Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.elsevier.com/locate/poamed

Case Report

Logopenic variant of primary progressive aphasia – Case report

Agnieszka Rakowska *, *Tomasz Matyskieła*, *Dorota Mroczkowska*, *Beata Zwiernik*, *Jacek Zwiernik*

Department of Neurology and Neurosurgery, Faculty of Medical Sciences, University of Warmia and Mazury, Poland

ARTICLE INFO

Article history:
Received 28 November 2014
Received in revised form
9 May 2015
Accepted 28 October 2015
Available online 19 December 2015

Keywords:
Primary progressive aphasia
Logopenic aphasia
Alzheimer's disease

ABSTRACT

Introduction: Speech disorders are often first symptoms of dementias with neurodegenerative basis. Differences in the clinical picture and different types of the speech difficulties may make diagnosis of this degenerative process easier.

Aim: To present an example of clinical evaluation of the patient with primary progressive aphasia (PPA) according to the newest diagnostic criteria. To shortly revise current knowledge about logopenic variant and its association with Alzheimer disease.

Case study: We present a case of a 75-year-old man suffering from progressive language difficulties, who was finally diagnosed as having primary progressive aphasia – logopenic variant. Clinical data, neuroimaging, psychological test batteries and speech therapist's examination based on the Boston Aphasia Test were used.

Results and discussion: In the first part we revise evolution of primary progressive aphasia diagnostic criteria and nomenclature, and focus on current approach to the patient with isolated, progressive speech difficulties. In the second part we attempt to summarize linguistic, neuropsychological and pathological findings that one may encounter in the case of logopenic variant of PPA.

Conclusions: Diagnosis of primary progressive aphasia requires a close cooperation between neurologist, speech therapists and psychologists. Clinical presentation, due to various level of cognitive decline at first stages of the disease and individualization of the clinical picture, is nonuniform. Recently created diagnostic criteria make both basic diagnosis and diagnosis of the primary progressive aphasia variants easier. This may lead choosing the rehabilitation methods easier in case of disordered language functions and other cognitive domains.

* *Correspondence to*: Department of Neurology and Neurosurgery, Faculty of Medical Sciences, University of Warmia and Mazury, Warszawska 30, Olsztyn 10-082, Poland. Tel.: +48 661950089.
E-mail address: aune.inez@wp.pl (A. Rakowska).

http://dx.doi.org/10.1016/j.poamed.2015.10.001

1. Introduction

Speech disorders are often first symptoms of dementias with neurodegenerative basis.

Differences in the clinical picture and different types of the speech difficulties may make diagnosis of this degenerative process easier.

In the article we present a case description of a patient suffering from speech difficulties characteristic for logopenic variant of the primary progressive aphasia (PPA). Clinical data, neuroimaging, psychological test batteries and speech therapist's examination based on the Boston Aphasia Test were used. This test is mainly utilized in the examination of aphasia after the ischemic stroke, but it may be used, as it contains elements required in the comprehensive assessment of the language deficiencies.

2. Aim

To present example of clinical evaluation of the patient with PPA according to the newest diagnostic criteria. To shortly revise current knowledge about logopenic variant and its association with Alzheimer disease.

3. Case report

The patient, 75-year-old male, retired warehouse-keeper (educational background: vocational school), with history of chronic obstructive pulmonary disease, arterial hypertension and slight prostate hyperplasia, was admitted to the Neurology Clinic in order to diagnose his language disorders, which has been persisting for about six years, connected with difficulties in the pronunciation of the words. Patient's wife describes them as: stumbling/faltering, losing appropriate words and searching for them. Difficulties connected with the beginning of the pronouncement. At the same time understanding of the language was relatively well kept. Firstly, language disorders were small and they did not affect daily activities. After, two years, the symptoms got stronger and became significant enough to limit patient's verbal contact with the others. Moreover, reading and writing difficulties occurred.

During the neurological examination, no sensory deficiencies, muscular strength deficiencies or pyramidal symptoms were revealed. There were no features of ataxia.

Head MRI (Fig. 1) revealed: widened lateral ventricle on the left, Sylvius sulci, widened sulci and narrowed gyri in the lower part of the parietal lobe and upper portion of the temporal lobe and also disseminated small vascular lesions.

Speech therapist's examination was conducted based on the Boston Aphasia Test, examining spontaneous speech, understanding of the language, oral expression, reading and writing. It revealed: word fluency disorders, difficulties in finding appropriate words both during the spontaneous speech and calling; handicapped repeating of sentences and words. In the spontaneous language and during calling there occurred many phonologic paraphasias. Deficits in executive functions such as writing are concerned, elements of dyscalculia and dyslexia occur.

