Fluent primary progressive aphasia: a pure language disorder distinct from semantic dementia?

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SUMMARY

Fluent primary progressive aphasia (fPPA) is very often confused with semantic dementia (SD), due to the ambiguity of the latter term. We present a case of fPPA with neuropsychological and neuroimaging data, not fulfilling the diagnostic criteria for SD. The language disturbance profile is presented, resembling transcortical sensory aphasia. The results of both standardized and experimental testing show speech comprehension disturbance with preserved fluency of speech as a core deficit. The specific character of fPPA is revealed by phonological and syntactic deficits, a modality effect in both oral and written naming, and preserved single-word comprehension in contrast to disturbed comprehension of complex sentences.

INTRODUCTION

The diagnostic criteria for primary progressive aphasia (PPA) were provided by Mesulam (2001). Any type of aphasia can appear in a progressive variant (Serrano et al., 2005; see: McNeil and Duffy, 2001 for detailed description), but the non-fluent forms have been most frequently reported. The relative scarcity of data on fluent primary progressive aphasia (fPPA) may be due to the existence of the term "semantic dementia" (SD), which is often
inconsistently applied, both in the literature and in clinical practice. On the one hand, SD refers to a frontotemporal dementia subtype with a dominant semantic deficit, accompanied by visual associative agnosia and prosopagnosia. On the other hand, it denotes the fluent subtype of PPA, with impaired speech comprehension as a core deficit (Mesulam, 2003). Both fPPA and SD (in its first meaning) are characterized by preserved fluency of speech and anomia (Sonty et al., 2003). The fluent PPA variant has been shown to include mild semantic deficits. It can be interpreted either as possible conversion to SD in later stages of the disease (Adlam et al., 2006; Knibb and Hodges, 2005), or as the generalization of cognitive symptoms to full-blown dementia. In the literature, the profile of language disturbance in fPPA has not been thoroughly described, in comparison to non-fluent PPA. Better verb than noun naming has been observed in fPPA (Hillis et al., 2004).

Neuroimaging studies of PPA, when abnormal, show isolated or predominant left hemisphere abnormalities, mainly in the temporal and/or frontal lobe. Non-fluent PPA is associated with evidence of atrophy of the inferior frontal gyrus, premotor cortex and anterior insula (Gorno-Tempini et al., 2004), whereas fPPA presents with changes in the left peri-Sylvian tempo-parietal region and adjacent superior temporal gyrus (Adlam et al., 2006). Neuroimaging studies in SD show temporal lobe atrophy, which is most often bilateral with left or right predominance. The anterior and infero-lateral parts of the temporal lobes are predominantly affected (Garrard and Hodges, 2000).

In this paper, we present a case of fPPA in which the accepted diagnostic criteria for SD are not met, so as to show some specific features that distinguish between fPPA and SD, such as impaired speech comprehension at the single-word or more complex level. Neuropsychological data describing aphasic symptoms and other aspects of cognitive function are combined with neuroimaging.

**Case description**

A 76-year-old, right-handed woman with around 13 years of formal education, who had previously worked as a teacher of needlecraft, noticed naming problems beginning about two years before our investigation. However, her family reports a 5-year history of progressive anomia. As a widow living alone, she initially experienced difficulty mainly in communication by telephone with her family and in handling formal conversations in different institutions. At present, the communication burden lies mainly on the interlocutor. The patient exhibits severe anomia with preserved fluency in spontaneous speech. Word-finding difficulties in daily life are effectively compensated by pantomime and circumlocutions. The patient is fully aware of word-finding deficits, but demonstrates poor awareness of her language comprehension problems. Likewise, her family failed to notice the speech comprehension problems.

The patient is a polite and socially well adapted woman. Her medical history is unremarkable. One of her sons was diagnosed with Parkinson's dis-
ease at the age of 50. One year before fPPA was diagnosed, she had been misdiagnosed with Alzheimer’s disease. At that time she started therapy with rivastigmine (3mg per day), but there was no improvement in language function. At the time of our assessment, she was well oriented in her familiar context and had no problems in daily living activities. Her day-to-day memory was well preserved.

