The Mild Cognitive Impairment of Primary Progressive Aphasia: A Case Series

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Abstract

The early and mild phases, termed mild cognitive impairment (MCI), of primary progressive aphasia (PPA) have been poorly described. This cross-sectional care series was conducted via retrospective chart review in order to further delineate the MCI stage during progression to PPA.

Nine cases of PPA with a secondary diagnosis of MCI were found, all of which had language as the primary domain affected. The most common deficits found were word finding difficulty (8 of 9 cases) and sentence repetition impairments (8 of 8 cases).

Our results showed that there is an MCI stage of PPA, characterized by word finding difficulty on observation and sentence repetition on neurocognitive testing. Early mild cognitive impairment for PPA will have language as the primary domain affected either with or without an amnestic component.

Introduction

Mild Cognitive Impairment (MCI) has been studied as a prodromal predictor of developing Alzheimer's disease (AD), Parkinson's disease (PD), and Dementia with Lewy Bodies (DLB)[1,8,9]. However, little research has been done to describe the MCI stage for a suspected etiology or Primary Progressive Aphasia (PPA). Pathology of subjects clinically diagnosed with MCI often show the same neuropathology as subjects diagnosed with dementia, indicating irreversible damage has already taken place even before a MCI diagnosis is made or symptoms are seen[10]. As such, there is a desire to understand the earliest stages of cognitive decline and research is trending toward studying subjects in the earliest phases of MCI and eMCI (early MCI), a stage in which the very first symptoms are recognized despite presence of a high level of functional status [2,4,11].

Primary Progressive Aphasia (PPA) is a dementia described by a progressive degeneration of language. While other cognitive symptoms may present throughout the course of disease, a clinical classification of PPA must include language impairment as the initial, most prominent symptom and must be the main contributor to deficits affecting activities of daily living. Early methods of diagnosing PPA demand a 2 year waiting period in which the language domain remained the primary area of decline[5,6,7]. This approach discourages early diagnosis which is known to be of benefit in treatment and research.

This study aims to identify the clinical phenotype of MCI as described by Petersen[9] as a prodrome to PPA. In addition, we hope to add to the understanding of very early symptoms and signs of PPA MCI by including 3 patient profiles with eMCI in which language is the main complaint and most prominently observed deficit.

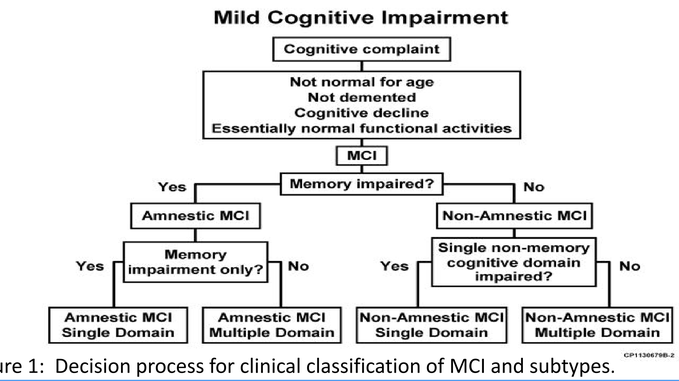

Figure 1: Decision process for clinical classification of MCI and subtypes.

Methods

Table 1. Demographic Data of Individual Cases (*Denotes EMCI)

	Age	Gender	Education	Disease Duration
*Case 1	77	Female	16	2
*Case 2	58	Female	18	2.5
Case 3	78	Male	13	5
*Case 4	61	Female	16	1.5
Case 5	71	Female	16	5.5
Case 6	73	Male	16	20
Case 7	83	Male	20	2
Case 8	68	Female	12	3.5
Case 9	71	Male	14	12

A query of the BSHRI memory disorder clinic database was conducted utilizing ICD9 codes 331.83 (MCI) and 784.3 (aphasia), generated 9 viable subjects for the study, patient demographics are summarized in Table 1. A retrospective chart review was conducted. A neurologist (MS) evaluated each patient through comprehensive neurological exam and a neuropsychologist (JP) confirmed domains of impairment as well as cognitive diagnosis by a battery of neuropsychological tests (Table 2).

Table 2. Neuropsychological Test Results Characteristics of Individual Cases (z-scores)

	Confrontation Naming (BNT)	Semantic Fluency	Phonemic Fluency (COWAT)	Written Spelling	Oral Spelling	Comprehension (Token) or (Complex Ideation)	Sentence Repetition
*Case 1	-1.00	-1.31	1.00	0.55	0.55	0.00	-1.04
*Case 2	1.00	0.20	-1.35	-0.25	-0.25	N/A	-1.48
Case 3	0.35	-1.31	-1.35	0.55	-1.55	0.80	-1.04
*Case 4	-1.35	-0.29	1.35	0.55	0.55	N/A	N/A
Case 5	1.00	-0.05	1.35	-0.84	-0.25	-2.00	<-2.35
Case 6	-1.00	-3.38	-1.65	-0.28	N/A	<-2.35	<-2.35
Case 7	-0.65	<-2.35	-0.35	-1.55	<-2.35	N/A	-1.89
Case 8	<-2.35	-2.65	-0.35	-1.55	-1.55	N/A	-1.48
Case 9	<-2.35	-1.71	0.35	-0.84	-1.04	-1.60	-1.48

*Denotes eMCI cases; Lightly shaded cells indicate suspect impairment as defined by >-0.5 to -1.49; Darkly shaded cells indicate impairment as defined by >-1.5 standard deviations below the mean.

