Language evaluation in semantic dementia

Bora Yoon, MD
Department of Neurology, Konyang University

CLASSIFICATION OF PPAS

Table 1. Descriptive and simplified criteria for classifying primary progressive aphasia.

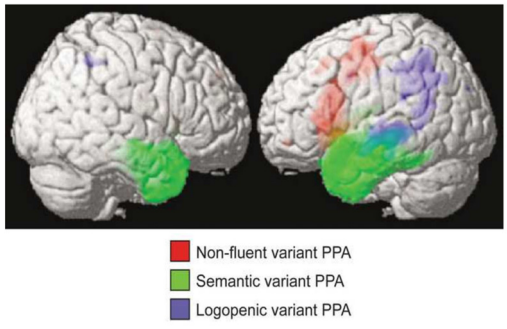
Diagnostic criteria for PPA	The following three conditions must all be present. 1. A new and progressive language disorder (aphasia) as documented by neuropsychologically determined abnormalities in one or more of the following domains: grammaticality of sentence production, word retrieval in speech, object naming, word and sentence comprehension, spelling, reading, repetition. Isolated impairments of articulation do not qualify. 2. Initial and relative preservation of episodic memory, executive functions, visuospatial skills and comportment as documented by history, medical records and/or neuropsychological testing. 3. Imaging and other pertinent neurodiagnostic test results that rule out causes other than neurodegeneration.
Agrammatic Subtype (PPA-G) **PNFA**	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) **SD**	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. Current classification systems require repetition impairments for diagnosing this subtype (19).
Anomic Subtype (PPA-A)	All features as in PPA-L except that repetition is intact.
Mixed Subtype (PPA-M)	Impaired grammatical structure and word comprehension, even at the early stages of disease.

LANGUAGE CHARACTERISTICS IN PRIMARY PROGRESSIVE APHASIAS

Table 1. Summary of speech and language characteristics in PNFA, SD and LPA

Ppa Variant	Most Common Deficits	Preserved Behaviors	Neuroanatomy	Pathology
Progressive non-fluent aphasia (PNFA)	• agrammatic output • motor speech impairments (apraxia of speech, dysarthria) • impaired comprehension of complex sentences • anomia (particularly for verbs)	• semantic processing • comprehension for single-words and simple sentences • auditory-verbal short-term memory (AVSTM)	Predominant left posterior fronto-insular atrophy	• Corticobasa degeneration • Progressive supranuclear palsy
Semantic dementia (SD)	• severe anomia • poor single-word comprehension (particularly for low-frequency items) • multi-modal semantic memory impairment • surface dyslexia	• motor speech • expressive syntax (fluency) • sentence comprehension	Predominant left anterior temporal lobe atrophy	Frontotemporal dementia-related pathology
Logopenic progressive aphasia (LPA)	• anomia • poor phase and sentence repetition due to impaired AVSTM • phonological errors in spontaneous speech and naming	• motor speech • semantic processing • single-word comprehension	Predominant left posterior perisylvian or parietal atrophy	Alzheimer's disease

NEUROANATOMY IN IMAGING



CASE 1

CASE 2

DIAGNOSTIC CRITERIA OF SD

Table V. Diagnostic features of semantic dementia (adapted from Gorno-Tempini et al.[4])

Clinical SD	Imaging-supported SD	SD with definite pathology
Impaired confrontational naming	Clinical diagnosis of SD	Clinical diagnosis of SD
Impaired single-word comprehension	Neuroimaging showing anterior temporal atrophy	Histopathological evidence of a specific neurodegenerative pathology
Impaired object knowledge	Predominant anterior temporal hypoperfusion or hypometabolism on PET or SPECT	Genetic evidence of a known pathogenic mutation
Surface dyslexia or dysgraphia		
Intact repetition and speech production		

PET = positron emission tomography; **SD** = semantic dementia (semantic variant primary progressive aphasia); **SPECT** = single-photon emission computed tomography.

Seltman RE & Matthews BR. CNS Drugs 2012;26:841-870

DIAGNOSTIC CRITERIA OF PNFA

Table IV. Diagnostic features of progressive non-fluent aphasia (adapted from Gorno-Tempini et al.[4])

Clinical PNFA	Imaging-supported PNFA	PNFA with definite pathology
Agrammatism	Clinical diagnosis of PNFA	Clinical diagnosis of PNFA
Apraxia of speech	Neuroimaging showing predominant left posterior frontoinsular atrophy	Histopathological evidence of a specific neurodegenerative pathology
Impaired comprehension of complex sentences	Predominant left posterior frontoinsular hypoperfusion or hypometabolism on PET or SPECT	Genetic evidence of a known pathogenic mutation
Intact single-word comprehension and object knowledge		

PET = positron emission tomography; **PNFA** = progressive non-fluent aphasia (non-fluent variant primary progressive aphasia); **SPECT** = single-photon emission computed tomography.

Seltman RE & Matthews BR. CNS Drugs 2012;26:841-870

Clinical & language characteristics of PPA variants

	SD	PNFA	LPA
Spontaneous speech	o	x	Δ
Single-word comprehension	x	o	o
repetition	o	x	x
naming	x	x	x
Sentence comprehension	x	Δ	x
Associated symptoms	Multimodal semantic impairment (faces, objects, sounds)/behavioral symptoms	Buccofacial & limb apraxia/mild contralateral motor symptoms	Acalculia/apraxia
Main lesions	Lt. inferior frontal/insular area	Lt.>>Rt. anterior temporal	Lt. Inferior parietal & posterior temporal

THANK YOU.