**Neurological examination**

The patient displays mild bilateral motor apraxia with some postural tremor. Verbal communication is severely impaired. She speaks fluently and effortlessly without agrammatism, but her spontaneous speech is empty of content. The patient cannot compensate for her speech comprehension problems. When confronted with a message that is not understandable, she exhibits a strong tendency to echolalia and echopraxis. Apart from that, neurological examination shows no abnormalities.

**Neuroimaging data**

Neuroradiological imaging showed a slight bilateral cortical and subcortical atrophy of the temporal and frontal lobes in CT scans, and significant atrophy of the left temporal lobe with the left lateral ventricle enlargement in MRI (see: Figure 1). MRI results rule out vascular or other non-neurodegenerative etiology.

An initial rCBF SPECT study with 99mTc- ECD, performed 10 months prior to diagnosis, showed substantially reduced perfusion mainly in the left inferior and middle temporal gyri, and in the left superior and inferior parietal gyri, along with subtle changes in the superior and middle frontal gyri bilaterally-

![Fig. 1. MRI of the brain, September 2007: (A) Coronal T2-weighted view – marked atrophy of left temporal and frontal lobes. (B) Axial FLAIR view – left perisylvian temporal – parietal atrophy](image)
more prominent on the left. SPECT re-examination (performed at the time of fPPA diagnosis) demonstrated progression in diminished radiotracer uptake in the left hemisphere, extending to the anterior part of the temporal lobe, the posterior part of the frontal lobe, and the superior part of the parietal lobe (see: Figure 2).

Electroencephalography was unremarkable.

Neuropsychological testing

The neuropsychological diagnosis aimed at describing the profile of language disturbances and other cognitive dysfunctions in the case presented so as to exclude cognitive symptoms characteristic for dementia syndromes other than fPPA. Language skills were assessed by means of tasks derived from the Boston Diagnostic Aphasia Examination (Goodglass and Kaplan, 1983), Boston Naming Test-short version (Kaplan et al., 1983), Frenchay Aphasia Screening Test (FAST) (Enderby et al., 1987) and Token Test part V (DeRenzi and Vignolo, 1962). Among the tasks used to assess other cognitive functions were elements of the Mini-Mental State Examination (Folstein et al., 1975), the Short Test of Mental Status (Kokmen et al., 1987), the Addenbrooke Cognitive Examination (Maturanath et al., 2000), A Visual Learning Test (Lamberti and Weidlich, 1998), the Block Design subtest from the WAIS-R (Wechsler, 1981), the Clock Face Filling task (Goodglass and Kaplan, 1983), the Rey Complex Figure Test (Rey, 1964), and calculia and praxis trials. The experimental investigation was designed to show that fPPA in this case is not equivalent to SD.
**Language processing**

**Spontaneous speech.** When talking to an examiner or caregiver she presented severe word finding difficulties. However, when she was shown the cookie theft picture from the BDAE, the problems were less obvious. The subject tended to use verbal stereotypes, such as "Mum and Dad", as verbal prostheses, which was not due to visual agnosia, as later on she correctly named a boy. Syntax errors were rare.

**Naming.** Confrontation naming was tested by two measures (see: Table 1). It is severely impaired without category specific deficits (see: Tables 2 and 3). The frequency of different naming errors was counted in all naming trials (both standardized and experimental). Among 57 aphasic errors (observed in all naming tasks), there were no spontaneous phonemic paraphasias (they appeared when the patient was provided with a phonemic cue), only 2 verbal paraphasias, 28% semantic paraphasias (16) and 70% circumlocutions (40). The effective pantomime usage and circumlocutions showed that naming deficit could not be attributed to poor visual recognition, and that the subject retained knowledge about the objects' usage (e.g. "you can sit on it" when showed a picture presenting a bench, pantomime of peeling a banana when asked to name "a banana", or of ironing when asked to name the action of "ironing"). Word-finding difficulties concerned mainly low and average frequency words. Responsive naming (naming in answer to orally presented definitions), which provoked echolalia, was worse than confrontation naming. Neither phonemic nor semantic cues were effective.