Psychiatric examination, which was conducted four years after occurrence of first symptoms revealed some undefined organic personality and behavior disorders caused by the dysfunction of the brain. The patient did not display any positive symptoms, but during the examination he revealed increased level of anxiety; he was clearly excited, and actively aggressive. Psychological research conducted using Mini Mental State Examination (MMSE; 27 points) did not reveal any symptoms of dementia. In the AVLT tests, decreased quickness of memorizing and the ability of recalling based on the reconstructing and recognizing language material were assessed. In the BVRT test, the presence of the slight visual–spatial functions' disorders was noticed. Lower efficiency of the executive functions was assessed using CTT-1 and CTT-2 tests (<10 centiles). During the next examination, conducted two years later, speech disorders were still predominant. Psychological research showed that recent memory disorders

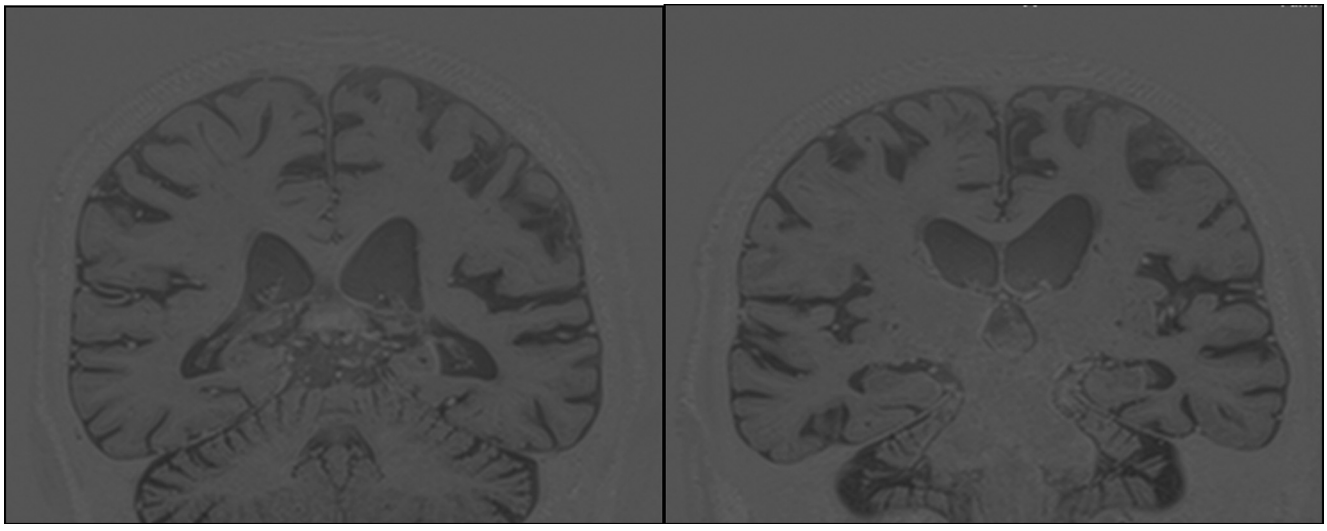

Fig. 1 – The MRI revealed entities: widened lateral ventricle on the left, Sylvius sulci, widened sulci and narrowed gyri in the lower part of the parietal lobe and upper portion of the temporal lobe and also disseminated small vascular lesions.

were stronger and also that ability of memorizing new information was decreased. Episodic memory was preserved, although the patient had problems with recalling dates. Also executive functions deficiencies occurred to be bigger, including intentional material screening, initiating of the activity, shifting of the attention and postponed reactions. Visual-constructive functions disorders were assessed as mild. In the emotional–motivational sphere high sensitivity, lack of action and negative attitude to the examination have been observed.

4. Results and discussion

4.1. *PPA – classification*

According to primal classification, PPA is a rare affection, which predominant symptoms are slowly progressive speech disorders. PPA is a clinical state connected with progressing losing of particular speech functions and with an initial abatement of the other cognitive domains.[1]

First description of the patient with an isolated losing of words comprehension belongs to Paul Serieux.[2] After the patient's death, during an autopsy test, Dejerine[3] found bitemporal cerebral cortex atrophy and neuronal loss. Another assessment of the biopsy material was conducted by Mesulam,[3] who also excluded an Alzheimer process. Based on previous observations of the cases of his own patients, he proposed name: ''slowly progressing aphasia without a general dementia''[4] and next ''primary progressive aphasia''.[5]

Most patients with PPA start to have first PPA symptoms before the age of 65.[6] Memory, visual transformation and personality during the first stages are relatively well kept. It is critical as far as differentiating between logopenic and semantic variants of primary progressive aphasia and Alzheimer disease and frontotemporal dementia is concerned. It was agreed that period when speech disorders are predominant in case of the patients with PPA should last about two years.[5,7,8] Neurodegenerative etiology of the disorders should be supported by neuroimaging, which allow to exclude focal lesions connected to the clinical picture.

