Clinical Features and Long-Term Outcome in Adult Stroke Patient due to Moyamoya Disease: A Single Subject Study

Yong Hyun Kwon¹, Chung Sun Kim²

¹Department of Physical Therapy, Yeungnam College of Science and Technology; ²Department of Physical Therapy, College of Rehabilitation Science, Daegu University

This case report described a single case of adult stroke patient due to Moyamoya disease through long-term follow-up observation, which included his demographics, brain images, and change of motor function and functional activities. The subject was the 54-year-old male diagnosed with left hemiparesis from a stroke due to multifocal encephalomalacia in both hemispheres. At the time of the stroke attack, he took brain surgery intervention including external ventricular drain. Physical and occupational therapy for stroke rehabilitation were admitted including muscle strengthening exercises, functional activity/ADL training, neurofacilitative techniques with bobath or proprioceptive neuromuscular facilitation concepts, and compensatory strategy. Patient's MRI showed that right frontal lobe, right peri-ventricular area, left parietal, and left occipital lobes were damaged, and MRA showed that abnormal collateral vessel was richly developed in both hemispheres by occlusion of proximal internal carotid arteries in both sides. His motor strength was improved from poor to good grade in all of upper and lower limb motions, that MBC was improved from stage 1 to stage 5. In FAC and barthel index, at the initial evaluation, he could not perform any functional movement, but his FAC and barthel index were on 3 and 14 points at present, respectively. During long-term follow-up for approximately 4 years, the subject's functional motor ability was improved, as similar with recovery progression of usual stroke patient. We believe that this single case report will provide clinical information and concern regarding Moyamoya disease with physical therapist, in terms of such as epidemiology, pathogenesis, diagnostic procedures, clinical features, recovery process, and prognosis.

Keywords: Moyamoya disease, Cerebral artery occlusion, Collateral vessel, Long-term outcome, Motor function

I. Introduction

Stroke is a major disease to cause cognitive and sensoriomotor disability, which is necessary for clinical rehabilitation service including physical and occupational therapy. Clinical features and symptoms of stroke is tremendous diverse depending on etiology, lesion location/volume size, pathophysiologic progress, and so forth. In addition, uncommon cases of stroke by specific or unknown causes have been variously observed in clinical setting, in terms of Behcet’s disease, Moyamoya disease, Kawasaki syndrome, epidermal nevus syndrome, Senden’s syndrome, Marfan’s syndrome, Sturje-Weber syndrome, etc. Of these uncommon types of stroke, stroke patient with Moyamoya disease is occasionally referred to physical therapist. Moyamoya disease is a chronic cerebroarteriopathy characterized by progressive bilateral obliteration of the major arteries in child and young adult. The occlusions usually originate with the internal carotid artery, and spread out the main cerebral arteries such anterior, middle, and posterior cerebral artery. As the stenosis of these arteries is gradually developed, it is replaced by a meshwork...
of the small collateral capillaries at the base of brain\(^8\,^9\). The named Moyamoya is originated from that appearance of these vessels on the angiography resembles a “puff of smoke”\(^8\,^9\). The first study reported the case of male adult who is 29 years old with bilateral dysplasia of the internal carotid arteries by Takeuchi and Shimizu\(^11\). The etiological course of Moyamoya disease still remained unknown. However, many factors have been suggested, including predisposition, systemic infection, focal factors, and platelet abnormalities\(^12\). The prevalence of Moyamoya is higher in Japanese than in Korean and Chinese, and European was nearly one–tenth of that reported in Japan\(^8\,^10\). Presently, the most effective therapeutic protocol for stroke with Moyamoya disease is established. However, surgical revascularization is recommended for effectiveness of symptom reduce and cerebral infarction prevention. Recent study reported that long–term clinical outcome of surgical treatment is relatively excellent\(^13\).

Moyamoya is not the unfamiliar disease for physical therapist, and relatively higher incidence in our country\(^14\,^15\). Im et al.\(^15\) reported that the prevalence rate was 5.2 per 100,000 persons, based on 2004 data from the National Health Insurance Corporation in Korea. However, there is little information regarding clinical features and rehabilitative outcome of Moyamoya disease, although long–term clinical outcome after surgical revascularization has been reported by many previous studies\(^10\,^19\). Here we described single case of adult stroke patient with Moyamoya disease through long–term follow–up observation for approximately 4 years. This case study reported his demographics, brain images, and change of clinical characteristics, including motor function and functional activities.

