurological symptoms in the motor sphere began to worsen. Despite good results in tests assessing cognitive functions, the wife of the subject began to notice significant changes in communication with the patient within one year of the accident: she had the impression that her husband did not understand the messages directed to him, and that his behavior lacked rationality.

As the neurological symptoms did not subside, the examinations were continued. Initially, the patient was diagnosed with Parkinson's disease, yet dopaminergic treatment proved ineffective, thus multiple system atrophy-parkinsonian type (MSA-P), a condition with predominant parkinsonian symptoms, was suspected.

The diagnosis was revised when the subject's wife noticed that he had developed a tendency to make quite specific head movements to look at something below or above his eyes. It was then that the suspicion of PSP was raised.

The subject was hospitalised again after approximately one and a half years due to exacerbation of the extrapyramidal syndrome symptoms. Neurological examination revealed dysarthria, abnormal upward movement of the eyeballs, hypomimia, bradykinesia, axial rigidity, increased muscle tone of extrapyramidal type in upper and lower limbs, Rossolimo's sign positive bilaterally, abnormal parkinsonian-ataxic gait with a tendency to fall to the left. A psychological consultation from this period indicates an organic basis for the disorders manifested by impaired cognitive processes, difficulty in focusing on the conversation, episodic and short-term memory disorders, impaired autopsychic and allopsychic orientation.

One year later, the patient was again admitted to a neurology clinic due to severe swallowing difficulties which persisted for a period of two weeks. A neurological examination confirmed PSP and all lesions were described as typical of cortical-subcortical dementia syndrome.¹⁶ No swallowing disorders were found in swallowing tests. The patient was prescribed a mush diet and meals were thickened with rice gruel. The neuropsychological examination diagnosed general memory impairment, organic mood disorder, the patient's state of consciousness was assessed as confused, with a tendency to risky behaviour.

Four years after the occurrence of the first symptoms of the disease, the patient scored 35 points on the Barthel scale. He was unable to perform self-care activities independently. The patient moves around in a wheelchair. His current medical records indicate the following diagnoses: PSP, Parkinson's disease, post-myocardial infarction status.

The subject is prescribed Madopar, sedative and anti-anxiety drugs as well as cholesterol-lowering and antihypertensive medications.

4. Research results

4.1. Motor assessment of articulatory muscles and selected primary functions

The dysarthria observed in the patient is of a spastic nature, although occasional features typical of the hyperkinetic-hypokinetic form can be seen.

The patient is diagnosed with an abnormal peak respiratory trajectory which causes a significant shortening of the respiratory phase and, as a result, the phonation phase. Examining the subject's phonatory abilities, the author observed some abnormalities in the ability to initiate the emission and pauses in phonation. Major

¹⁶ Vascular dementia was excluded on the basis of a proton spectroscopy study.

changes were also observed in the quality of the voice: the patient's voice is effortful, hoarse, quiet, with no possibility of modulation or gradation of its diameter.

Changes in prosody mainly concern slowing down the speech rate, standardisation of the accent and exaggerated amplification of the sound of speech in moments of nervousness.

The musculature of the face and the articulatory apparatus of the subject at rest indicates the presence of increased muscle tone. It involves the strongest of the masticatory muscles, namely the masseter and temporalis muscles, causing severe trismus. The temporomandibular joints are also affected, causing a significant distortion of the sounds produced.

Large dysfunctions were observed in the area of the the orbicularis oris muscle, the superior and inferior incisive muscles and the zygomaticus major muscles. The observed excessive tension in, among others, the risorius and the levator labii superioris alaeque nasi muscle results in the persistence of facial expressions indicating disgust, anger and distress. Significant muscle tension is also present in the cheek muscles and the depressor anguli oris, giving an expression of bitterness and resignation. The subject is unable to make purposeful movements that require the depressor labii inferioris and the mentalis to work together. Increased muscle tone is observed in the areas of the platysma muscle as well as the suprahyoid and infrahyoid muscles, which may result in the appearance of pressed voice.

Negative changes were indicated by the tests assessing the work of the tongue muscles (external muscles – genioglossus, hyoglossus or styloglossus and internal – longitudinal, inferior longitudinal lower, transverse and vertical muscles). Increased muscle tone was observed in the area of the levator and the tensor veli palatini.

