J. Phys. Ther. Sci.
25: 1647–1649, 2013

Case Study

Long-term Outcome of Motor Function in a Child with Moyamoya Disease: A Case Report

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Abstract. [Purpose] This observational study provides a retrospective description of changes in motor function of a 10 year old child who suffered from motor weakness caused by Moyamoya disease (MMD) over an approximately 3 year follow-up observation period. [Methods] The child was diagnosed as MMD due to multifocal encephalomalacia in both frontal and parietal cortices. After the ischemic attack, the child received physical therapy the based on stroke rehabilitation, including muscle strengthening exercises, training of functional activity/ADL, and neurodevelopmental treatment. [Results] The child's MRI showed areas of ischemic infarction in both the frontal and parietal lobes. Steno-occlusive findings for both the anterior cerebral artery and middle cerebral artery were observed on cerebral angiography. Regarding changes of motor function during the three-year follow-up, significant improvements, in the Motricity index, Modified Brunnstrom Classification, manual function test, and functional ambulatory category were observed. [Conclusion] The basic motor function and functional abilities of the child showed improvement with conservative treatment over approximately three years. The functional motor ability of children with MMD may be similar to the recovery progression of pediatric stroke patients, if there is no re-occurrence of ischemia.

Key words: Moyamoya disease, Cerebral artery occlusion, Motor function

(This article was submitted Jun. 11, 2013, and was accepted Jul. 3, 2013)

INTRODUCTION

Moyamoya disease (MMD) is a chronic occlusive cerebral arteriopathy characterized by progressive steno-occlusion of the arteries of the Willis circle[1–5)]. The progressive occlusion generally begins in the proximal portion of the internal carotid artery, gradually spreading to numerous collateral vessels. Appearance of occlusion in the collateral vessels in angiographic studies is described as a "puff of smoke". The prevalence of MMD due to unknown causes has been frequently reported in Japan and East Asia, and idiopathic and genetic factors are frequently related[6, 7)].

MMD is also one of the most common causes of neurological disability, such as cognitive and motor dysfunctions, arising from cerebrovascular disease in childhood[8)]. According to prior studies[1, 9, 10)], children predominantly show high incidence of ischemic pathogenesis due to inappropriate perfusion, whereas in adults, cases of hemorrhage related to the fragility of neovessels are common. In addition, the incidence of MMD shows two age-related peak one at approximately 5 years old, and the other in the mid-40s[1, 3, 4)]. Despite this, MMD has not received significant attention in the field of clinical physical therapy due to it being regarded as an uncommon brain disease, as its incidence rate is lower than that of cerebral palsy. This single-case study reports the changes in motor function and functional ability of a child with Moyamoya disease, over a long-term clinical observation of three years.

SUBJECTS AND METHODS

Our subject was a 10 year-old male child, who was diagnosed as having Moyamoya disease by a pediatric neurologist, and transferred to the physical therapy department at a local university hospital. At the first onset on August 20, 2010, the child complained of a burst-onset headache in the frontal area, along with vomiting, and tremor of the right arm. The result of brain MR imaging showed areas of infarction in the bilateral frontal and parietal lobes. In addition, the result of the electroencephalogram, for confirmation of seizure occurrence, showed non-specific finding. The child was finally diagnosed as having Moyamoya disease, caused by occlusion of both the anterior and middle cerebral arteries according to angiography findings (Fig. 1). The boy had no past medical history of diabetes, hypertension, tuberculosis, or other surgical procedures. Interview of his parents also indicated no family history of Moyamoya disease. According to the child's medical record, he had received conservative therapy using anticoagulants and

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thrombolytic agents, similar to the management of ischemic stroke without neurosurgical procedure. The child's neurological pathology caused severe motor weakness and cognitive dysfunction. When his medical condition was stable, the child was transferred to the physical therapy department for rehabilitation treatment. To date, he has been involved in a physical therapy program, including strengthening exercises, training of functional activity/ADL, and neurodevelopmental treatment using the Bobath concept. The parents of the child gave their written informed consent to participation in this case report. The study protocol was approved by the ethics committee of a university hospital.

Figure 1 shows T2-weighted MR images (A) and cerebral angiography (B). T2-weighted MR images show ischemic infarctions in both the frontal and parietal lobes. Cephalocaudal (B-1) and anterioposterior (B-2) views of cerebral angiography show that the anterior and middle cerebral arteries were occluded, resulting in the development of abnormal collateral vessels.

