

Case Report

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Cognitive-linguistic diagnostic profile of anomie aphasia in moyamoya disease: A case study***Corresponding Author: Sandhya K**

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Abstract

Moyamoya syndrome is a type of progressive cerebro vascular disorder characterised by progressive narrowing of arteries of the circle of Willis. Arteries get clogged because of growth of prominent small vessel due to which blood supply to the brain is reduced [1]. A 31 years old male was a pre-diagnosed case of Moyamoya Disorder (MMD) with hemiplegia of right side. His MRI reports revealed Magnetic Resonance Imaging brain angiogram was done which showed patchy left Middle Cerebral Artery (MCA) territory infarcts with involvement of left anterior basal ganglia, left insular cortex and patchy frontal and parietal infarcts as well as left Internal Carotid Artery (ICA) occlusion. He presented with a complaint of difficulty in recalling names. Based on Medical history, formal and informal evaluations, the patient was diagnosed as having Anomic Aphasia with Cognitive deficit secondary to MMD. The case study has attempted to provide a comprehensive picture of the patient's cognitive linguistic profile.

Keywords: Moyamoya disorder; MMD; Anomic aphasia; Cognitive-linguistic disorder.

Introduction

Moyamoya syndrome is a type of progressive cerebro vascular disorder characterised by progressive narrowing of arteries of the circle of Willis. Arteries get clogged because of growth of prominent small vessel. As a result, blood supply to the brain is reduced [1]. Despite the fact that it can be found all over the world, it is more widespread in East Asian countries. Two peaks of onset are observed in Moyamoya syndrome where one is in childhood and the other is in young adulthood [2]. The ischemic type has been shown to be predominate in childhood, while the hemorrhagic type is more often observed in the adult population [3].

This disorder is characterised by hazy appearance on angiography resembling a puff of smoke in the air and has thus been given the name Moyamoya, a Japanese term meaning the same. Japan has the highest incidence of MMD, with three cases per 100,000 [1]. The clinical presentation of MMD includes Transient Ischemic Attacks (TIA), ischemic stroke, hemorrhagic

stroke, headache, epilepsy and cognitive dysfunction [4]. Clinical profile of MMD includes headache, cognitive impairment, hypertension, temporary or permanent blindness, hemiplegia, general paresis of the insane, loss of sensation, aphasia, and mechanisms related to ischemia in the frontal, parietal, and temporal lobes [5].

Researches have evidenced that Moyamoya disease is associated with Cognitive deficits and aphasia. An estimated one million individuals in the United States have aphasia and an estimated 80,000 new cases are thought to be diagnosed each year (American Speech-Language-Hearing Association [ASHA], 2014b) [6]. Aphasia is an acquired condition caused by a disease or trauma to parts of the brain involved in language processing. According to ASHA (2014b) [6], the most prevalent cause of aphasia is stroke. An estimated 25-40% of stroke survivors are diagnosed with aphasia (National Aphasia Association, 2014) [7]. It may affect auditory comprehension, verbal expression, reading, and writing and word-finding abilities along with sen-

sory-motor impairments. Anomic aphasia can be a difficulty in retrieving precise nouns, often intended words, but occasionally verbs, when creating sentences. Although the grammatical features of the sentence remain intact, word retrieval difficulty persists that leads to insertion of filler words, long pauses or choice of wrong words (paraphasias) [8]. Though there are a couple of case reports on cognitive deficits in adults with MMD, limited Indian researches available which documents the structured assessment of language and cognitive abilities and hence, the present study gives a picture of detailed evaluation of a patient of MMD.

Case presentation

A 31 years old male, presented with sudden onset of right side weakness and inability to speak since that morning. The patient is married and works as a Software Engineer. The patient reported to the department of Speech and Hearing with a complaint of difficulty in recalling names. He was a pre-diagnosed case of MMD with hemiplegia of the right side.

Medical history

On arrival, his Blood Pressure (BP) was 150/100 mmhg, Glasgow Coma Scale (GCS) E4M5V1, pupils 2 mm. He complained of slurring of speech and inconsistent response to name call. Magnetic Resonance Imaging brain angiogram was done which showed patchy left Middle Cerebral Artery (MCA) territory infarcts with involvement of left anterior basal ganglia, left insular cortex and patchy frontal and parietal infarcts as well as left Internal Carotid Artery (ICA) occlusion. The patient was intubated and admitted for Digital Subtraction Angiography (DSA) and emergency mechanical thrombectomy. DSA showed severe narrowing of Left supraclinoid ICA, right terminal ICA and right MCA severe narrowing, left MCA occlusion - likely Moyamoya syndrome.

Mechanical thrombectomy was abandoned in view of DSA finding, Neurosurgery opinion was obtained and patient was admitted in Medical Intensive Care Unit (MICU) started on anti-edema, anticonvulsant, antiplatelets and other neuroprotective measures. Gradually his GCS was improved to E4M6 (to visual cues). CT brain showed Acute- to -subacute infarct in left fronto-temporal operculum, insular cortex, external capsule, caudate and lentiform nuclei. Mass effect with compression of left lateral and 3rd ventricle and midline shift of 5 mm to right, chronic lacunar infarct in right hemisphere and bilateral ethmoid and sphenoid sinusitis was noted. It also showed no significant interval change in infarcts volume and mild effacement of left frontal sulci and gyri.