The subject is diagnosed with dysphagia.¹⁷ He has very serious problems processing a bite of solid food and transporting semi-liquid and liquid food towards the throat. Part of the food intake re-

¹⁷ Conf. P. Stręk, *Diagnostyka dysfagii ustno-gardłowej*, „Terapia” 2002, no. 10(2), pp. 12–15; J. Tomik, B. Solowska, *Zaburzenia połykania*, „Neurolingwistyka Praktyczna” 2015, no. 1, pp. 27–41.

mains in the mouth, chewing time is prolonged, liquid food is spit up. Although the swallowing reflex is initiated, it is significantly slowed down. There is a significant weakening of the defensive reflexes, such as grunting and coughing.

4.2. Mini Mental State Examination

The test results indicate the presence of mild dementia. The subject has preserved auto- and allopsychic orientation.

4.3. Clock Completion Test

The subject had significant problems with the correct placement of the hands of the clock and the correct marking of the indicated time, which suggests the existence of, *inter alia*, impaired ability to plan and visualise, visual memory and motor skills disorders, and consequently visual-spatial disorders as well as abstract-conceptual thinking disorders.¹⁸

4.4. Assessment of the quality of non-verbal communication

The subject obtained 2 points in the Nonverbal Communication Scale.¹⁹ Due to significant motor dysfunctions, he does not usually use non-verbal communication.

4.5. Assessment of comprehension

Tests aimed at assessing speech comprehension showed no change in the comprehension of single words, simple sentences and short stories. The patient did not recognise linguistic situational humour. The observed difficulties in responding adequately to

¹⁸ Conf. S. Krzywiński, *Test rysowania zegara*, „Postępy Psychiatrii i Neurologii” 1995, no. 4 I(2), pp. 21–30.

¹⁹ M. Pączalska, *Afazjologia*, Wydawnictwo Naukowe PWN, Warszawa-Kraków 1999, p. 270.

long extended instructions result from impaired concentration and memory, and – to a lesser extent – from a lack of understanding of grammatical and semantic relations between words.

4.6. Evaluation of dialogue and monologue skills

The structure of the dialogue co-created by the patient was very disturbed.²⁰ The patient's speech was significantly impoverished and he did not initiate independent, spontaneous verbal contact. He limited himself to signalling his immediate needs verbally (in the form of imperatives and simple sentences). He tended to divide longer utterances to make up for the lack of air, but usually failed to answer questions that required a longer utterance.

The patient did not construct monologues and answered leading questions reluctantly and with single words. He did not have the ability to make a holistic assessment of the image which he saw, he failed to perceive and take into account cause and effect. The way in which he speaks indicates a clear impoverishment of his active vocabulary.

The speech of the subject revealed a lack of word fluency, semantic paraphrases, perseverations, a tendency to template utterances, the use of a telegraphic style, chanted speech, sometimes pronominal speech, a lack of word readiness, palilalia, logoclonia, a tendency to echolalic repetition, the presence of indefinite phrases and grunts.

4.7. Assessment of the implementation of automated statements

No pathological changes were indicated by tests aimed at assessing the extent to which the subject is able to reproduce verbal automatisms that require the involvement of right-hemispheric strategies.²¹ During the realisation of the sequences, it was possible to

²⁰ Conf. J. Warchala, *Dialog potoczny a tekst*, Wydawnictwo UŚ, Katowice 1991.

²¹ E. Szepietowska, J. Lipian, *Fluencja słowna neutralna i afektywna u chorych z uszkodzeniem prawej, lewej lub obu półkul mózgu*, „Psychiatria Polska” 2012, no. XLVI(4), pp. 539–551.

observe articulatory distortions of the produced sounds caused by dysarthric disorders, a slowing down of speech pace as well as a lack of coordination of breathing, phonation and articulation.

4.8. Assessment of repetition

The subject correctly repeated isolated vowels that did not require tongue elevation, similar results were obtained with regard to the repetition of syllables. He repeated 2-syllable and 3-syllable words relatively well. Deficits were observed in longer forms, containing consonant clusters, which were most often "broken" by a vowel. The number of words repeated was limited (usually up to three), chiefly due to impaired attention and memory as well as weakened emotional-motivational processes. The patient repeated only one structural neologism, the ability to repeat them being considered a measure of pure phonological processing.

4.9. The naming test

The subject was presented with 50 illustrations representing 10 semantic categories, of which he named 26 illustrations correctly (8 of them with a significant delay) and 24 incorrectly. The errors observed concerned the use of the superordinate category name (4), an incorrect name but belonging to the same semantic category (4), an incorrect name not belonging to the semantic category concerned (2), perception errors (2), perseveration (1), the use of descriptive constructions (9), no answer or the answer "I don't know" (2).

The subject did not use gesticulation and he did not use ideomotor praxis.