Conclusions

Word finding difficulty and memory complaints are the most common self-reported characteristics and word finding problems are the most commonly observed speech behavior to appear in the MCI state for clinically suspected PPA. Neuropsychological testing revealed that sentence repetition, semantic fluency, and confrontation naming were the most impaired objective measures. Of the 9 patients, 5 were non-amnestic and 4 were amnestic. Of those with eMCI, all were non-amnestic single domain (language). These findings are similar to those previously described by Mesulam[6] where word finding difficulties were the most typical complaint and objectively, word finding hesitation and sentence repetition were the most common deficits.

Acknowledgements

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Results

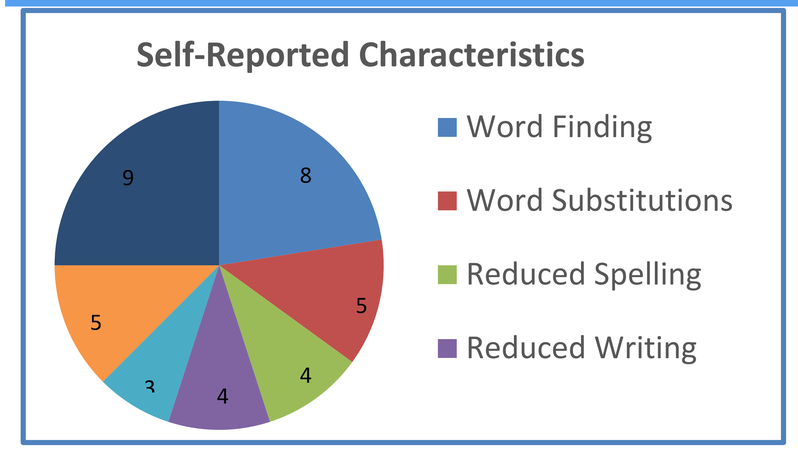

Figure 2. Self Reported Characteristics of Individual Cases

Word finding was the most prevalent observation. Substitutions and comprehension were second. Few patients had paraphasias and Re-takes. None displayed neologisms or agrammatisms.

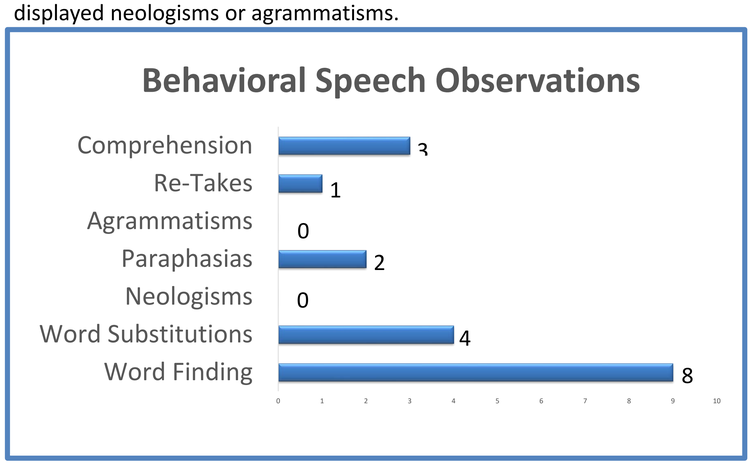

Figure 3. Behavioral Speech Observations of Individual Cases

Memory and word finding were the most prevalent complaints. Second were word substitutions and reduced comprehension. Least common were reduced spelling, writing and reading.

These results highlight the importance of subjective complaints, behavioral observations, and objective testing when working with suspected PPA. While sentence repetition deficits alone may not be specific enough to differentiate between dementia subtypes, we suggest that a possible diagnosis of prodromal PPA should be considered if word finding difficulties are the primary subjective complaint and reduced sentence repetition is found on neuropsychological testing, with or without the presence of memory impairment

Table 3. Neuropsychological Diagnosis

	Neuropsychological Diagnosis of Individual Cases
Case 1	Nonamnestic, single-domain early MCI (language)
Case 2	Nonamnestic, single-domain early MCI (language)
Case 3	Nonamnestic, multiple-domain MCI (language, attention)
Case 4	Nonamnestic, single-domain early MCI (language)
Case 5	Nonamnestic, single-domain MCI (language)
Case 6	Amnestic, multiple-domain MCI (language, executive, visuospatial)
Case 7	Amnestic, multiple-domain MCI (language, executive)
Case 8	Amnestic, multiple-domain MCI (language)
Case 9	Amnestic, multiple-domain MCI (language)