**Comprehension.** Single word comprehension was quite well preserved when compared with naming (see: Table 2). In the case of complex messages, the patient could understand 2-step commands when comprehension did not require understanding the relations conveyed (Commands from BDAE). However, comprehension of complex sentences was severely impaired (Token Test part V). The subject could understand commands such as "Touch the blue circle and the red square," but not "Before touching the yellow circle, pick up the red square". The level of complexity was a better predictor of the patient's performance than the length of the sentence.

**Repetition.** Repetition of single phonemes, syllables, syllable pairs and words containing up to 5 syllables, when the structure of the word was simple and familiar to the patient, was unimpaired. Sentence repetition was impaired with a nonsignificant difference for high and low frequency sentences (see: Table 1). The repetition of sentences showed phonemic paraphasias and neologisms, which were absent in naming trials (apart from responding to a phonemic cue) and spontaneous speech. Such errors demonstrate disturbed phonological processing.

**Automatized sequences.** With slight help to initiate the subject could produce the days of the week and count (from 1 to 23, with one error).

**Reading.** The patient could read high frequency words; sentence reading was slightly impaired. The subject could understand simple written commands.
Table 1. Language processing data for a patient with PPA

<table>
<thead>
<tr>
<th>Function</th>
<th>Measure</th>
<th>score range</th>
<th>patient’s score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Naming</td>
<td>Boston Naming Test- short version</td>
<td>0-15</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>BDAE – Confrontation naming</td>
<td>0-105</td>
<td>81</td>
</tr>
<tr>
<td></td>
<td>BDAE – Responsive naming</td>
<td>0-30</td>
<td>8</td>
</tr>
<tr>
<td>Repetition</td>
<td>BDAE – Sentence repetition- high frequency</td>
<td>0-8</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>BDAE – Sentence repetition- low frequency</td>
<td>0-8</td>
<td>0</td>
</tr>
<tr>
<td>Comprehension</td>
<td>BDAE – Word discrimination</td>
<td>0-72</td>
<td>70</td>
</tr>
<tr>
<td></td>
<td>BDAE – Commands</td>
<td>0-15</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>BDAE – Complex ideational material (1-4)</td>
<td>0-4</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Token test – part V</td>
<td>0-22</td>
<td>5</td>
</tr>
<tr>
<td>Reading</td>
<td>BDAE – Word reading</td>
<td>0-30</td>
<td>29</td>
</tr>
<tr>
<td></td>
<td>BDAE – Sentence reading</td>
<td>0-10</td>
<td>8</td>
</tr>
<tr>
<td></td>
<td>FAST – Reading comprehension</td>
<td>0-5</td>
<td>3</td>
</tr>
<tr>
<td>Writing</td>
<td>BDAE – Writing words to dictation</td>
<td>0-10</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>BDAE – Written confrontation naming</td>
<td>0-10</td>
<td>7</td>
</tr>
</tbody>
</table>

Table 2. Confrontation naming and Word discrimination from the BDAE for a patient with PPA

<table>
<thead>
<tr>
<th>Category</th>
<th>Confrontation naming patient’s score (max. score)</th>
<th>Word discrimination patient’s score (max. score)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Objects</td>
<td>9 (18)</td>
<td>10.5 (12)</td>
</tr>
<tr>
<td>Letters</td>
<td>18 (18)</td>
<td>12 (12)</td>
</tr>
<tr>
<td>Geometric forms</td>
<td>6 (6)</td>
<td>9.5 (12)</td>
</tr>
<tr>
<td>Actions</td>
<td>11 (18)</td>
<td>6 (6)</td>
</tr>
<tr>
<td>Numbers</td>
<td>15 (18)</td>
<td>10.5 (12)</td>
</tr>
<tr>
<td>Colors</td>
<td>13 (18)</td>
<td>10.5 (12)</td>
</tr>
<tr>
<td>Body parts</td>
<td>9 (18)</td>
<td>-</td>
</tr>
<tr>
<td>Global Score</td>
<td>81 (105)</td>
<td>70 (72)</td>
</tr>
</tbody>
</table>
but not complex ones, when comprehension of relations was required (see: Table 1).