The motricity index (MI) and Modified Brunnstrom Classification (MBC) were used to assess the motor capacity and muscle tone of both paralyzed limbs. The manual function test (MFT) was adopted for evaluation of upper limb movement and hand manipulative function in each segmental joint of the affected limb. In addition, the child's ambulatory ability was tested using the functional ambulatory category (FAC). Owing to their good reliability and validity[11–13], these clinical assessments are often used in clinical rehabilitation.

RESULTS

At the initial evaluation at the end of August, in 2010, MI of the upper and lower limbs (the right/the left) was 19/54 and 19/19, respectively. MBC was 1 on the right side and 5 on the left side. MFT was 4 on the right side and 9 on the left side. FAC was 0, because the child could not walk independently. At the second evaluation at the end of May, in 2011, MI of the upper and lower limbs (right/left) was 55/64 and 33/33, respectively. MBC scores of the right and left side were 4 and 5, respectively. MFT scores were 10 on the right side and 25 on the left side. FAC was assessed as grade 1. At the third evaluation at the end of May, in 2012, MI of the upper and lower limbs (right/left) was 60/70 and 52/52, respectively. MBC scores of the right and left sides were 4 and 5, respectively. MFT scores were 14 on the right side and 26 on the left side. FAC was assessed as grade 2. At the most recent evaluation, made in the middle of May, 2013, the patient showed the same motor function as that observed at the third evaluation.

DISCUSSION

This study retrospectively describes a single case of a 10 year old child with Moyamoya disease, who complained of motor weakness caused by cerebral infarction of both the frontal and parietal lobes, over a follow-up observation of approximately three years. The child's MRI/Angiography showed revascularization of collateral vessels in both cerebral hemispheres, which led to cell death in the corresponding brain areas. The child has received conservative medical treatment and physical therapy for three years since onset, which resulted in noticeable motor improvement in terms of MI, MBC, MFT, and FAC. These changes indicate that the child has recovered basic motor function and functional motor skills.

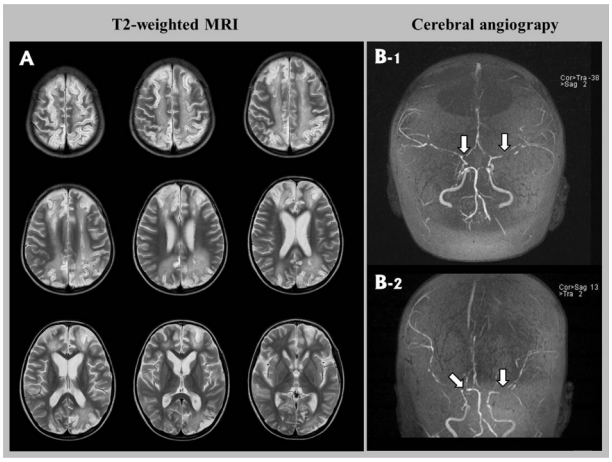

Fig. 1. Brain images of a child with Moyamoya disease

This recovery process was not dissimilar to that of other children with stroke[14–17]. We assumed that this is because there was no recurrence of ischemic or hemorrhagic attack, and the presence of neurosurgical revascularization from the onset. Our case is similar to other case studies[5, 18–20], which have reported that children with MMD show good functional and social outcomes, as the result of long-term follow-up. According to the study of Weinberg et al.[5], functional and neurocognitive outcomes differed between children and adults with MMD. They reported that the cognitive function most affected in children with MMD was intelligence, whereas it is executive function in adult cases. In addition, Phi et al.[18] reported that patients with pediatric MMD who reached adulthood showed good social adaptation. Therefore, it is likely that patients with MMD have a high possibility of achieving good functional outcome through a spontaneous healing process and rehabilitation treatment.

Cases of MMD in children are not rare in the field of clinical pediatric physical therapy in East Asia, compared to other regions. Therefore, an understanding of the clinical features and the recovery process of children with MMD would help physical therapists in the East Asian region to perform appropriate evaluation and intervention. To the best of our knowledge, few long-term follow-up reports of children with MMD have been published. We hope that our present findings contribute to clinical decision making for children with MMD. However, this single observational study is limited by difficulty with generalization of the results. In future study, randomized clinical trial with a large sample size will be required of follow-up of pediatric patients with MMD.

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