Moyamoya Disease in an Extremely Old Patient

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Abstract-

Purpose: The oldest patients diagnosed with moyamoya disease (MMD) in the USA may have been as old as 85+ years, and 68 years in Taiwan; therefore, MMD is generally thought not to occur in extremely old patients in Taiwan. Herein, we report this case to revise the common thinking.

Case Report: An 82-year-old woman had suffered twice from a right cerebral infarction. A digital subtraction angiogram demonstrated abundant collateral arterial networks from the bilateral proximal middle cerebral arteries despite the absence of arteriosclerotic stenosis of the bilateral extracranial internal carotid arteries. Aspirin has effectively controlled her symptom since diagnosis and, as a result, a director indirect revascularization procedure is unnecessary.

Conclusion: MMD does occur in extremely old Taiwanese, as old as over 80 years old, and thus should be a differential diagnosis of cerebral infarction in an extremely old patient. Digital subtraction angiography is the gold standard for the diagnosis.

Key Words: moyamoya disease, cerebral infarction, hemiplegia, rete mirabile, extremely old person

INTRODUCTION

Moyamoya disease (MMD) is a cerebrovascular disease involving progressive occlusion that is characterized by stenosis or occlusion at the terminus of the internal carotid artery (ICA) or the proximal areas of the anterior/middle cerebral arteries with concurrent formation of pathognomonic collateral arterial networks (retemirabile) near the occlusive or stenotic lesions(1,2). Definite MMD is diagnosed when there are patients with bilateral lesions and probable MMD is diagnosed in unilateral lesions. The typical clinical features are cerebral infarction, recurrent transient ischemic attacks, and convulsions together with involuntary movements and headaches in most children and young patients; furthermore, there is intracranial bleeding in about half of adult patients despite the absence of any intracranial aneurysm(3). The incidence is 0.086 per 100,000 in USA(4), and 0.01 to 0.048 per 100,000 in Taiwan(5). Although MMD often attack children less than ten years old(2), the oldest patient may have been 85+ years.
old in USA \(^{(4)}\), and 68 years old in Taiwan \(^{(5-8)}\). Therefore, MMD would seem to be rare in older patients and is generally thought not to occur in extremely old patients in Taiwan. Herein we report an 82-year-old woman diagnosed with MMD as a rare curiosity.

**CASE REPORT**

An 82-year-old woman did not suffer from hypertension, diabetes mellitus or other systemic disease; however, she had suffered a right cerebral infarction one year earlier and had been bothered with left hemiplegia since then. She was sent to our hospital because the left hemiplegia had been deteriorating for two weeks. The consciousness level was E4V4M5. Blood pressure was 131/70 mmHg with a heart rate of 65/min. Muscular power was grade 5/5 (Medical Research Council Scale) in the right upper and lower limbs, and grade 1/5 in the left upper and lower

![Figure 1](image-url)

*Figure 1.* (A): Magnetic resonance imaging, including T1, diffusion weighted imaging (DWI) and apparent diffusion coefficient map (ADC map). (B): Symptomatic time-of-flight angiogram. (C): Digital subtraction angiogram of the bilateral internal carotid arteries.
limbs. Deep tendon reflexes were all positive for all limbs, but were increased for the left upper and lower limbs. Pinprick, vibration and light touch sensations were all intact on the trunk and all limbs. There was no involuntary movement or signs of cranial neuropathy.

Magnetic resonance imaging showed encephalomalacia at right frontal lobe (Fig. A, filled arrows), and diffusion weighted imaging showed a hyperintense area at right superior parietal lobe (Fig. A, hollow arrows). A time-of-flight angiogram showed that the either ICA was normal, but the bilateral middle cerebral arteries were occlusive and the basilar artery was arteriosclerotic (Fig. B). Thus, she was diagnosed with acute right parietal infarctions, which were stage II (Brunnstrom classification) at left upper extremity and stage III at left lower extremity. A common blood examination, biochemistry, immune index and urine analysis were unremarkable.

Over the following week of hospitalization, she took 100 mg aspirin daily. Her consciousness improved to E4V5M6, and her muscular power improved to grade 3/5 for the left upper and lower limbs. A digital subtraction angiogram demonstrated the presence of abundant "puff-like smoke" collateral arterial networks (Fig. C, hollow arrow-heads) from bilateral proximal middle cerebral arteries despite of the absence of atherosclerosis of either extra-cranial ICA. These findings suggested stage IIb MMD (Suzuki and Takaku classification modified by Fukuyama and Umezu(9,10). Over the following half year, she took aspirin every day and received regular rehabilitation. The left hemiplegia has continued to be mild and has not deteriorated.

**DISCUSSION**

Digital subtraction angiography has been the goldstandard for the diagnosis of MMD, and is useful when assessing the development of collateral arterial networks(10). MMD has to be distinguished from moyamoya syn-drome (MMS), which can be associated with certain systemic conditions such as atherosclerosis, sickle cell disease, chronic basilar meningitis, neurofibromatosis, radiation, homocysteinuria, brain neoplasm, head trauma, Down's syndrome and autoimmune disease(2,3,11). Although a direct or indirect revascularization procedure is beneficial to reducing ischemic symptoms and improving neurological outcome than leaving the situation to its natural course or to medical treatment(2,13), conservative treatment would be recommended for an extremely old patient more than a younger patient; thus, surgery was unnecessary with our case because aspirin was able to effectively control her symptom over time. Nonetheless, this patient needs long term follow up. In conclusion, MMD does occur to extremely old Taiwanese, as old as over 80 years old, and thus be a differential diagnosis of cerebral infarction in an extremely old patient, whom might have more vascular diseases than a younger patient. Digital subtraction angiography is the gold standard for the diagnosis.

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