Moyamoya Disease: a ray of hope from a psychosocial perspective

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ABSTRACT

Moyamoya disease (MMD) is a rare blood vessel disorder occurring in people from Japan and other Asian countries, but people in other countries also have been diagnosed with MMD. Impaired neuropsychological functioning is a common sequelae of MMD in children as well as adults. We report a male, diagnosed as MMD at 8 years who was referred for neuropsychological evaluation (NPE) at 12 year, revealed impaired intellectual functioning with moderate retardation on social adaptive functioning. After 2 years of follow up, post psychosocial intervention, the patient showed remarkably upward trend in his social adaptive functioning, with shift in his intellectual functioning by 21 I.Q. Points thereby brining him to mild category of mental retardation. Therefore, this rare case shows improved neuropsychological functioning, highlighting importance of interplay between nature and nurture roles. Hence, NPE is sensitive in comparing and drawing inferences for neuropsychological rehabilitation and pharmacological management for such patients.

KEY WORDS: Neuropsychology, Cognition, Childhood Stroke, Psychosocial Intervention, India

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Introduction

Moyamoya Disease (MMD) is a rare cerebrovascular disease of unknown etiology where arteries in the brain are constricted. On X-rays, these collateral vessels have the appearance of a "puff of smoke".¹ It primarily affects children but can also occur in adults. In children it tends to cause strokes or seizures whereas in adults may result in strokes or bleeding.

Individuals with MMD may have disturbed consciousness, speech deficits, sensory and cognitive impairments, involuntary movements, and vision problems, researchers also believe that MMD is the result of inherited genetic abnormalities.¹

International literature shows that MMD patients are likely to have impaired neuropsychological functioning. A study done to see the effect of MMD on neuropsychological functioning, especially executive functioning in adults, suggests that it can impair cognition but that the effect is not as severe as in paediatric cases. Memory and intellect are spared, to a large extent.²

Review from India doesn't show much literature on neuropsychological functioning. A case of an 8-year-old child is reported but the neuropsychological functioning had not been assessed.³ Assessment of neurological and neuropsychological outcome following revascularization showed that surgical procedures were effective in halting the neurological progression and resulted in neuropsychological improvement in some patients.⁴

This is the first Indian case study which analyzes the neuropsychological profile, which has shown improvement, following psychosocial intervention.

Case history

The patient is 14 year old, male, who was diagnosed with MMD with intractable epilepsy in 2008 (at 8 years of age), by the Dept of Neurology. The current frequency of the seizures is approximately 1–2 times per day. He was referred to Department of Clinical Neuropsychology, in 2011 (at 12 years of age). The

patient belonged to a middle class socioeconomic status living in a nuclear family presently doing schooling from open school.

The MRI of the brain done on 26th August 2011 revealed a case of MMD with an evidence of atrophy and gliosis involving the right temporal, occipital and to lesser extent parietal and frontal lobes with white matter hyperintensities.

MR Angiography of the Brain reveals attenuation of both supraclinoid ICA with of bilateral M1 and M2 segments. The right PCA is also attenuated.

The parents were advised about the surgery by the neurologist but the parents did not want to go ahead with the surgery. It was a decision made by the parents.

Pharmacotherapy at present: Laconext 100 mg (2 times a day), Cloba 10 mg (2 times a day), Lamosyn 100 mg (2 times a day), Oxitol 150 mg (2 times a day) and Ecospirin 150 mg.

The patient was provided services in Clinical Neuropsychology as per the ABA design (where A (Pre Neuropsychological Assessment), was done in 2011, B (Psychosocial intervention), A (Post Neuropsychological Assessment)). The tests administered on the patient in 2011 were Gesell's Drawing Test,⁵ and Vineland's Social Maturity Scale.⁶ Due to a lower mental age, testing for higher level functioning could not be carried out in the initial assessment. The psychosocial intervention included 23 sessions of 45 minutes each. It was carried out for 2 years where initially weekly sessions were held with the patient and the parents. This for carried out for 1 month. After that monthly follow ups were maintained for the remaining time. A repeat evaluation was done in 2013. Looking at a change of scores in the post assessment on social adaptive functioning higher mental functioning tests were administered i.e. Malin's Intelligence Scale for Indian Children⁷ as well as the Childhood Psychopathology Measurement Scale⁸

Results

The results (Table 1) show an improvement in the test scores after the psychosocial intervention (Table 2). The psychosocial