4.10. Assessment of verbal fluency

The subject mentioned 6 names in the category of animal names, of which 3 were repetitions, he named 4 words in the category of sharp objects, of which 2 were semantic errors; he produced only

3 words beginning with the “a” sound and 3 words beginning with the “k” sound in a formal fluency test. The level of performance in semantic and formal verbal fluency tests demonstrates impaired access to lexical and semantic long-term memory resources, significant impairment of executive functions as well as the functions of voluntary attention and working memory, which coordinate and monitor task performance. The subject’s responses are characterised by considerably impoverished fluency, which is manifested by a small number of elements mentioned, the presence of rather long pauses and repetitions.

4.11. Assessment of reading

During reading attempts, very severe segmental and suprasegmental abnormalities were observed, the reading speed was significantly slowed down. Numerous phonetic distortions were noted. The subject did not usually attempt to read words with more than 3-4 syllables or read only the initial word sounds, and he found it difficult to read words saturated with consonant clusters, which he usually broke up with a vowel.

The observed disorders suggest that the lexical deficits are due to cognitive, executive and motor dysfunctions involved in the reading process.

4.12. Assessment of handwriting

In all the conducted tests (rewriting, dictation, writing from memory, spontaneous writing), it was possible to observe a significant reduction in the legibility of the patient’s handwriting, numerous graphic errors, a very significant slowing down of the writing pace, difficulty in keeping the handwriting in one line, very little fluency and freedom of movement of the lines, micrographia and high fatigue of the subject. Only occasional and very short dictated texts were transcribed correctly, although they were significantly distorted graphically. When writing sentences, the subject omitted

letters and words. The patient did not attempt creative writing, he was unable to construct a sentence independently.

5. Speech therapy diagnosis

Conducting a general assessment of the subject's functioning in the context of cognitive and non-cognitive changes, it should be noted that the changes observed result from dementia that develops together with the progression of PSP. This dementia is reminiscent of the picture of behavioural frontotemporal dementia with pathological changes in language and communication skills as well as dysarthria and dysphagia.

Functioning of the patient is predetermined by disorders in the emotional sphere: the lack of efficient motivational mechanisms contributes to the loss of interest in the world around him and in any activity. The subject lacks the ability to plan both simple and complex activities, which is indicative of severely disturbed executive mechanisms. His functioning is impeded by impaired concentration, reduced speed of information processing, limited ability to process the verbal material and reduced learning efficiency.²² The patient has demonstrated selective disorders of linguistic competence as well as disorders of motoric pronunciation mechanisms. Furthermore, it is possible to notice the onset of anomie, impaired ability to construct sentences, read and write as well as loss of fluency of speech and syntactic reduction in the messages formulated by the subject.

Conclusions

The article presents a speech therapy case study of a patient with progressive supranuclear palsy. As for interactional competence and

²² Conf. D.I. Dominguez, B. De Strooper, *Novel therapeutic strategies provide the real test for the amyloid hypothesis of Alzheimer's disease*, „Trends in Pharmacological Sciences” 2002, no. 23, p. 324.

skills, the subject has a preserved ability to understand non-verbal kinetic, prosodic and proxemic behaviour. Deficits in understanding verbal and non-verbal messages depend on the complexity of the messages. Creating both non-verbal and verbal behaviour is impaired. In terms of communicative competence and skills, deficits are observed in understanding and applying linguistic social, situational and pragmatic rules. The examination of language competence and skills allows the author to conclude that there are fewer deficits in the recognition of units of the phonological subsystem, prosodic features and the understanding of the morphological subsystem units than in the understanding and building syntactic structures, understanding and conducting metalinguistic operations or the realisation of phonological subsystem units, prosodic features, the ability to fluently update units of the morphological system. In addition, pathological changes result in impaired ability to perform other language tasks, such as writing or reading.

Attention is drawn to the uncharacteristic slow course of the disease in the examined patient in the light of data in the literature.²³ The research paper confirms the thesis that dementia disorders overlap with motor disorders in the course of PSP.

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²³ P. Liberski, T. Sobów, B. Sikorska, *Postępujące...* p. 539; M. Pąchalska, H. Kurzbauer, B. Grochmal-Bach, B.D. MacQueen, A. Urbanik, I. Herman-Suchar-ska, *Nietypowe zaburzenia języka i mowy u pacjentki z klinicznym rozpoznaniem zespołu Steele'a-Richardsona-Olszewskiego*, [in:] *Choroby otępienne. Teoria i praktyka*, ed. by J. Leszek, Continuo, Wrocław 2011, p. 271.

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