Writing. Written confrontation naming was better than writing to dictation. Omission errors occurred when copying a sentence.

Summary Language comprehension impairment was accompanied by a moderately severe naming deficit. Repetition and automatized language sequences were relatively preserved. Understanding single words was better than understanding complex sentences. Reading and writing short and simple words was partially preserved. Spontaneous speech was fluent but not informative due to word finding difficulties. The degree of language impairment according to BDAE was 2. The pattern of language disturbance resembles transcortical sensory aphasia.

COGNITIVE FUNCTION

No signs of psychomotor slowing were noticed. In the Digit Symbol subtest from the WAIS-R, the slightly lowered scale score (7) was due to poor reading of digits. The subject was well oriented in time, which was evidenced e.g. by her choosing a photo of the current president and pope among many other photos presented. Her performance in such tasks showed no features of prosopagnosia.

Visuospatial functions were preserved: the subject could copy the design from the MMSE and a cube (from the Short Mental Status Examination). She performed exceptionally well in the Rey Complex Figure Test (32 points- first trial, 32 points- second trial- 50th percentile; two copy trials were given, as the subject was dissatisfied with her first drawing). The subject could spontaneously indicate subtle flaws in her drawing. She performed at an average level in the Block Design subtest (scale score 9). She performed well in identifying masked letters (4/4) and counting dots from Addenbrooke's Cognitive Examination (4/4), and also in Poppelreuter's multiple choice format task (4/4).

Visuospatial memory was tested by means of the Benton Visual Retention Test with multiple choice format, and the visuospatial learning task (Lamberti and Weidich, 1998). In the first test, the subject's score was 6 (max. score 10), but she changed 4 out of 10 correct answers into incorrect ones, which reflected a selection problem, rather than short-term visuospatial memory impairment. In the visuospatial learning task the subject was presented with 9 figures, and after each of 6 presentation trials she was instructed to arrange them using 5 wooden sticks. The learning curve is very low (0,2,1,1,1,2, recognition 7, with 2 false recognitions; after a 10-minute delay, 2, delayed recognition 6, with 4 false recognitions), but no forgetting was observed after a filled delay. Planning and cognitive control deficits could have influenced her poor performance on this task.

The subject could name and write numbers up to 2 digits. Adding and subtraction was possible on numbers up to 2 digits. Other operations were
impossible to execute. The acalculia seemed to be related to the language disturbances.

Limb praxis was preserved, as shown by experimental trials. A variety of naming trials showed intact ideomotor praxis, since the patient used adequate gestures to demonstrate the use of a variety of tools, such as a hammer or a saw. Efferent motor praxis (in Luria's hand-position sequencing task) was severely impaired (first sequence, 1/5; second sequence, 0/5).

Executive function was assessed mainly qualitatively. In the Complex Figure Test the subject demonstrated poor planning (additive details approach). In a less complex task, such as clock face filling (putting all the numbers on the clock face), no planning deficits were observed. The subject committed only one single perseverative mistake in copying Luria's alternate design. Executive dysfunction seems to be mild in the context of language problems.

**Summary.** Language disturbance was accompanied by mild visuoconstructive deficits and executive dysfunction. Acalculia symptoms were secondary to language disturbance. Aphasic symptoms constituted the core deficit.

**Experimental investigation**

Two experiments were designed to analyze the patient's deficits. The first trial was intended to demonstrate the specificity of sentence repetition problems, and to explain the appearance of phonemic paraphasias in repetition and when provoked by phonemic cues. The subject was asked to repeat high frequency sentences containing 3-10 syllables (2 sentences for each length were provided; the task was discontinued after two failures for a given sentence length, as in the Digit Span subtest from the WAIS-R). As expected, phonemic paraphasias appeared when the sentence length exceeded the immediate memory span, which was 8 syllables long.