PPA currently includes heterogeneous group with various clinical, pathological and radiological presentation. Terminology and criteria in this respect are constantly evolving. Previously commonly used in the world literature division mentioning primary progressive non-fluent aphasia and semantic dementia is no longer in use. The term ''primary progressive aphasia'' should be a root diagnosis,[8] which means that one should diagnose PPA at first and then indicate its actual subtype. In accordance with Gorno-Tempini and coauthors' diagnostic criteria from 2011,[9] three variants of PPA can be distinguished: non-fluent variant (nfvPPA), logopenic PPA and semantic dementia (SD). According to later criteria, proposed by Mesulam,[10] one can, however, assign patients with PPA to one of five subtypes: non-grammatical (PPA-G), semantic (PPA-S), logopenic (PPA-L), anomic (PPA-A) and mixed (PPA-M). Both classification systems require the basic diagnostic criteria of PPA to be met.

Diagnostic criteria of PPA according to Mesulam include (see also Table 1):

- presence of speech disorders with slow beginning and gradual progressive course, mainly as difficulties in calling things and using last names;
- all of the previous activities are performed properly in the period of at least two years since the occurrence of the symptoms;
- before the occurrence of the disease symptoms, speech functions are fully preserved;
- within first two years of the disease, apathy, frontal lobe syndrome, visual–spatial orientation disorders or symptoms of sensory disorders are not observed;
- within first two years of the disease acalculia and apraxia may occur, but they do not affect daily activities;
- other cognitive functions may gradually get worse after two years of the disease, however aphasic disorders are predominant.

4.2. *Logopenic variant of PPA*

In 2004 Gorno-Tempini and cooperatives chose a group of people among the patients who represented disorders typical neither for Alzheimer disease nor for the semantic dementia. Due to slow speech with numerous pauses for finding appropriate words, the same as Mesulam in his reports, they called this group ''logopenic''.[11]

Table 1 – Current types of PPA.[20]

Subtype of PPA	
Agrammatic subtype (PPA-G)	Impaired grammatical structure of spoken or written language in the absence of significant word comprehension impairments. Output is usually of low fluency but does not have to be dysarthric or apraxic.
Semantic subtype (PPA-S)	Impaired word comprehension in the absence of significant impairment of grammar. Object naming is severely impaired. Output is motorically fluent but contains word finding hesitations, paraphasias and circumlocutions.
Logopenic subtype (PPA-L)	No significant grammar or word comprehension impairment. Speech contains many word-finding hesitations and phonemic paraphasias. Object naming may be impaired and may constitute the only significant finding in the neuropsychological examination. Repetition impairments are required for diagnosis.
Anomic subtype (PPA-A)	All features as in PPA-L except that repetition are intact.
Mixed subtype (PPA-M)	Impaired grammatical structure and word comprehension, even at the early stages of disease.

Logopenic variant of PPA, contrary to non-grammatical PPA, is distinguished by the lack of agrammatism and by kept prosody.[10] Speech, although it is not fluent, is not telegraphic. Patients find it difficult to update the words both during the spontaneous speech, and calling things. Contrary to SD, understanding of single words is relatively well kept. Phonological paraphasias in spontaneous speech and calling, without changing of the speech sounds are peculiar.[9,10,12] Understanding and repeating in the logopenic variant of PPA, contrary to non-grammatical type of PPA, are limited in case of long sentences and phrases and do not depend on their grammatical complexity. That is why the short-lasting phonological memory defect is a postulated speech disorder mechanism.[13]

Disturbances coexisting with speech disorders include, among others, dyscalculia, phonological dyslexia, limb apraxia and changes of behavior such as: anxiety, sensitivity, excitement or apathy.[14,15] It was also revealed that there is a connection between the early disorders of episodic memory in the course of PPA and Alzheimer disease pathology.[16]

PPA-L is connected with peculiar lesions in the neuroimaging tests. In case of MRI and SPECT, atrophy or lower flow was revealed in the back side of the upper and middle left temporal gyrus and in the lower parietal lobe.[13] Similar pattern of atrophy is observed in the Alzheimer disease, especially with the early beginning,[17] although in PPA-L lesions are located mainly in the left temporal area.[18] Connection between PPA-L and Alzheimer disease seems to be confirmed by the accumulation of Beta amyloid in the typical for PPA-L cerebral cortex areas[19] and decrease of the concentration of AB42 and accumulation of tau protein in the cerebrospinal fluid.[15]

One should remember that the main criterion, which allows to differentiate between PPA-L and Alzheimer disease is, in case of predominant speech disorders and cognitive deficiencies, at least two years period of speech disorders dominance.

