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 oldas 85+ years, and 68 years in Taiwan; therefore, MMD is generally thought not to occur in extremelyold 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 middlecerebral arteries despite the absence of arteriosclerotic stenosis of the bilateral extracranial internalcarotid 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 bea differential diagnosis of cerebral infarction in an extremely old patient. Digital subtraction angiogra-phy is the gold standard for the diagnosis.

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

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INTRODUCTION

Moyamoya disease (MMD) is a cerebrovascular disease involving progressive occlusion that is character-ized by stenosis or occlusion at the terminus of the inter-nal carotid artery (ICA) or the proximal areas of theanterior/ middle cerebral arteries with concurrent forma-tion of pathognomonic collateral arterial networks (retemirabile) near the occlusive or stenotic lesions[1,2]. Definite MMD is diagnosed when there are patientswith bilateral lesions and probable MMD is diagnosedin unilateral lesions. The typical clinical features arecerebral infarction, recurrent transient ischemic attacksand convulsions together with involuntary movementsand headaches in most children and young patients; fur-thermore, there is intracranial bleeding in about half ofadult patients despite the absence of any intracranialaneurysm[3]. The incidence is 0.086 per 100,000 in USA[4], and 0.01 to 0.048 per 100,000 in Taiwan[5]. AlthoughMMD often attack children less than ten years old[2], theoldest patient may have been 85+ years

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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 thoughtnot to occur in extremely old patients in Taiwan. Hereinwe report an 82-year-old woman diagnosed with MMDas 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 yearearlier and had been bothered with left hemiplegia sincethen. She was sent to our hospital because the left hemi-plegia had been deteriorating for two weeks. The con-sciousness level was E4V4M5. Blood pressure was131/70 mmHg with a heart rate of 65/min. Muscularpower was grade 5/5 (Medical Research Council Scale) in the right upper and lower limbs, and grade 1/5 in theleft upper and lower