The second experiment, inspired by the work of Adlam et al. (2006), was devised to test semantic knowledge and language processing. Forty-eight gray scale pictures presenting items from 6 categories were prepared (see: Appendix). There were 3 categories of organic objects (fruit, vegetables, insects) and 3 non-organic ones (vehicles, tools, furniture). Each category contained 8 items. There were 6 tasks based on the same picture set. First, semantic fluency trials were administered (60 seconds for each category). Secondly, the subject had to match the appropriate color to a grayscale picture of a given object (10 trials). Then there was a naming trial for all pictures presented in random order (as indicated in the Appendix). Subsequently, the participant had to classify the pictures into 6 categories, where examples for each one were provided. This was followed by a picture finding trial, when the examiner told the patient to point to a given object (a half point was given when the category was correct or when a category cue was provided, as in the BDAE Word discrimination subtest). The last trial consisted of 12 ques-
tions referring to semantic knowledge about the objects. There were 2 multiple-choice format questions for each category, one about the object’s function and another one about other aspects of object knowledge. For instance, one of the questions concerning insects was: "Which insect catches flies?". The subject had to indicate the right picture among six given (spider, dragonfly, butterfly, ladybird, grasshopper).

The results presented in Table 3 (compared to a neurologically intact 84-year old control subject with 7 years of education) indicate that access to lexicon was severely impaired, as demonstrated by naming and verbal fluency trials. Word comprehension was significantly better than naming, which together with circumlocutions and correct pantomime usage (e.g. when unable to name tools) calls into question the possibility that the naming deficit could be attributed to poor visual recognition. The classification task showed slight confusion between fruit and vegetables, which is of diagnostic importance, as such difficulties did not appear in the control person. On no occasion was there an error caused by confusion between organic and non-organic things. In the semantic knowledge trial, the patient could answer 10 out of 12 questions. The failure in 2 questions was probably due to speech comprehension difficulties, as one of these question contained a negative, while another one required understanding relations. In the color attribution task, the subject committed 2 errors: throughout the trial the patient had difficulties in selecting the right color. For each item she had to be asked by the examiner to choose only one color. Semantic knowledge seemed to be relatively preserved, in contrast to severe word-finding difficulty.

In order to determine if word discrimination was impaired in relation to word-finding difficulty or a categorization deficit, tau B Kendall correlation coefficients were computed among word discrimination, naming and categorization. Word discrimination was related to naming (tau B Kendall=0.39; p<0.01), but not to categorization (tau B Kendall=-0.14; p=0.33). Naming did not correlate with categorization (tauB Kendall=0.184; p=0.21). Since cate-

Table 3. Experimental data on language processing and semantic knowledge in a patient with PPA and a control subject

<table>
<thead>
<tr>
<th>Category</th>
<th>Task patient/control (max. score)</th>
<th>Semantic knowledge-questions¹</th>
<th>Semantic knowledge-colors¹</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Verbal fluency</td>
<td>Naming</td>
<td>Word Comprehension</td>
</tr>
<tr>
<td>fruit</td>
<td>0/18</td>
<td>2/8 (8)</td>
<td>7,5/8 (8)</td>
</tr>
<tr>
<td>vegetables</td>
<td>3/14</td>
<td>3/8 (8)</td>
<td>6/8 (8)</td>
</tr>
<tr>
<td>insects</td>
<td>0/6</td>
<td>1/5 (8)</td>
<td>4/7 (8)</td>
</tr>
<tr>
<td>tools</td>
<td>0/11</td>
<td>1/6 (8)</td>
<td>6/8 (8)</td>
</tr>
<tr>
<td>vehicles</td>
<td>1/6</td>
<td>3/8 (8)</td>
<td>5.5/8 (8)</td>
</tr>
<tr>
<td>furniture</td>
<td>0/12</td>
<td>3/7 (8)</td>
<td>5.5/8 (8)</td>
</tr>
<tr>
<td>Global score</td>
<td>4/67</td>
<td>13/42 (48)</td>
<td>34/47 (48)</td>
</tr>
</tbody>
</table>

¹due to scarcity of items detailed scores for all categories are not presented
DISCUSSION

