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Case Report

Relato de Caso

Moyamoya disease: impact on the performance of oral and written language

Doença de Moyamoya: impacto no desempenho da linguagem oral e escrita

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Keywords

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Child Development
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ABSTRACT

Moyamoya disease is an unusual form of occlusive, cerebrovascular disorder that affects the arteries of the central nervous system, causing acquired language alterations and learning difficulties. The study aim was to describe the oral/written language and cognitive skills in a seven-year-and-seven-month-old girl diagnosed with Moyamoya disease. The assessment consisted of interviews with her parents and application of the following instruments: Observation of Communicative Behavior, Peabody Picture Vocabulary Test, Academic Performance Test, Profile of Phonological Awareness, Raven's Progressive Matrices Test, Special Scale, Wechsler Intelligence Scale for Children, Bender Visual Motor Gestalt Test, and Wisconsin Card Sorting Test. Two episodes of stroke in the left and right temporal-parietal and left frontal areas occurred until the age of six years and five months. Revascularization surgery and medication treatment were conducted. The audiologic and ophthalmologic assessments indicated normality. At the time of the study, the girl was attending the second grade of elementary school. She presented changes in oral and written language (syllabic-alphabetic), non-naming of all graphemes, low arithmetic and writing means, reading skill below first grade level and psycholinguistic delay, and pre-school level phonological processing skills. The psychological evaluation indicated satisfactory intellectual level; however, it also showed cognitive performance impairment in verbal and execution tasks and limitations on graphic-perceptual-motor skills and sequential logic organization. The stroke episodes influenced the performance of learning processes, affecting the analysis, integration, and interpretation of relevant visual and auditory information.

Descritores

Doença de Moyamoya
Desenvolvimento Infantil
Acidente Vascular Cerebral
Linguagem Infantil
Aprendizagem
Cognição

RESUMO

A Doença de Moyamoya (DMM) é uma forma incomum de doença cerebrovascular oclusiva que acomete artérias do sistema nervoso central, acarretando alterações adquiridas de linguagem e dificuldades na aprendizagem. O objetivo foi descrever habilidades de linguagem oral/escrita e cognitivas em menina com diagnóstico de DMM de sete anos e sete meses. A avaliação constou de entrevista com pais, Observação do Comportamento Comunicativo, Teste de Vocabulário por Imagem Peabody, Teste de Desempenho Escolar, Perfil de Habilidades Fonológicas, *Wechsler Intelligence Scale for Children*, Teste Gestáltico Visomotor Bender, *Wisconsin Card Sorting Test*. Até os seis anos e cinco meses, ocorreram dois episódios de acidente vascular encefálico (AVE) em região temporoparietal esquerda e direita e área frontal esquerda. Realizou cirurgia para revascularização e tratamento medicamentoso. Avaliações audiológicas e oftalmológicas indicaram normalidade. Cursa o segundo ano do ensino fundamental. Apresentou alterações na linguagem oral e escrita (silábico-alfabética); não nomeação de todos os grafemas; escrita e aritmética média-inferior e leitura inferior à primeira série; habilidades do processamento fonológico em nível pré-escolar. A avaliação psicológica indicou nível intelectual satisfatório, porém prejuízo no desempenho cognitivo em tarefas verbais e de execução, limitações nas competências gráfico-percepto-motoras e na organização sequencial lógica. Os episódios de AVEs trouxeram interferências no desempenho dos processos de aprendizagem pelas especificidades das áreas afetadas, interferindo na análise, integração e interpretação de informações auditivas e visuais relevantes para os processos de aprendizagem.

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INTRODUCTION

Moyamoya disease (MMD) is an unusual form of chronic, occlusive, cerebrovascular disorder that affects the arteries of the central nervous system causing thrombosis, ischemia, and intraparenchymal hemorrhage. The blockages result in small-caliber, inefficient vascular neoformation[1].

MMD is a rare disorder that presents etiological aspects and insufficiently known pathophysiological mechanisms[2,3]. It is mostly prevalent in Japan, but it has been increasingly diagnosed worldwide and is a major cause of cerebrovascular accident during childhood. This condition accounts for approximately 6% of the stroke cases in children in Western countries[3]. Reports on its occurrence show a female-to-male ratio of 2:1[3,4].

The term Moyamoya is of Japanese origin, meaning "something hazy" such as a "puff of smoke". It refers to the appearance of findings observed in angiography, which is the criterion used for confirmation of its diagnosis[3,4].