\section*{II. Case description}

\subsection*{1. History and systems review}

We searched stroke patient with multiple brain damages diagnosed as Moyamoya disease, of stroke patients who has been long–termly followed up to physical therapy room of university hospital at present, since stroke onset. As result of review of their medical records, we selected the 54–year–old male diagnosed with left hemiparesis from a stroke due to multifocal encephalomalacia in both hemispheres. Before stroke onset, he carried on a private business and was physically active, regularly engaging in tennis and golf. At the time of the stroke attack in the end of January in 2009, he complained severe headache and mental decline after climbing mountain, and was transferred to an emergency room of university hospital. By intra–cerebral hemorrhage in computerized tomography and magnetic resonance image of the brain, he took brain surgery intervention including external ventricular drain, By arisen from hydrocephalus after neurosurgery operation, ventriculoperitoneal shunt operation was performed. As medical status was getting stable, he took general physical therapy interventions including passive exercise and bed side activity program to his caregiver, for prevention of limited range of motion and predictable abnormal posture. Until present, he was hospitalized five times at the department of neurology or rehabilitation medicine, whenever medical problems were occurred. However, he did not take any neurosurgical operation. Physical therapy and occupational therapy were involved with the rehabilitation program, generally at frequency of five times per week during hospitalization or two times per week during out–patient treatment. Conventional intervention for stroke rehabilitation were admitted including muscle strengthening exercises, functional activity/ADL training, neurofacilitative techniques with bobath or proprioceptive neuromuscular facilitation concepts, and compensatory strategy. His pas medical history was free from hypertension, diabetes mellitus, and cardiac problem.

In acute stage, mini–mental status examination could be not assessed due to his poor cooperation. In the end of 2009, as his cognitive function was improved, he could carried on one step command, but other items could be not assessed due to his poor attention and demotivation. From the middle of 2011 to the present, in mini–mental status examination, his cognitive abilities were feasible to perform five orientation items and two memory recognition, two step commands, and one repetition of language.

\subsection*{2. Brain imaging}

T2–weighted image of magnetic resonance image (T2–MRI)
and magnetic resonance angiogram (MRA) with other brain imaging techniques were conducted whenever it was needed depending on clinical decision of patient’s doctor in charge. In this study, we selected the T2–MRI and MRA images that best expressed patient’s neurological status. Figure 1 indicated T2–weighed MRI (A), anteroposterior view image (B) and lateral view image (C) of MRA, following 9 months after onset. Patient’s MRI showed that right frontal lobe and peri-ventricular area were damaged, and that left parietal and occipital lobes were infarcted (A). Anterioposterior view of MRA (B) showed that abnormal collateral vessel was richly developed in both hemispheres (a), and that proximal internal carotid arteries in both of sides were narrowed, which may lead the anterior cerebral artery and middle cerebral artery to be disappeared. In addition, lateral view of MRA (C) indicated abnormal collateral vessel (a) and occlusion of proximal internal carotid artery (b).

3. Basic motor function

Motor strengthening in the left hemiparetic upper and lower limb was tested with manual muscle test (MMT). The Modified Brunnstrom Classification (MBC) was used to assess the muscle tone of hemiparetic upper arm and its changes. MBC was classified into six stages as follows: stage 1 indicated that the upper limb was completely paralyzed, stage 2 had no active motion but reflexive movement, stage 3 performed his joint movement up to 50 percentile of normal range in at least one joint, stage 4 had over 50 percentile or more of movement in one part of the arm, stage 5 performed nearly full range of motion but no completely, and stage 6 was fairly closed to normal motion. MBC has generally been used in clinical rehabilitation, on account of its good reliability and validity. MMT and MBC were regularly tested when he had been hospitalized or ambulatory visits.

In acute stage, it was impossible that MMT was assessed accurately, because of his poor cognitive function. However, based on his voluntary movement, it was likely that his left upper and lower limbs be poor grade in all of limb motions, in terms of shoulder abduction, elbow flexion, finger extension, hip flexion, knee extension, and ankle dorsiflexion, MBC was at stage 3. His motor strength was improved to fair grade in all of upper and lower limb motions following 10 months after stroke attack. MBC was improved to stage 4. Following two and half years since onset, MMT was changed to good grade in all of limb movements and MBC was improved to stage 5. These basic motor abilities have been lasted until now, with fluctuation of motor function according to his general condition or presence of medical complication.

4. Functional motor ability

For evaluation of motor performance in each segmental joint of affected limb, manual function test (MFT) was used, which included 32 testing items to assess the upper limb movement and hand manipulative function, using eight measurement tools. In scoring, 1 point is graded when each test item is successfully performed, and scored 0 when the test is not completed. Finally, each of test items is summed up. The maximum score of MFT is 32 points. At the initial evaluation of onset, MFT could not be tested due to his poor mental status. After that, in June of 2011, his MFT was 13, and from July of 2011 to present, he has been 17.