Regardless of the ultimate outcome in this case, the insidious onset, predominant language disturbance and progression of aphasia justify the diagnosis of fPPA as distinct from a generalized dementive process. Neuroimaging data did not reveal any other possible etiology of the disorder. Moreover, rCBF SPECT and MRI revealed changes mainly in the left temporal lobe, which are consistent with a predominant speech comprehension deficit. Neuropsychological assessment identified comprehension problems as the core deficit, accompanied by disturbed naming. Understanding complex sentences was impaired, while single-word comprehension was almost intact, which is in contrast to the profile characteristic for SD. Semantic knowledge was relatively preserved, when compared to severe word finding difficulty. The patient does not meet the criteria for SD, since:

- no signs of prosopagnosia or visual associative agnosia were identified;
- single words comprehension was mostly preserved;
- semantic knowledge seemed preserved.

Moreover, symptoms of aphasic acalculia and phonological disturbances were present. Naming was much better when provided with visual input (confrontation naming) relative to verbal input, while no modality effects are expected in SD (Garrard and Hodges, 2000). This pattern is consistent also with observations from two writing tasks with the same input modalities. The alexia and agraphia present in our patient were not typical for SD, as the errors were phonological. The neuropsychological data are consistent with neuroimaging results showing left temporal lobe atrophy and perfusion deficits in the language area. Both contribute to the diagnosis of fPPA.

Case reports published before 1990 suggested that the aphasia in PPA was most often fluent, anomic or Wernicke-like. However, since 1990, non-fluency has been considered by some to be a prominent hallmark of the syndrome (McNeil and Duffy, 2001; p. 476). The criteria proposed by Mesulam (2001) do not emphasize the distinction between fluency and nonfluency. In contrast, consensus statements on FTD emphasize fluency as one of several key features that distinguish two distinct forms of progressive aphasia: progressive non-fluent aphasia (PNFA) in PPA, and fPPA as an element of SD (Neary et al., 1998). The neuropsychological examination of our patient and neuroimaging data do not support this claim. Our case description supports the earlier observations of a broader clinical spectrum of language symptoms in PPA, and favors the view that the syndrome is not exclusively limited to the non-fluent aphasia type, as suggested by Neary et al. in 1998.

Some authors suggest that patients who meet the diagnostic criteria for
fPPA may also meet the diagnostic criteria for early-stage SD, provided that the impact of concept familiarity and typicality is taken into account. These patients, presenting with fPPA, had reduced grey matter density in the temporal lobes bilaterally (more widespread on the left), with the severity of atrophy in the left inferior temporal lobe being significantly related to performance on both verbal and non-verbal measures (Adlam et al., 2006). In our patient, neuroimaging data suggest a predominant left hemisphere dysfunction, with the involvement of anterior parts of the perisylvian language areas, as in PNFA, with inferior and middle temporal damage, as in fPPA (Gorno-Tempini et al., 2004). The atrophy we found in the anterior part of left temporal lobe seems typical for SD, however. In spite of the fact that abnormalities in neuroimaging are rather diffuse in our patient, they are restricted to the traditional "language area." In this case, neuroimaging does not allow for unquestionable confirmation of fPPA versus SD. However, in the context of the cognitive data, the neuroimaging results - mainly the asymmetry and localization of changes revealed by both MRI and SPECT - support the former diagnosis.

The comparison of the profile of our subject's language disturbances to other cases of fPPA described in the literature is hindered by the scarcity of data on fPPA, which is possibly due to its confusion with semantic dementia. It has been shown that in the fluent form, aphasic disturbance in PPA resembles Wernicke's aphasia (Radanovic et al., 2001) or transcortical sensory aphasia (Serrano et al., 2005). In our patient, the aphasic disturbance is phenomenologically close to transcortical sensory aphasia.

In a study by Radanovic et al. (2001), the average performance on language tests was better in the fluent groups regardless of the type of the task (oral comprehension, reading comprehension, naming, repetition, reading), whereas in classical aphasic syndromes the pattern of deficit would vary according to the aphasia syndrome. It may be that the comparison of language disturbance in fluent and non-fluent PPA patients may be obscured by, for example, the time from onset to language testing, and by the use of SD and fPPA as diagnostic labels in an inconsistent way. To shed light upon the differences between SD and fPPA, more prospective studies are needed, which could, in some cases, show conversion of fPPA to SD or PNFA.