5. Conclusions

Current state of knowledge about PPA is based on the descriptions of single cases. Diagnosis requires a close cooperation among neurologist, speech therapists and psychologists. Clinical presentation, due to various level of cognitive decline at first stages of the disease and individualization of the clinical picture, is nonuniform. Recently created PPA diagnostic criteria make both basic diagnosis and diagnosis of its variants easier. This may lead choosing of the rehabilitation methods easier in the case of disordered language functions and other cognitive domains.

Conflict of interest

None declared.

REFERENCES

1. Harciarek M, Kertesz A. Primary progressive aphasias and their contribution to the contemporary knowledge about the brain–language relationship. *Neuropsychol Rev*. 2011;21(3):271–287.
2. Sérieux P. Sur un cas de surdité verbale pure. *Rev Med*. 1893;13:733–750.
3. Dejerine J, Sérieux P. Un cas de surdité verbale pure terminée par aphasie sensorielle, suivi d'autopsie. *CR Seances Soc Bio Paris*. 1897;49:1074–1077 [in French].
4. Mesulam MM. Slowly progressive aphasia without generalized dementia. *Ann Neurol*. 1982;11(6):592–598.
5. Mesulam MM. Primary progressive aphasia – differentiation from Alzheimer's disease. *Ann Neurol*. 1987;22(4):533–534.
6. Mesulam MM, Wieneke C, Thompson C, Rogalski E, Weintraub S. Quantitative classification of primary progressive aphasia at early and mild impairment stages. *Brain*. 2012;135(Pt 5):1537–1553.
7. Mesulam MM. Primary progressive aphasia. *Ann Neurol*. 2001;49(4):425–432.
8. Mesulam MM. Primary progressive aphasia: a 25-year retrospective. *Alzheimer Dis Assoc Disord*. 2007;21(4):S8–S11.
9. Gorno-Tempini ML, Hillis AE, Weintraub S, et al. Classification of primary progressive aphasia and its variants. *Neurology*. 2011;76(11):1006–1014.
10. Mesulam MM. Primary progressive aphasia: a dementia of the language network. *Dement Neuropsychol*. 2013;7(1):2–9.
11. Gorno-Tempini ML, Dronkers NF, Rankin KP, et al. Cognition and anatomy in three variants of primary progressive aphasia. *Ann Neurol*. 2004;55(3):335–346.
12. Wilson SM, Henry ML, Besbris M, et al. Connected speech production in three variants of primary progressive aphasia. *Brain*. 2010;133(Pt 7):2069–2088.
13. Gorno-Tempini ML, Brambati SM, Ginex V, et al. The logopenic/phonological variant of primary progressive aphasia. *Neurology*. 2008;71(16):1227–1234.
14. Rohrer JD, Ridgway GR, Crutch SJ, et al. Progressive logopenic/phonological aphasia: erosion of the language network. *Neuroimage*. 2010;49(1):984–993.
15. Rohrer JD, Warren JD. Phenomenology and anatomy of abnormal behaviours in primary progressive aphasia. *J Neurol Sci*. 2010;293(1/2):35–38.
16. Munoz DG, Woulfe J, Kertesz A. Argyrophilic thorny astrocyte clusters in association with Alzheimer's disease pathology in possible primary progressive aphasia. *Acta Neuropathol*. 2007;114(4):347–357.
17. Frisoni GB, Pievani M, Testa C, et al. The topography of grey matter involvement in early and late onset Alzheimer's disease. *Brain*. 2007;130:720–730.
18. Migliaccio R, Agosta F, Rascovsky K, et al. Clinical syndromes associated with posterior atrophy: early age at onset AD spectrum. *Neurology*. 2009;73(19):1571–1578.
19. Rabinovici GD, Jagusi WJ, Furst AJ, et al. Abeta amyloid and glucose metabolism in three variants of primary progressive aphasia. *Ann Neurol*. 2008;64(4):388–401.
20. Mesulam MM, Weintraub S. Spectrum of primary progressive aphasia. *Baillieres Clin Neurol*. 1992;1(3):583–609.