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A Systemic Analysis Language Deficits in Moyamoya Disease with Childhood Onset and Adolescent Onset

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ABSTRACT

Moyamoya is a Japanese word for a "cloud of smoke" or "haze". Moyamoya disease also known as Nishimoto-Takeuchi-Kudo disease is a rare, progressive vascular disorder in which the carotid artery in the skull becomes blocked or narrowed, reducing blood flow to the brain. The onset of Moyamoya disease in children is 5-10 years and in adult is 30-50 years. The signs and symptoms of Moyamoya disease will have difference according to their age of onset. The chances of cognitive impairments are more in childhood onset especially in a prelingual period when compared to adolescent and adulthood onset. Later these cognitive impairments may lead to several developmental disabilities and communication breakdowns. In adolescent or adulthood onset the chances of cognitive impairment are less and they may show an aphasic or dysarthric type of communication impairment. Aim of this study is to compare the communication impairments of Moyamoya disease with childhood onset and adolescent onset. The study was done by taking two female participants with Moyamoya disease. The first subject was with a childhood onset at the age of 3.9 years and the second subject was with an adolescent onset at the age of 16 years. Both the participants' speech and language skills were assessed using standardized tools for communication assessments. The study concludes that, Moyamoya disease with childhood onset will lead to language learning deficits accompanied by intellectual disabilities and in case of adolescent onset the communication impairments will be in the form of Aphasic and Dysarthric type of disturbances.

Key words: Moyamoya disease; Aphasia; Dysarthria; Intellectual Disabilities

INTRODUCTION

Moyamoya is a Japanese word for a "cloud of smoke" or "haze". Moyamoya disease also known as Nishimoto-Takeuchi-Kudo disease is a rare, progressive vascular disorder in which the carotid artery in the skull becomes blocked or narrowed, reducing blood flow to the brain[1] and it was first described by Takeuchi and Shimizu in 1957. Tiny blood vessels then open up at the base of the brain in an attempt to supply with blood. It is referred as a form of large

vessel occlusion affecting the intracranial supra clinoid carotid artery, in which a secondary network of proliferating collateral arterioles is seen to arise from the carotid artery proximal to the occlusion. The exact cause of Moyamoya disease is unknown. Moyamoya disease is more common in Japan, Korea and China, but also in other parts of the world. Primary Moyamoya disease may be genetically transmitted in an autosomal recessive trait, and account for approximately 10% of all cases in Japan. Secondary Moyamoya disease occurs in association with a number of different underlying disorders or conditions, including certain infections involving the central nervous system, neurofibromatosis type 1, sickle cell disease and down syndrome. The incidence of the disease is more in Japan. Prevalence and incidence of the disorder there has been reported to be 3.16 cases and 0.35 case per 100,000 people, respectively[2]. The male to female ratio is 1:1.4.

Signs and symptoms of this disease are Stroke with hemorrhage from abnormal blood vessels, headache, seizures, weakness, numbness or paralysis in face, hemiparesis, visual disturbances, difficulty in speaking, developmental delay, involuntary movements etc. The onset of Moyamoya disease in children is 5-10 years and in adult is 30-50 years[2]. Children with such an abnormality present with hemiplegia, cerebral ischemia, reduced brain blood supply, including stroke, transient ischemic attacks, headaches, seizures, involuntary movements, or occasionally progressive developmental delay and adults were present with signs and symptoms of brain ischemia, they also have a greater tendency to suffer intracranial hemorrhage. Although dementia, dysphagia, pyramidal or extrapyramidal signs and cerebral atrophy have also been documented. The signs and symptoms of Moyamoya disease will have difference according to their age of onset. The chances of cognitive impairments are more in childhood onset especially in a prelingual period when compared to adolescent and adulthood onset. Later these cognitive impairments may lead to several developmental disabilities and communication breakdowns. In adolescent or adulthood onset the chances of cognitive impairment are less and they may show an aphasic or dysarthric type of communication impairment. The diagnosis of Moyamoya disease is primarily based on neurological evaluation, signs and symptoms, family history, medical history, physical examination and neuroimaging studies.

Need of the Study

The rehabilitation procedure of the persons with Moyamoya disease should vary according to their nature impairment. The treatment of an adult with developmental delay and acquired communication disorders are different. Hence, as there are evidences of Moyamoya disease with an onset in different age levels, the understanding of difference in the characteristics of this disease in various age of onset is vital in the rehabilitation.

Aim of the Study

Aim of this study is to compare the communication impairments of Moyamoya disease with childhood onset and adolescent onset.