The variable incidence rates between different ethnic groups and the familial recurrence support the role played by genetic factors on MMD[2]. Countless candidate genetic factors for MMD have been identified in several chromosomal regions (e.g., 3p24.2p26, 6q25, 8q23, 12p12, and 17q25)[2] in the past years, which justifies its phenotypic variability.

In this disease, there is bilateral occlusion of the internal carotid arteries, and because this occlusion is slow and gradual, multiple anastomoses are formed between the internal and external carotid arteries, which are composed of the anterior and posterior lateral choroidal arteries, the basilar artery, and the meningeal arteries[3-5].

The etiology of MMD is still unclear[2,6] and its incidence is higher in the first decade of life (50% in pre-school age); nevertheless, it can also occur in the second or third decades of life. The natural history of MMD entails significant morbidity and mortality[6]. Pediatric patients can present silent attacks, transient or permanent ischemic attacks, and cerebral infarctions in the territory of the internal carotid artery, particularly in the frontal lobe[4]. Unilateral lesions may progress to bilateral lesions[6]. Children may experience headaches, convulsive seizures, involuntary movements, and cognitive alterations[1,6].

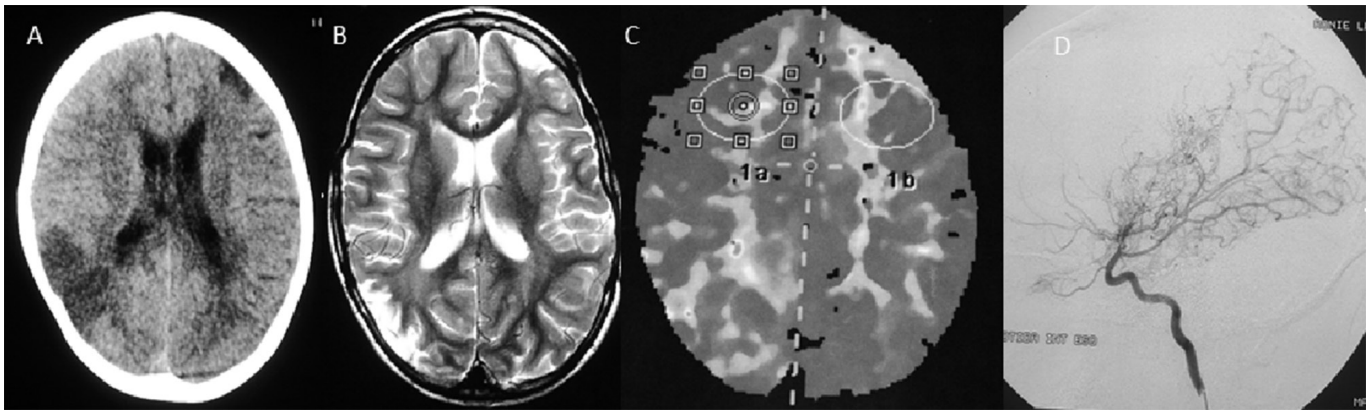
The clinical manifestations of brain injuries are variable. Depending on the areas affected, they can cause dysarthria, aphasia, hemiparesis and/or convulsive seizures[3-5]. Studies have reported[1-7] neurocognitive and psycholinguistic changes that result in losses for learning and quality of life. Revascularization surgery can significantly reduce the subsequent risks to cerebral ischemic events[4,6] and protect the intellectual abilities of children with MMD[7].

The objective of the present study was to describe the oral/written language and cognitive skills in a seven-year-and-seven-month-old girl diagnosed with Moyamoya disease.

CASE REPORT

The patient's family signed an Informed Consent Form according to resolution no. 466/12 of the National Research Ethics Committee.