Table 1: The Details of Both The Assessments done in 2011 and 2013

A (Pre Neuropsychological Assessment)	S.No.		Details		2011			2013		Remarks
					Score	Category	B (Psychosocial Intervention) See Table 2	Score	Category	
	1.		Mental Age (MA) on GDT	7	Low ↓	8		Low ↓	Improved	
	2.		Performance Quotient (PQ) from GDT	58	Mild	64		Mild	Improved	
	3.		Social Quotient (SQ)	36	Moderate	57		Mild	Improved (Improvement of 21 IQ points)	
	4.		Social Age (SA)	4 Years & 3 Months	Low ↓	8 Years		Low ↓	Improved	
	5.	On Social Adaptive Functioning: (VSMS)				On Social Adaptive Functioning: (VSMS)				
		A	Self-Help General	2 Years & 7 Months ↓		9 Years & 3 Months↓		Improved		
		B	Self-Help Eating	2 Years & 5 Months↓		9 Years & 3 Months↓		Improved		
		C	Self-Help Dressing	4 Years↓		6 Years & 8 Months↓		Improved		
		D	Locomotion	4 Years & 5 Months↓		6 Years↓		Improved		
		E	Occupation	2 Years & 9 Months↓		3 Years & 7 Months↓		Improved		
		F	Communication	3 Years↓		8 Years & 8 Months↓		Improved		
		G	Self-Direction	Nil↓		Nil↓		Improved		
	h	Socialization	5 Years↓		7 Years & 8 Months↓		Improved			
6 Malin's Intelligence Scale for Indian Children (MISIC)*										
Verbal Ability										
General Knowledge							77	Borderline		
Comprehension Ability							77	Borderline		
Working Memory							69	Mild		
Attention and Concentration							69	Mild		
VIQ/Verbal Intelligence Quotient							73	Borderline		
Performance Ability										
Abstract Ability							65	Mild		
Practical Ability							62	Mild		
PQ/Performance Quotient							64	Mild		
Mean IQ							64 (SQ + VIQ + PQ /3)	Mild		
Compare above(MISIC) findings to an average of 90–110 I.Q. Points										
7 Childhood Psychopathology Measurement Scale (CPMS)										
	A	Low Intelligence with Behavioural Problems					1.74	Mild		
	B	Conduct Disorder					–0.21	Not Evident		
	C	Anxiety					–0.62	Not Evident		
	D	Depression					4.20	Severe		
	E	Psychotic Symptoms					1.45	Mild		
	F	Special Symptoms					1.60	Mild		
	G	Physical illness with emotional problems					1.23	Mild		
	H	Somatisation					0.60	Not Evident		
		Sample was compared to the Indian Norms (prepared on a sample of 200 Children) (Malhotra et al 1988)								

*Since there was improvement in comparison to earlier report on Intellectual screening scores (GDT) and improvement on Social Quotient (VSMS), therefore the patient was evaluated on higher order neuropsychological functioning i.e. Verbal and Performance tests (MISIC) which specifically evaluated ability for school going children and Childhood Psychopathology (CPMS).

Table 2: Showing Psychosocial Intervention Used on the Patient⁹

1	Behavioural Strategies	<ul style="list-style-type: none"> • Communication Enhancement Techniques
2	Family Therapy/Counselling	<ul style="list-style-type: none"> • Understanding • Comforting • Action oriented
3	Informational Strategies	<ul style="list-style-type: none"> • Informational (schools & vocational services) • Guidance regarding other facilities

intervention which followed the first neuropsychological examination included strategies for behavioural management, informational strategies, vocational guidance and family therapy, supportive counselling (Table 2).⁹ There is an upward shift in the social adaptive functioning from **moderate** mental retardation to **mild** mental retardation as per ICD 10.¹⁰

The Details of Both the Assessments done in 2011 & 2013. As can be seen in Table 1, the assessment done in 2011 only has social adaptive functioning which could be done at that time where the child was at Moderate MR level (36) but due to the improvement seen in social adaptive functioning in 2013 where the assessment revealed a marked improvement from 36 to 57 which shows an improvement of 21 IQ points and moving him to the mild MR level. Higher level cognitive functioning was done in 2013. The Mean IQ including scores on VSMS and MISIC show a mean IQ of 64 which puts the child in the category of Mild MR which depicts an overall change of 27 IQ points which is considered to be significant.