METHOD

The study was done by taking two female cases with Moyamoya disease as subjects. The first subject was with a childhood onset at the age of 3.9 years and the second subject was with an adolescent onset at the age of 16 years. The assessment of communicative and cognitive skills was done at the age of 12 years for the subject 1 and 19 years for the subject 2. The diagnosis of Moyamoya disease was under the basis of neurological investigations and neuroimaging studies. The subject 1 had three massive attacks of stroke from 3·9 to 4 years of age and underwent left Encephalo-Duro-Arterio-Myo-Synangiosis (EDAMS) at the age of four years. Subject one's communication skills are assessed through

perceptual analysis of speech and language skills, administration of Frenchay Dysarthria Assessment[3] and Linguistic Profile Test[4] (LPT) Malayalam version. The assessment of cognitive abilities was done by using Raven's Coloured Progressive Matrices[5] (RCPM). The study also considered Malin's Intelligence Scale for Indian Children (MISIC) values to identify the subject's intelligence quotient.

The subject 2 had an apparently normal development till the age of 16 years and three attacks of cerebrovascular accident reported from 16 to 17 years. The assessment of communication impairments was conducted by perceptual assessment of semantic, pragmatic and cognitive skills, language comprehension and expression, perceptual and subjective assessment of speech skills, assessment of dysarthria component using Frenchay Dysarthria Assessment[3], assessment of aphasia component using Western Aphasia Battery[6] Malayalam translated version. To know the cognitive functioning level of the subject, psychological assessment reports based on Mini Mental State Examination[7] (MMSE) and the Post Graduate Institute of Psychiatry - Battery of Brain Dysfunction (PGI-BBD) were considered.

RESULTS

The results showed that the subject 1 and 2 is having a severe impairment in communicative abilities. The subject 1 had difficulties in both speech and language skills associated with cognitive deterioration. On the assessment of speech skills, subject showed mild voice problems whereas articulation and fluency were normal. Also, subject showed irregularity in laryngeal functioning after evaluated with Frenchay Dysarthria Assessment[3].

While coming to the language aspects the of the subject 1, child had an inadequate pragmatic and cognitive skills with a language age of 8+ years on Linguistic Profile Test[4], while the semantic relations were adequate. The cognitive assessment was done with Raven's Coloured Progressive Matrices[5] and the score was 16 out of 36 which indicates mild cognitive dysfunction. The subject had an Intelligence Quotient of 67 when evaluated with Malin's Intelligence Scale for Indian Children by a psychologist. On Computerized Tomography hypodensity noted in white matter in both parietal lobes and frontal lobes.

The subject 2 also showed impairments both speech and language skills whereas the cognitive skills were better preserved. The speech skills were affected with a deterioration in phonation, articulation and fluency. The respiratory support for speech is impaired as the subject showed reduced Maximum Phonation Duration. Also, the alternative and successive movements of the articulators are affected from Diadochokinetic rate. The phonatory deficiency of the subject was evident on the analysis of s/z ratio. The Frenchay Dysarthria Assessment[3] results revealed a dysarthric nature of speech and non-speech skills and the mostly affected skills are laryngeal, lips, tongue and intelligibility. On language aspects, the subject 2 showed word finding problems, difficulties in comprehending complex sentences, repetition problems, pragmatic issues etc. On Western Aphasia Battery[6] the subject had an Aphasia Quotient 87.2 which revealed aphasic pattern of language disturbance. The subject's cognitive skills are affected mildly when assessed by a psychologist using Mini Mental State Examination[7] and the post Graduate Institute of psychiatry - Battery of Brain Dysfunction. Magnetic Resonance Imaging showed cortical swelling with grey-white matter "blurring" and cortical and subcortical T2 and FLAIR hyperintensity in left parietal and bilateral frontal lobes with subtle pial and cortical enhancement and also left tempero-parieto-occipital volume loss with subcortical gliosis and ex-vacuo dilatation of the posterior horn of the left lateral ventricle.

	Reflex			Respiration		Lips					Jaw		Soft Palate			Laryngeal				Tongue						Intelligibility		
a																											
b																											
c																											
d																											
e																											
	COUGH	SWALLOW	DRIBLE	AT REST	IN SPEECH	AT REST	SPREAD	SEAL	ALTERNATE	IN SPEECH	AT REST	IN SPEECH	FLUIDS	MAINTENANCE	IN SPEECH	TIME	PITCH	VOLUME	IN SPEECH	AT REST	PROTRUSION	ELEVATION	LATERAL	ALTERNATE	IN SPEECH	WORDS/REPETITION	SENTENCE/DESCRIPTION	CONVERSATION

Table 1: Frenchay Dysarthria Assessment[3] of the subject 1.

