



CASE SERIES: DIFFERENT VARIANTS OF PRIMARY PROGRESSIVE APHASIA

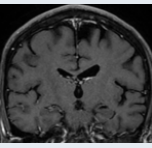
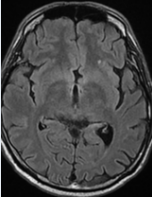
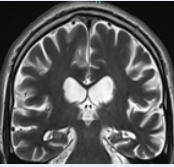
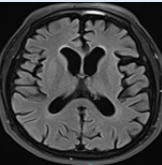
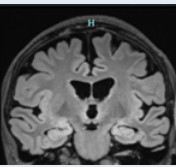
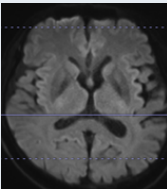
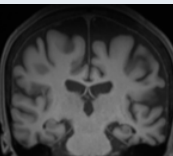
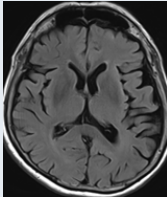
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Introduction

The primary progressive aphasia (PPA): a group of disorders consisting of logopenic, semantic, and nonfluent/agrammatic variants. Although it is being diagnosed with increasing frequency, PPA is still a rare syndrome. The PPA syndrome arises when the language-dominant (usually left) hemisphere becomes the principal target of neurodegeneration. It is often difficult to identify the underlying neurodegenerative disease etiologies for patients with progressive speech disorders. Here we report on 4 cases of Progressive Primary Aphasia of different variants.

	Case 1. Male, 54 yo	Case 2. Male, 59 yo	Case 3. Female, 50 yo	Case 4. Female, 72 yo
HISTORY	Halting speech and word-finding difficulties for 1 year, followed by confusion and decline in daily function.	Progressive anomia and semantic memory loss over 3–4 years, with emotional lability.	Slow, effortful speech and behavioral withdrawal over 2 years, with language and memory decline affecting daily function.	Word-finding difficulty and sentence formulation issues over 1 year. Initial memory was relatively preserved.
NEUROLOGICAL EXAM	Within Normal Limit	Within Normal Limit	Within Normal Limit	Within Normal Limit
NEUROPHYSIOLOGICAL EXAM	ATTENTION			
	Impaired	Normal	Impaired	Impaired
	LANGUAGE			
	Nonfluent, anomia, impaired complex comprehension and repetition, preserved reading, impaired writing	Fluent, anomia, comprehension intact, impaired repetition, normal reading & writing	Nonfluent, anomia, impaired comprehension, impaired repetition, slow reading, impaired writing	Fluent, anomia, impaired comprehension, repetition, reading (paralexia), writing (paragraphia), paraphasia
	VERBAL FLUENCY (SEMANTIC / PHONEMIC)			
	3/0	11/NA	3/1	1/5
	BNT			
	8/15	7/15	7/15	3/15
	TOKEN TEST			
	15.5/36	Not Available	11/36	10.5/36
	MEMORY			
	Impaired in immediate, recent, recall, recognition memory	Impaired in recent, recall, recognition memory	Impaired in recent, recall, recognition memory	Impaired in immediate, recent, recall, recognition memory
	EXECUTIVE			
	Impaired	Normal	Impaired	Impaired
	VISUOSPATIAL			
	Normal	Normal	Impaired	Normal
	TOTAL CERAD			
	27/100	23/100	26/100	20.5/100
	MMSE			
	14/30	17/30	10/30	13/30
	MOCA-INA			
	8/30	14/30	2/30	5/30
BPSD	None	Agitation	None	None
BRAIN MRI	 	 	 	 
DIAGNOSIS	Logopenic variant PPA	Semantic Variant PPA	Non-Fluent Variant PPA	Logopenic variant PPA

Discussion

The PPA syndrome is diagnosed when 3 criteria are met, insidious onset and gradual progression of a language impairment (i.e., aphasia), aphasia should initially arise as the primary impairment, and diagnostic testing should point to a neurodegenerative, and therefore progressive, process as the only underlying cause. Three patients had early-onset dementia. All of them had aphasia at the beginning of their symptoms but came to the hospital when the symptoms had worsened and memory became impaired, making it difficult to assess. The underlying pathology of our patients suggests either Alzheimer’s disease or of the frontotemporal dementia pathologies, therefore it is important to evaluate the biomarker probability for AD etiology.

Conclusion

This case series emphasizes the necessity of evaluating patients with PPA through a diagnostic framework to classify the variant and therefore, to work out the underlying etiology. It is important to assess aphasia early in the disease course.

References:

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