Case Report

Primary Progressive Aphasia (A Case Report)

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Abstract

Primary progressive aphasia (PPA), was first defined by Mesulam in 1982 as a "slowly progressive language dysfunction without generalized dementia". PPA is a neurodegenerative syndrome usually presented with anomia and reduced speech fluency, it is characterized by deterioration in language for least 2 years. Focal left perisylvian degeneration is thought to be responsible for this clinical syndrome. We present a 65 years old male patient, who presented with a progressive decline of language function, mainly in naming skills. Others mental functions were relatively preserved as well as an independence in the activity of daily livings. His cranial MRI imaging displayed a focal asymmetric left atrophy in the left anteriomedial temporal lobe. Asymmetrical hypoperfusion in the left parieto-temporal lobes were seen in his SPECT study. This case with clinical and radiological evidence of PPA is presented.

Keywords: Primary Progressive Aphasia, aphasia, language dysfunction

INTRODUCTION

The clinical criteria of Fronto-temporal lobe degenerations, which was published in 1998, includes 3 subcategories of this disorder. The most common form is Fronto-temporal Dementia (FTD) with changes in personality, behavioral problems and / or executive impairment. The other categories includes fluent aphasia (semantic aphasia) and non-fluent aphasia, both a subclass of primary progressive aphasia (PPA). In Semantic Dementia patients lose abilities to name and understand words and to recognise the significance of faces, objects and other sensory stimuli.
Non-fluent primary progressive aphasia (PPA) is is a disorder predominantly of expressive language, in which severe problems in word retrieval occur in the context of preserved word comprehension (1,4,11). It presents as an isolated, progressive language dysfunction for at least 2 years, without deterioration in other cognitive domains other than praxis (9,14). Although there is no specific type of language dysfunction for PPA, the most commonly affected language skills include word finding, verbal fluency and object naming (6,13,16).

Aphasia typically starts in presenium (before the age of 65), insidiously with a gradual worsening of symptoms over 5-20 years (12).

The neuropathological changes which include neuronal loss with gliosis and mild spongiform changes have been reported to be more pronounced in the left frontal, perisylvian and temporal cortices. Fewer than 20% of the patients show the pathology of Alzheimer’s disease (10,13).

We describe the clinical findings, cognitive profile and neuroradiological findings of a patient diagnosed with PPA.

**CASE PRESENTATION**

A 65 year old retired accountant reported a 3 year history of progressive language detoriation, mainly word finding and naming difficulties. He was otherwise healthy and his family history was negative. He retired 4 years ago with his own request. At that time he had some difficulty finding words but he was still capable of bookkeeping and he denies any cognitive or social problems at that time. His comprehension remained good. Recently he noticed some memory problems, he was especially forgetting names. He was also having difficulties ‘expressing himself’ which lead to anxiety, and he recieved treatment for panic attacks and depression.

On examination he was alert and anxious. His conversational speech was fluent with occasional word finding difficulties, word substitutions and some semantic paraphasias. He had impaired object naming with only 50% of correct answers. His comprehension, repetition, reading, writing and calculation functions were intact.

His Mini Mental State Exam (MMSE) was 27/30. During the Boston Naming Test (BNT) he was only able to name 7/31 objects correctly, he was able to describe another 14 and answered them correctly with the given choices. He gave wrong answers for 7 objects and was mute for 3. He displayed poor performance in Rey Auditory Verbal Learning test. He displayed a very poor performance during the verbal fluency tasks. He was only able to find 4 animals during one minute and 2 word pairs in separate categories. He described the Cookie Theft picture very briefly, with 3 short sentences and few words (Table 1). He completed 8 categories in the Wisconsin card sorting test. His performance on Stroop test, Trail making A test, Benton line orientation test, construction and abstract thinking were within the normal limits. He had 1/30 mistakes in pictorial recognition memory for faces and 4/30 mistakes in topographical recognition memory test. Apraxia was not present.

