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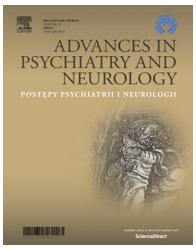

Original research article/Artykuł oryginalny

Overlapping and distinguishing features of descriptive speech in Richardson variant of progressive supranuclear palsy and non-fluent progressive aphasia

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ARTICLE INFO

Article history:
Received: 20.04.2015
Accepted: 12.06.2015
Available online: 21.06.2015

Keywords:
- Progressive supranuclear palsy
- Primary progressive aphasia
- Alzheimer's disease
- Dementia
- Speech analysis

Słowa kluczowe:
- postępujące porażenie ponadjądrowe

ABSTRACT

Background: Progressive supranuclear palsy (PSP) has overlapping clinical features with primary progressive aphasia (PPA). Our study aimed at comparing classical PSP phenotype – Richardson syndrome (PSP) with three different variants of PPA in terms of descriptive speech. **Methods:** Fifty-eight patients participated in the study: 18 with the clinical diagnosis of PSP-RS, 14 with nfvPPA, eight with logopenic variant of PPA (lvPPA), 8 with semantic variant of PPA (svPPA) and 10 with Alzheimer's disease (AD). **Results:** Lexical access in descriptive speech is comparably good in PSP-RS and nfvPPA. However, speech rate is faster in PSP-RS than in nfvPPA and patients with PSP-RS are more likely to construct complex sentences than individuals with nfvPPA. **Conclusion:** Overlapping linguistic features were noted between PSP-RS and nfvPPA, but not with lvPPA or svPPA.

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http://dx.doi.org/10.1016/j.pin.2015.06.003

- afazja pierwotna postępująca
- choroba Alzheimera
- otępienie
- analiza mowy

Introduction

Progressive supranuclear palsy (PSP) was originally described as a prototypic form of subcortical dementia [1]. However, the overlap between PSP and frontotemporal lobar degeneration (FTLD) has been increasingly recognized [2] and PSP is currently regarded as a cluster of clinical syndromes, rather than a unitary entity. The originally reported PSP phenotype is referred to as Richardson's syndrome of PSP (PSP-RS). It shares neuropsychological, behavioural and neuroradiological features with behavioural variant of frontotemporal dementia (bvFTD). Another PSP phenotype, sharing clinical features with FTLD spectrum is PSP variant with progressive apraxia of speech evolving into progressive non-fluent aphasia (PSP-PNFA) [3].

Primary progressive aphasia (PPA) is currently divided into three variants: non-fluent (nfvPPA; PNFA), semantic (svPPA) and logopenic (lvPPA). Non-fluent and semantic variants are usually associated with FTLD, while logopenic variant usually has underlying Alzheimer's disease (AD) pathology. In nfvPPA parkinsonism and features of corticobasal syndrome may appear in the disease course [4].

This paper addresses the potential overlap of linguistic features between PSP-RS and nfvPPA. Descriptive speech of patients with PSP-RS is compared against the speech of patients with nfvPPA, svPPA, lvPPA and AD. It is hypothesized that speech in PSP-RS shares more features with nfvPPA than with other PPA variants and AD.

Patients and methods

Participants

Fifty-eight patients participated in the study: eighteen (13 men, 5 women) with the clinical diagnosis of PSP-RS according to Litvan et al. criteria [5] (13 probable, 5 possible), fourteen (3 men, 11 women) with nfvPPA, eight (3 men, 5 women) with lvPPA, eight (4 men, 4 women) with svPPA (diagnosed at level I according to Gorno-Tempini et al. criteria [4]) and ten (4 men, 6 women) with AD (diagnosed according to McKhann et al. [5]). All patients were diagnosed in centres specializing in the diagnosis of neurodegenerative disorders.

The patients' age averaged 67 ± 10 years in PSP-RS, 67 ± 10 years in nfvPPA, 69 ± 4 years in lvPPA, 64 ± 7 years in svPPA and 74 ± 9 years in AD. The time since symptom onset ranged from 1 to 5 years in PSP-RS, from 0.5 to 5 years in nfvPPA, from 1 to 9 years in lvPPA, from 1 to 7 years in svPPA and from 3 to 9 years in patients with AD. The groups were matched in terms of age ($p = 0.170$) and years of education ($p = 0.143$). All participants volunteered for this study and provided informed consent to participate. The study procedures were approved by local bioethics committee.

Methods

To assess the patients' descriptive speech the description of Cookie theft picture from Boston Diagnostic Aphasia Examination-3 or A beach scene by Prof. EK Warrington [6] was administered by a neuropsychologist (EJS, AB or DW). All picture descriptions were transcribed and scored by two independent raters specializing in speech pathology (KKK and MK). The raters were aware of the spectrum of disorders being analysed, but they were blinded to the clinical diagnosis in each patient. Divergent scores were discussed with the third rater (EJS) and scores reported were reached by consensus. For all patients with PSP-RS, AD, lvPPA and 8 patients with nfvPPA original recordings were available at the time of the analysis. For the remaining six patients with nfvPPA and patients with svPPA, there were only written transcripts available at the time of the result analysis. Thus, lexical and syntactic aspects were compared using the complete data set and for analysis of motor aspects of speech only patients' data with available recordings were included in the analysis. For each parameter assessed raw scores (number of occurrence) were used in the analysis: sample duration, number of words, lexical content (number of nouns, verbs, definite and indefinite pronouns), occurrences of palilalia, phonemic paraphasias, non-fluencies (at word and sentence level separately). Raters were also asked to detect features suggestive of aprosody and dysarthria. Subsequently, so as to make the results independent of the variable duration of speech samples, several variables were computed as ratio or proportion of raw scores (e.g. proportion of nouns to total number of words used).

Results

Lexical and syntactic aspects of descriptive speech

On the lexical level, speech samples produced by patients with PS-RS contained similar percentage of nouns to samples from nfvPPA group. Moreover PSP-RS patients used significantly more nouns than patients with lvPPA and svPPA (Tab. I). A similar pattern of results was observed for the content words ratio (proportion of nouns and verbs to total number of words), having been higher in both PSP-RS and nfvPPA groups.

The use of in(definite) pronouns in PSP-RS group was much less common than in patients with lvPPA and svPPA.