Language evaluation

On perceptual evaluation, it was noted that he was able to follow commands and give relevant answers to the questions asked. His working memory was intact whereas semantic memory, processing speed and his reasoning were impaired. Speech mechanism examination revealed normal structure and function of articulators. Additionally, he exhibited impaired performance in verbal and visual working memory tasks, while his episodic memory was normal. WAB -K (Kannada version) was administered and AQ of 88.6 was obtained. He performed well in all other domains except Naming.

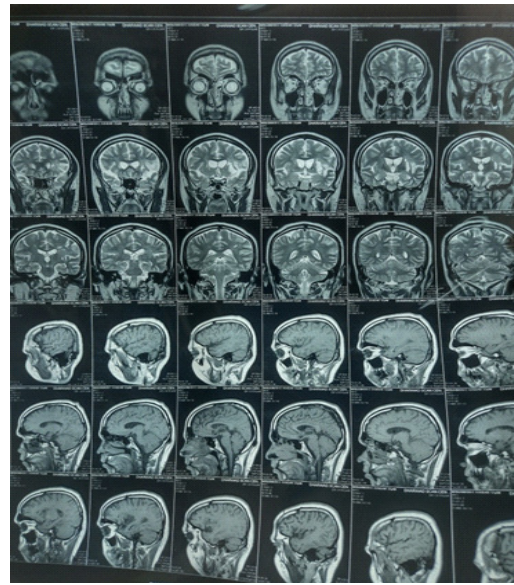


Figure 1: CT scan image.

Table 1: WAB scores of the patient.

WAB domains	Max score	Patient score	AQ
Spontaneous Speech			
Information Content	10	8	
Fluency	10	10	18
Comprehension			
Yes/No question	60	60	
Auditory word recognition	60	60	9.7
Sequential commands	80	74	
Repetition	100	92	9.2
Naming			
Objective Naming	60	56	
Word fluency	20	4	
Sentence completion	10	6	7.4
Responsive speech	10	8	
Aphasia quotient			88.6

It was observed that in general conversation he had difficulty in word retrieval. Analysis of speech sample revealed semantic paraphasias (Substitution of another word of same category) and circumlocutions (a deliberate attempt to express a particular word), hesitations and word finding pauses. Naming domain of WAB-K showed impaired word fluency and sentence completion abilities whereas confrontation naming and responsive speech were relatively spared. Based on observation and test results a diagnosis of Anomic Aphasia was made at the Department of Speech and Hearing OPD.

Linguistic Assessment Protocol developed by Aruna (2001) [9] was administered to assess his Cognitive abilities. The CLAP consists of four domains including attention (visual & auditory category), memory (episodic, working & semantic), problem solving (sentence disambiguation, sentence formulation, comparing and contrasting two objects, predicting the outcome of a described situation, Predicting cause of a described situation, answering 'why' questions & sequential task analysis) and organization (categorization, analogies & sequencing of events).

With regard to the patient's cognitive profile, the above mentioned scores show a greater difficulty in attention, perception, discrimination, memory, reasoning and problem solving and organization. Based on all these observations, the patient was diagnosed as having Anomia Aphasia with Cognitive deficit secondary to Moyamoya Syndrome and was subjected to Speech and language therapy.

Table 2: Scores of CLAP of the patient.

Domain	Test item	Maximum	Patient's score
I. Attention, Perception and Discrimination			
Visual	Letter Cancellation	10	5
	Contingent Letter Cancellation	10	4
	Word Cancellation	10	6
Auditory	Sound count Letter-Pair	10	2
	discrimination Months	5	3
	discrimination Word –Pair	5	2
	backwards naming	10	2
II. Memory			
Episodic Memory	Orientation and recent memory questions	10	8
Working Memory	Digit Forward	5	5
	Digit backward	5	4
Semantic Memory	Co-ordinate naming	5	3
	Super ordinate naming	5	3
	Word-naming fluency	5	2
	Generative naming	5	2
	Sentence repetition	10	6
	Carry out commands	10	5
III. Reasoning & Problem Solving			
	Sentence disambiguation	10	6
	Sentence formulation	5	3
	Predicting outcome	10	5
	Compare and contrast	10	5
	Predicting cause	10	6
	Why Questions	5	2
	Sequential Analysis	10	6
	IV. Organization	Categorization	10
	Analogies	10	5
	Sequencing events	40	25

Source: Cognitive-linguistic Assessment Protocol for adults (Aruna, 2001) [9].

Discussion

The current study gives a complete Cognitive linguistic profile of a patient of Anomic Aphasia with Cognitive deficits following an MMD induced stroke. The patients with MMD have an increased risk of stroke development, which may be aggravated by pain or anxiety. It is necessary to avoid cerebral ischemic attacks caused by fluctuating BP and respiration. Pre-surgical neuropsychological testing results for persons with MMD reveal that they showed executive functioning impairment with a reasonably unimpaired memory function [10], whereas the current study contradicts the previous evidences. However, several studies have shown significant relationships between language comprehension, for example, and different facets of cognitive control, such as working memory [11] and inhibition [12-14]. A

research [11] has shown that performance on a modified listening span task was linked to language comprehension abilities, although only in people with nonfluent aphasia, and not for people with fluent aphasia. Findings of the present investigations appear to be in agreement with previous researches.

Clinical significance

Prior to developing the treatment plan, it is essential to conduct a complete assessment of all relevant criteria, including the degree of involvement and medical history. Additionally, neuropsychological assessments help in formulating intervention plans for maintaining current cognitive performance. Such procedures are essential in continuous rehabilitation efforts and enhance the quality of life for person with Cognitive linguistic disorders. The external validity of the current study, which focuses on a single case profile, is constrained when applied to other MMD cases, but the author expects that this proof will add to the information already available.

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