<table>
<thead>
<tr>
<th>Table 1: Description of the Cookie Theft picture from the patient.</th>
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<tbody>
<tr>
<td>Child climbed on a <strong>cookie</strong> to show a cookie. Giving something related to the cookie to the girl. <strong>Something about cleaning the cloths the lady is doing</strong></td>
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</table>

Magnetic Resonans Imaging Scan displayed left anteromedial temporal lobe atrophy (Picture1,2). Reduced perfusion more pronounced on the left was seen in both parietal and temporal lobes in the Single Photon Emission Computed Tomography (SPECT) study (Picture 3,4).
DISCUSSION

PPA was first described in 1982 by Mesulam, in a study of 6 patients who experienced slowly progressive language disorder and atrophy of the left perisylvian region. It was considered to be a focal cerebral degeneration restricted to language areas. The name “PPA” was proposed on a later review by Mesulam and Weintrub in 1992 (2). The diagnostic criteria for PPA was described by Mesulam in 2001 (9). (Table 2)
Table 2: The diagnostic criteria for PPA

1. Insidious onset and gradual progression of word-finding, object-naming or word comprehension impairments as manifested during spontaneous conversation or as assessed through formal neuropsychological testing of language.

2. All limitation of daily living activities can be attributed to the language impairment, for at least 2 years after onset.

3. Intact premorbid language functions (except for developmental dyslexia).

4. Absence of significant apathy, disinhibition, forgetfulness for recent events, visuospatial impairment, visual recognition deficits, or sensorimotor dysfunction within the initial 2 years of illness. This criterion can be fulfilled by history, survey of daily living activities, or formal neuropsychological testing.

5. Acalculia and ideomotor apraxia can be present even in the first 2 years. Mild constructional deficits and perseveration are also acceptable as long as neither visuospatial deficits nor disinhibition influence daily living activities.

6. Other domains may become affected after the first 2 years, but language remains the most impaired function throughout the course of the illness and deteriorates faster than other affected domains.

7. Absence of specific causes such as stroke or tumor as ascertained by diagnostic neuroimaging.

We examined a 65 year old patient, 3 years after the onset. His main problem was word finding and naming difficulties. Even though he mentioned recent memory problems the results of his memory tests were normal. Activities of daily living were preserved. He was diagnosed with PPA according to Mesulam’s criteria for PPA.

Memory, visuospatial abilities, reasoning and social functioning are all preserved in early PPA. Although language dysfunction may cause poor performance in some memory tests, these patients mostly function normally during their daily lives.

Most errors were found during the BNT and verbal fluency tests in our patient. Memory functions were preserved but he still had trouble during the verbal memory tasks. He was able to describe the words correctly but he did have trouble naming them, probably related by naming difficulties.

Studies in patients with PPA have shown that the executive and visuospatial skills are preserved during the first few years of the disease. Our patient was successful during the Wisconsin cart sorting test which is a test of the integrity of frontal lobe functions and measures attention, perseveration, working memory, executive functions and abstract thinking. Our patient completed another frontal lobe function test, the stroop test slowly (due to his word finding difficulties) but correctly. Abstract thinking and visuospatial skills were normal.

In PPA patients neuropathological studies displayed neuronal loss and focal spongiform changes in the left perisylvian, frontal and temporal cortex areas rather than in the hippocampal or entorhinal areas which are the areas most affected in Alzheimer’s disease.
Neuroimaging with cranial computed tomography (CT) images may be normal in PPA patients for the first 3 to 8 years, MRI, positron emission tomography (PET) and SPECT may be more valuable for detecting the left hemisphere changes and focal atrophy (2,7). Focal perisylvian atrophy in the left lateral temporal lobe (superior and inferior gyrus) has been shown in most MRI studies, with nonspecific or generalized atrophy in other studies (2,5,12,15). Hypometabolism focally or generalized in the left hemisphere has been reported in more than 70% of patients in PET and SPECT studies (2,12). It has also been shown that the focal left temporal and perisylvian changes which can be seen early in the disease course spreads to the parietal and frontal areas with the progression of the disease (15).

Our case is a good example for PPA clinically with isolated word finding and naming difficulties for 3 years and radiologically with focal left anteromedial temporal lobe atrophy on MRI and bilateral parietal and temporal lobe hypometabolism, more pronounced on the left hemisphere in SPECT.

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