The study participant was a seven-year-and-seven-month-old female child, daughter of non-consanguineous parents, diagnosed with Moyamoya disease. The girl presented psychomotor development in accordance with regulatory criteria. She had several episodes of chronic ear infection during the first year of life. Her parents reported that, by the age of four, she began to complain of tinnitus and headaches. Audiologic and ophthalmologic assessment indicated normality. Around the age of five, she manifested involuntary tightness in the lips and numbness in the hands. At that time, under neurological monitoring, she performed imaging exams that showed signs of ischemic cerebrovascular accident (ICVA) with involvement of the left temporal-parietal area. At the age of six years and five months, a second ICVA occurred involving the right temporal-parietal and left frontal areas. The patient was submitted to revascularization surgery and medication treatment. Figure 1 shows the data obtained in the following image exams conducted after surgery: Computed Tomography (CT), Magnetic Resonance Imaging (MRI) with



Caption: (A) Computed Tomography (CT): showing hypodense area in both brain hemispheres, in the right and left parietal-occipital regions, and in the left frontal area, compatible with ischemic cerebrovascular accident (ICVA); (B) T2-Magnetic Resonance Imaging (MRI): showing old cortical infarction in the territory of the left middle cerebral and right posterior cerebral arteries; (C) Magnetic Resonance Angiography (MRA): depicting a classic example of Moyamoya disease; (D) Magnetic Resonance Angiography (MRA): showing compatibility with MMD
Figure 1. Results of the Computed Tomography (CT), Magnetic Resonance Imaging (MRI), and Magnetic Resonance Angiography (MRA)

perfusion study, and Magnetic Resonance Angiography (MRA) of arteries and intracranial veins.

At the age of seven years and seven months, she was attending the second grade of elementary school at a private institution. Although she was receiving pedagogical support and was in speech-language pathology and psychology therapy, her academic achievement was below average. There were no complaints about behavior.

The speech-language and neuropsychological assessments consisted of interviews with parents and educators and the application of specific instruments of assessment according to the child's age. The following protocol was used: Observation of Communicative Behavior (OCB)[8]; Peabody Picture Vocabulary Test (PPVT)[9]; Academic Performance Test (APT)[10], Profile of Phonological Awareness (Pro-PA)[11], Raven's Progressive Matrices Test - Special Scale (RAVEN)[12], Wechsler Intelligence Scale for Children - Third Edition (WISC-III)[13], Bender Visual Motor Gestalt Test (BG)[14], and Wisconsin Card Sorting Test (WCST)[15].

Table 1 shows the results of the speech-language pathology assessment. The participant presented difficulty in naming isolated graphemes.

The neuropsychological assessment (Table 2) indicated intellectual level satisfactory for the age; however, it also showed cognitive performance impairment in verbal and execution tasks, with limitations on the management of attentional focus, graphic-perceptual-motor skills, sequential logic organization, and expressive language; these impairments caused important losses in the acquisition and development of academic skills. The participant showed collaborative behavior during the administration of the assessments.

DISCUSSION

Changes in information processing skills involving working memory, phonological awareness, and access to mental lexicon, as well as cognitive delay affect the learning process[5]. The performances obtained suggest risk of significant losses in academic skills for school progress.

MMD sequelae caused by ICVAs are variable and dependent on the specificity of the brain areas affected[1,3]. After brain injury, the neurological deficits, which are invariably permanent, require frequent therapeutic monitoring, mainly to cope with changes resulting from the injury. In the present case, the affected areas are extremely important for the functioning of language and learning. A previous study reported that cognitive functions and language performance are crucial in the processes of learning and social integration, especially during school age[5].

Table 1. Skills, performance, corresponding age, and ratings obtained using the instruments of clinical assessment

Instruments	Performance	
OCB	Communicative intention; good interaction; initiates and maintains communicative shifts; produces words and simple sentences without syntax changes. Presents the language functions of informing, protesting, requesting, and imitating. Understands simple instructions. Reduced attention span. Difficulties to narrate facts and perform logical and temporal sequences. Does not present productive phonological processes.	
PPVT	Higher-low rating.	
APT	Skills	Rating
	Reading	Mean below 1st grade
	Writing	Mean below 1st grade
	Arithmetic	Mean below 1st grade
Pro-PA	Pre-school level phonological processing skills	

Caption: OCB = Observation of Communicative Behavior; PPVT = Peabody Picture Vocabulary Test; APT = Academic Performance Test; Pro-PA = Profile of Phonological Awareness

Table 2. Performance, intelligence quotient, and classification obtained using the instruments of neuropsychological assessment