For test of ambulatory ability in stroke patient, Functional ambulatory category (FAC) was used, FAC consists of six grades as following, when stroke patient walk on a 15 meter–even floor: 0=no ambulatory, 1=requirement of consistent assistance by a person, 2=requirement of intermittent assistance by a person, 3=requirement of verbal instruction, 4=requirement of supervision or physical assistance.
on stairs and uneven surfaces, and 5=walk independently. At the point of the initial evaluation, he did not have physical function, worthy of performing of FAC. At the evaluation in December of 2009, his FAC was 1, and he has been still on 3 points on FAC from November of 2011 to present.

The barthel index was test to assess functional activities of daily life in our stroke patient. The 10 testing items include feed, move and return from wheelchair to bed, groom, transfer to and from a toilet, bath, walk on level surface, go up and down stairs, dress, and continence of bowels and bladder. Higher score is graded, as patient performs the activity more independently. In December of 2009, his barthel index was 14, and developed to 16 from July of 2011 to present.

The reliability and validity of all tests in terms of the MFT, FAC, and barthel index are well established.21-23

III . Discussion

In this study, we described single case of adult stroke with Moyamoya disease for approximately 4 years since stroke onset, who is 54-year-old male patient with mainly left hemiparesis by multiple infarction in both hemispheres. T2-weighted MRI and MRA of brain were tested for diagnosis of stroke due to Moyamoya disease and for examination of its progress, in addition, basic motor function and functional activity were assessed: the former included MMT for motor strength and MBC for normalized synergic pattern, and the latter consisted of MFT for hand motor function, FAC for walking ability, and barthel index for ADL function. His MRA indicated that proximal internal carotid artery was occlusive, and that it led to revascularization of collateral vessels in both cerebral hemispheres. In addition, these pathologic processes caused to ischemic cell death of right frontal lobe, right peri-ventricular area, left parietal lobe, and left occipital lobe. For long-term follow-up with conventional neurological and rehabilitative treatment, his functional motor ability was improved, as not different from recovery progression of usual stroke patient. As for these functional improvements, it would be attributed that additory ischemic attack or its related neurosurgical intervention was not occurred after onset. These findings were accordance with several previous studies12, 24-26, reporting long-term follow-up experiment of motor function and social adaptation in Moyamoya disease, Moore et al12, introduced four female stroke patients due to Moyamoya in an acute rehabilitation setting, and compared between at the initial and discharge scores of Functional Independence Measure. As results, the patients with Moyamoya had a higher Functional Independence Measure score, Longer length of stay, and slower rate of progress than other stroke patients, Mukawa et al26, conducted survey study for long-term follow-up of 208 juvenile patients who had surgical revascularization due to Moyamoya disease. Their long-term survey results indicated that surgically treated juvenile patients with Moyamoya maintain good ADL outcome, although new cerebrovascular events occurred significantly over 10 years after the initial surgical intervention. In addition, Phi et al27, reported that patients with Moyamoya showed favorable social adaptation, as results for details of social adaptation and satisfaction in adults who took surgical revascularization during childhood. Moyamoya disease is a chronic cerebrovascular pathologic condition characterized by progressive stenosis of the major brain arteries. The fundamental causes are unknown yet. Genetic background exist the high prevalence among East Asian people, especially in Japan, Korea, and China8, 28. However, low prevalence is found among Caucasians28. In general, Moyamoya is often occurred in child under 10 years or 30's adult, and rather higher incidence rate in female. Main clinical symptoms include transient ischemic attack, headache, seizure convulsion, disturbance of consciousness, etc8, 25. According to clinical features and long-term outcome in adult patients with Moyamoya disease by retrospective longitudinal study of Bao et al34, the median age for the onset of disease was 36.8 (18-56) years, and familial occurrence was 2.3%. In addition, the incidence of postoperative ischemic or hemorrhage was 5.9%, and involvement of posterior cerebral artery and transient ischemic attack at older age was identified as predictors of adverse post-operation.

Stroke patient with Moyamoya is not rather rare case of neurological disorder, compared to other uncommon causes of stroke, but often referred to rehabilitation service.
Therefore, clinical information regarding Moyamoya disease will be important for physical therapist, such as epidemiology, pathogenesis, diagnostic procedures, clinical features, recovery process, and prognosis. However, in the field of physical therapy, there was little report on clinical characteristics regarding Moyamoya disease in domestic or overseas scientific journals. We believe that this current study will provide clinical information and concern regarding Moyamoya disease with physical therapist. However, our limitation is that it is difficulty for generalization of final long-term outcome of motor function, because of just a single case of study regarding stroke with Moyamoya. However, long-term follow-up study of stroke patient with Moyamoya is clinically is not somewhat common, and recruitment of the type of patient is not easy. In further study, we expect that original work with large sample size or a series of case for pediatric patients will be conducted, considering effectiveness of physical therapy intervention and therapeutic guideline depending on physical functional ability of patient.

References