The use of verbal-visual material (matching the right picture to a verbally presented question) in one of the semantic tasks in the experimental investigation is a possible shortcoming of our study. However, the fact that the patient's performance in this task was hindered by the speech comprehension deficits only in 2 of the 12 questions, taken together with the global results and consistent with other semantic trials (picture- color matching, categorization), allows us to conclude that semantic knowledge is better preserved than language function.

Language function testing was much more extensive than semantic/cognitive trials, which is another shortcoming of our assessment. In our patient, there were slight disturbances in semantic categorization, observed in the
classification experimental tasks, which could partially support the hypothes-
thesis of early-stage SD deficits in fPPA. However, the prevalence of circumlo-
cutions and presence of occasional phonemic paraphasias preclude a pri-
mary semantic deficit, as in this case semantic paraphasias would be more
frequent. At the time of our assessment, the language disturbance was much
more prominent than any other dysfunction that was revealed in the cognitive
testing. Since in fPPA the dementing process tends to generalize in the
course of the disease, it may comprise a semantic deficit that is comparable
to other aspects of cognitive dysfunction. This should be distinguished from
the dominant semantic disturbance in SD. Therefore, only follow-up can
show either conversion to SD or generalized dementia in this patient.

We suggest that error analysis in naming trials (especially the proportion
of semantic paraphasias and circumlocutions) could help in differentiating SD
from fPPA. Moreover, the comparison of confrontation naming and respon-
sive naming could provide information on modality-specific effects, when
worse responsive naming could serve as an indicator of fPPA, rather than
SD. Phonemic paraphasias in response to phonemic cues are also unlikely in
SD, while possible in fPPA.

It is recommended that the term semantic dementia be used in accord-
dance with the suggestions provided by Mesulam (2003), so as not to con-
found it with fPPA. Differential diagnosis could be supported by experimental
testing, assessing both the verbal and non-verbal knowledge of the same set
of objects, as presented in our study. A prospective analysis of semantic
deficits in fPPA should be undertaken in relation to other aspects of cognitive
deterioration.

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## Appendix: materials used

<table>
<thead>
<tr>
<th>Category</th>
<th>Items</th>
<th>Semantic questions</th>
</tr>
</thead>
</table>
| **Fruit**      | grapes (3), pineapple (9), banana (13), cherries (19), strawberry (26), pear (40), plum (42), apple (48) | What do we need to make apple-pie?  
Which fruit is imported from distant countries? |
| **Vegetables** | radish (5), lettuce (11), potato (17), carrot (25), peas (29), pepper (31), leek (33), cauliflower (36) | From which of these can juice be extracted?  
Which of these is pulled out of the soil? |
| **Insects**    | dragonfly (7), spider (8), mosquito (14), fly (16), butterfly (22), ant (32), grasshopper (39), ladybird (43) | Which insect catches flies?  
Which insects have a queen? |
| **Furniture**  | armchair (2), table (10), bookcase (18), sofa (23), chest of drawers (30), wardrobe (38), desk (41), bed (47) | Which piece of furniture is indispensable in the bedroom?  
What do we work on? |
| **Vehicles**   | bicycle (1), car (6), truck (12), coach (20), tractor (28), airplane (35), ship (44), helicopter (46) | Which vehicle is used for work on the farm?  
Which vehicle doesn’t have an engine? |
| **Tools**      | clippers (4), pliers (15), pincers (21), hammer (24), screwdriver (27), electric drill (34), saw (37), axe (45) | Which of these tools needs electricity?  
What do we use to hammer nails? |

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1. the numbers represent the order in which the items were administered in the naming, word comprehension and classification trials
2. the Polish equivalent of apple pie does not include the word “apple”
3. the Polish word for “bedroom” does not include the word “bed”
4. each object’s name is one word in Polish
5. the Polish word for an electric drill does not include the word “electric”
6. the Polish equivalent of the verb “to hammer” is not similar to the Polish noun for “hammer”