	Reflex			Respiration		Lips					Jaw		Soft Palate			Laryngeal				Tongue						Intelligibility		
a																											
b																											
c																											
d																											
e																											

	COUGH	SWALLOW	DRIBLE	AT REST	IN SPEECH	AT REST	SPREAD	SEAL	ALTERNATE	IN SPEECH	AT REST	IN SPEECH	FLUIDS	MAINTENANCE	IN SPEECH	TIME	PITCH	VOLUME	IN SPEECH	AT REST	PROTRUSION	ELEVATION	LATERAL	ALTERNATE	IN SPEECH	WORDS/REPETITION	SENTENCE/DESCRIPTION	CONVERSATION

Table 2: Frenchay Dysarthria Assessment[3] of the subject 2.

Western Aphasia Battery		
	Maximum Score	**Subject's Score**
Spontaneous Speech		
Information Content	10	9
Fluency	10	9
Total	20	18
Auditory Verbal Comprehension		
Yes/No Questions	60	60
Auditory Word Recognition	60	50
Sequential Commands	80	80
Total Divided by 20 For AQ	10	9.5
Repetition	10	7.7
Naming		
Object Naming	60	60
Word Fluency	20	5
Sentence Completion	10	9
Responsive Speech	10	10
Total Divided by 10 For AQ	10	8.4
Aphasia Quotient	100	87.2

Table 3: Western Aphasia Battery (Andrew Kertesz,1982) of the subject 2.

	Subject 1	**Subject 2**
Age of Onset	3.9 years	16 years
Developmental Milestones	Normal till 3.9 years and regression noticed	Normal
Fine & Gross Motor Skills	Weakness on right lower limb while walking	All the fine & gross motor skills were affected with right hemiparesis
Oral Peripheral Mechanism Examination	All structures and functions were adequate	On structure left deviation of lips and on functions all the were affected except for teeth
Vegetative Skills	Adequate	Affected blowing and IOBP
Prelinguistic Skills	Mild impairment	Adequate
Semantic Relations	Adequate	Adequate
Pragmatic Skills	Inadequate	Inadequate
Cognitive Skills	Inadequate	Adequate
Articulation	Adequate	Poor speech intelligibility and distortions were present
Fluency	Adequate	Slow rate, unfilled pauses and repetitions
Voice	Reduced loudness Reduced pitch range	Reduce pitch, reduced loudness and mild hoarseness
DDK	Adequate	Inadequate
MPD	Adequate	Inadequate
Language Skills	On LPT 8+ years	On WAB AQ: 87.2
FDA	Mildly affected laryngeal functions	All most all functions were affected
Handedness	Right-handed	Right hemiparesis
Formal Cognitive Assessment	IQ is 67 on the basis of MISIC	Mild cognitive impairment on MMSE & PGI-BBD

Table 4: Comparison of Results

SUMMARY AND CONCLUSIONS

The study summarized that the Moyamoya pattern of cerebrovascular disease developed a deterioration in the speech and language skills associated with impairments in cognitive skills for the subject 1. The language skills of the child showed 4 years delay on Linguistic Profile Test[4]. The cognitive skills of the subject were also impaired with an intelligence quotient of 67 on Malin's Intelligence Scale for Indian Children. The subject doesn't show any noticeable deficits for speech skills except for voice. Hence the results concludes that the communication impairments of the subject 1 is due to the language learning problems. In subject 2 the cognitive impairments are negligible and the remaining may be due to the naming difficulty and minimal deficits in auditory verbal comprehension. The subject's both speech and language skills were impaired. On Western Aphasia Battery[6] the Aphasia Quotient was 87.2 and on Frenchay Dysarthria Assessment[3] the subjects almost all speech and nonspeech tasks were inadequate.

So, the study concludes that Moyamoya disease with childhood onset will lead do language learning deficits accompanied by intellectual disabilities and in case of adolescent onset the communication impairments will be in the form of Aphasic and Dysarthric type of disturbances. Hence the manifestation of speech, language and communication impairments have difference on the basis of age of onset and it should be considered while selecting rehabilitation procedures.

Limitations of the Study

- There is only single subject in each age group
- The study doesn't include geriatric, adult and infantile onset

Further Recommendations

- Conduct this study in a greater number of subjects
- The study should conduct in all age levels

Declaration by Authors
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