Instruments	Performance
Mental Level: RAVEN – Raven's Progressive Matrices Test - Special Scale	Mental level compatible with chronological age (Rating: III - Medium). Identifies similarities; establishes analogies and relationships of cause and effect.
Learning Level: WISC-III – Wechsler Intelligence Scale for Children - Third Edition	Cognitive skills for the verbal (VIQ = 88) and execution (EIQ = 85) activities presented medium-lower levels, with medium-inferior overall performance classification for the chronological age. Satisfactory performances were identified in assessments related to visual-perceptual discrimination with respect to understanding and social adequacy. Presented median level in assessments that required processing speed; however, the factor indices related to verbal understanding and perceptual and attentional organization showed below-average profiles for development. These data represent impact on the fundamental skills for formal learning.
Perceptual-Motor Level: BG – Bender Visual Motor Gestalt Test (Santucci/Koppitz)	Highly impaired graphic-perceptual-motor performance, showing below age and school levels (Md = 2a) and limited resources in the integration of lines and construction of angles, which represent losses in formal drawing and writing. 57% indicative signs of neurological immaturity for handwriting.
Execution Level: WCST – Wisconsin Card Sorting Test	The execution reasoning, used in the resolution of situations of conflict, choice, and adaptation to the context (absent completed trials), was below average, representing lack of knowledge in managing the focus of attention and difficulty in memorizing (working memory) for the related activities.

The parietal lobe was one of the areas affected by the ICVA in the present case. This area is divided into two functional regions: one region is involved in sensation and perception, whereas the other is responsible for integrating sensory input, primarily with the visual system. The first function integrates sensory information to form a single perception (cognition). The second function builds a system of spatial coordinates to represent the world around us. Individuals with damage to the parietal lobes may present slight to deep deficits, related to abnormalities in body image and spatial relations[7]. Damage to the left parietal lobe can cause confusion between left and right, writing (dysgraphia) and mathematical thinking (dyscalculia) difficulties, language disorders (aphasia), and inability to perceive objects normally (dysgnosia). Damage to the right parietal lobe can result in disabilities that affect the individual's care with the body, such as dressing and bathing, and compromise independence in activities of daily living (ADL). The clinical history presents these findings, that is, the child showed difficulty in accomplishing ADLs independently.

Bilateral lesions interfere with the integration of visual attention and motor efficiency[6], which is characterized by manifestations related to dyspraxia, losses in formal drawing and writing activities, construction of angles, and integration of lines. The child in question presented particular difficulty with these skills.

The temporal lobe was another area affected by the ICVA. The temporal lobes are related to memory, hearing, processing and perception of sound information, ability to understand language, and higher order visual processing. The psycholinguistic changes observed are reported in the literature[7]. Lee et al.[5] found that lesions in the left brain resulted in reduced verbal skills, whereas lesions in the right brain resulted in lower scores on neurocognitive functions such as memory, visual skills, and spatial relations.

Lesions in the frontal lobe may be related to difficulty in the maintenance of sustained attention, memory, and planning, as well as mood disorders. The latter was not observed in the case studied. Pediatric MMD patients can benefit from surgical revascularization, especially those with progressive neurological symptoms or evidence of inadequate blood flow and/or collateral circulation, as well as those without surgery contraindication[4-5]. The child in this study was submitted to this surgical procedure.

Considering the developmental plasticity in the infant brain, it is expected that extrinsic influences, such as participation in speech-language pathology and psychology intervention processes, coupled with intrinsic influences can alter the cortical organization and regenerate damaged connections, improving the performance of children in compromised skills.

Although MMD does not present high incidence, it is a major cause of ICVA in children and, therefore, needs to be better understood, particularly regarding the development of these children, due to the impact caused on brain functioning, which affects the lives of these individuals[6].

MMD is a complex and heterogeneous disorder, with varying phenotypes and genotypes, that has rapidly expanded worldwide[1,2]. Thus there is need for more comprehensive knowledge on the disease in order to develop more specific and efficient risk stratification systems, including deep clinical and radiological phenotyping and identification of biomarkers that could improve the prognosis capacity to develop tailored intervention procedures adapted to individual necessities[2].

The management of treatment of MMD sequelae requires a multidisciplinary approach, especially in childhood, to minimize the deleterious effects caused by brain insults, which will bring consequences for the school and social life of these children and their families.

FINAL COMMENTS

Moyamoya disease influences the language processes and neurocognitive functioning, with losses in the learning process owing to the specificities of the affected areas, as well as in the skills of analysis, integration, and interpretation of essential visual and auditory information.

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Author contributions

DACL was responsible for the study design, collection and analysis of data, and writing of the manuscript; CCR was responsible for the speech-language pathology assessment, literature review, and revision of the manuscript; PMDPF was responsible for the neurological assessment; MLMT was responsible for the psychological assessment and writing of the manuscript.