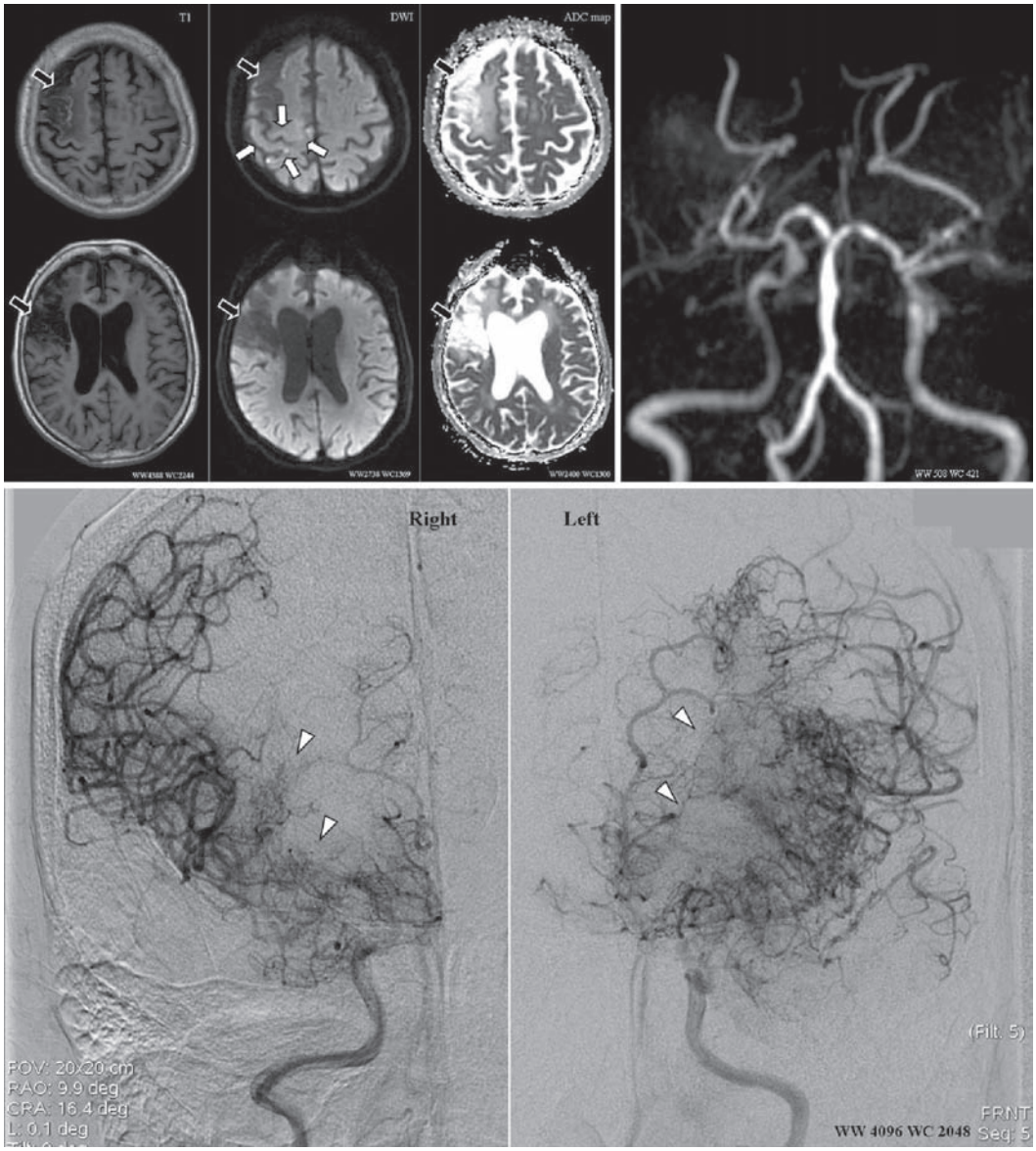

Figure 1. (A): Magnetic resonance imaging, including T1, diffusion weighted imaging (DWI) and apparent diffusion coefficient map (ADCmap). (B): Symptomatic time-of-flight angiogram. (C): Digital subtraction angiogram of the bilateral internal carotid arteries.

limbs. Deep tendon reflexes wereall positive for all limbs, but were increased for the leftupper and lower limbs. Pinprick, vibration and lighttouch sensations were all intact on the trunk and alllimbs. There was no involuntary movement or signs ofcranial neuropathy.

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

Over the following week of hospitalization, she took100 mg aspirin daily. Her consciousness improved to E4V5M6, and her muscular power improved to grade3/5 for the left upper and lower limbs. A digital subtrac-tion angiogram demonstrated the presence of abundant"puff-like smoke" collateral arterial networks (Fig. C, hollow arrow-heads) from bilateral proximal middlecerebral arteries despite of the absence of atherosclerosisof either extra-cranial ICA. These findings suggestedstage IIIb MMD (Suzuki and Takaku classification mod-ified by Fukuyama and Umezu)[9,10]. Over the followinghalf year, she took aspirin every day and received regularrehabilitation. The left hemiplegia has continued to bemild and has not deteriorated.

DISCUSSION

Digital subtraction angiography has been the goldstandard for the diagnosis of MMD, and is useful whenassessing the development of collateral arterial networks[3]. MMD has to be distinguished from moyamoya syn-drome (MMS), which can be associated with certain sys-temic conditions such as atherosclerosis, sickle cell disease, chronic basilar meningitis, neurofibromatosis, irradiation, homocysteinuria, brain neoplasm, head trauma, Down's syndrome and autoimmune disease[2,3,11]. Withour patient, a preexisting or age-related vascular diseasewas possible because no vascular studies have been doneprior to the symptomatic onset; besides, the previousright cerebral infarction relating encephalomalacia (Fig. A, filled arrows) did not necessarily attribute to MMD; however, those MMS conditions could be excludedbased on history, blood examinations and imaging studies.

MMD is of unknown etiology, and possibly geneticor associated with infection. Intraoperative and patho-logical observations have shown that the outer diametersof the relevant ICA terminus is markedly reduced due tointimal fibrocellular thickening, an uneven waving of theinternal elastic lamina and attenuation of the media; then, pathognomonic collateral arterial networks areextensively developed and seem to be closely associatedwith the onset of infarction and hemorrhage[3]. In addition, an essential arterial changes could occur to the ver-tebrobasilar system due to the compensatory verte-brobasilar hypertension secondary to the ICA occlusionor stenosis[9,12]. With our patient, the basilar arterial arte-riosclerosis (Fig. B) is suggestive of hemodynamicchanges and consequent tissue responses secondary tothe ICA terminus stenosis.

MMD patients with milder symptoms are usually treated by medical treatments, including vasodilators, antiplatelet agents, antifibrolytic agents, and fibrinolyticagents, but their efficacies are doubtful[13]. Although adirect or indirect revascularization procedure is benefi-cial to reducing ischemic symptoms and improving neu-rological outcome than leaving the situation to its naturalcourse or to medical treatment[2,13], conservative treat-ment would be recommended for an extremely oldpatent more than a younger patient; thus, surgery wasunnecessary with our case because aspirin was able toeffectively control her symptom over time. Nonetheless,this patient needs long term follow up. In conclusion,MMD does occur to extremely old Taiwanese, as old asover 80 years old, and thus be a differential diagnosis ofcerebral infarction in an extremely old patient, whomight have more vascular diseases than a younger patient. Digital subtraction angiography is the gold standard for the diagnosis.

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