Behavioural strategies

Target symptoms were lack of communication, self-absorbed and other inappropriate behaviour. a) Parents were encouraged to gradually introduce the child to situations where he would have to interact with other children. They were told that individual attention would be of great help. The therapists also tried to establish a rapport with the child and interact with him, as a means of demonstrating to parents what needed to be done. The need for consistent handling was emphasized b) the use of activities suggested were the ones that the child enjoyed. Parents were told that such activities were to follow a set schedule, would have to be simple, accompanied by clear instructions and visual cues. Demarcating areas for specific activities was also suggested.

Family Therapy/Counselling

Use of Supportive techniques which helped parents cope with the emotional impact of diagnosis. Their disappointment and frustration on learning about the diagnosis and its implications was acknowledged. Ways of handling the stress without resorting to maladaptive coping were discussed. Positive methods such as discussing problems and their solutions among themselves, or with others, seeking help from professionals etc. were suggested.

Informational strategies

Educational and informational strategies were used to improve parent's understanding of the nature of the disease and its treatment. Parents were educated about possible prognosis,

and treatment/rehabilitation. Other aspects such as learning disability were also discussed. Information about facilities for special schooling was also provided.

Discussion

A psychosocial intervention was carried out with parental involvement in the present case. Post psychosocial intervention, at a follow up after 2 years showed improved neuropsychological functioning, which resulted in an upward trend in social quotient from and SQ of 37 to an SQ of 57. This shows an increase of 21 IQ points which is considered to be significant. Review shows an evidence of deficits in neuropsychological functioning in adults as well as paediatric cases. There is a lack of evidence regarding cognitive and clinical outcomes, due to the rarity of the disease.¹¹ MMD leads to a drastic decline in neurocognitive functions indicating a role for early surgery to save the intellectual abilities of patients.¹² In the present case, suggested Interventions such as behavioural strategies where the therapist worked with the child as a means of demonstration to parents about what was needed to be done. The importance of communication enhancement and structuring activities included choosing activities like identifying an area of house (free from distractions), interacting on one to one basis, maintaining eye contact. Family therapy to deal with emotional aspect included use of techniques like listening, comforting, action oriented techniques including planning action, behaviour modification and desensitization (overcoming anxiety). Informational strategies such as providing basic information about various aspects of the disease, and also vocational guidance were used.⁹ Increased environmental stimulation and the overall learning is believed to have led to improvement. As reported by the child's parents, the child currently has a normal interaction with friends and teachers. The child also has developed familiarity to the latest technology like computers and mobile phones which helps in cognitive stimulation.

Improvement in the scores may be attributed to his developing brain. Improvement is seen despite any reduction in the dosage of medications. There is need to assess the same by comparing the developing brain of this disease with adult MMD brain. It would probably be inappropriate to draw inferences about the child's neuropsychological ability on the basis of available literature on the said disease in adults. An evaluation on CPMS, shows the child is performing well despite the disease. It is pertinent to point that out just as in a case it was stated that MMD does not necessarily result in an incapacitated state.¹² This can also be the cause of improved social adaptive functioning along with improved intellectual functioning (both verbal and performance). Consistent with our results, a study which was done to see the effect of MMD on neuropsychological functioning, showed impairment is not as severe in paediatric cases.¹³

The critical issue is to identify brain-related dysfunction, so that these academic and behavioural interventions can be initiated in a timely manner. Early appropriate intervention can reduce the likelihood that the child will experience continued failure, which may lead to more severe emotional or behavioural difficulties. These types of interventions may include environmental modifications or introduction of compensatory strategies. The present case highlights the importance of environmental stimulation in aiding cognitive and intellectual functioning. The mother emphasised on the colouring and cursive writing skills of the child which have improved from before while the child maintains a

diary of activities. The mother mentioned the use of the white board to improve verbal and writing ability. This case of MMD shows improvement on neuropsychological assessment, post psychosocial intervention, emphasising the importance of overall characteristics of a person's environment along with genetic factors. Neuropsychological rehabilitation could go a long way in helping the child for improving the neuropsychological functioning which will improve his overall wellbeing.

Limitations

Neglected yet extremely important perspective of MMD has been described. In this progressive organic disease, can alone psychological intervention contribute to a significant improvement in the child's psychological development without taking into account the response to revascularization such as the residual cerebral blood flow and ischemia the Suzuki's stage of the disease, the effect on anticonvulsants on the outcome of intractable seizures has not been examined. During the ABA process the child was going to school and therefore to know that improvement in the social maturity is attained only due to intervention and there are no other factors (such as age education). All these factors have not been examined.

Authorship Contributions

Ashima Nehra: Carried out the initial assessment and interventions given to the patient and the family along with the compilation of the article, **Harsimarpreet Kaur:** Helped in the post assessment and compilation of the article.

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