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Case Report

# Moyamoya Disease Presenting with Hemichorea: A Case Report

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We report an unusual case of a 13-year-old boy with moyamoya disease whose predominant manifestation was right hemichorea with initial dystonic spasms of the right limbs and face. A diffusion-weighted MR image of the brain in the initial stage showed no infarcted or other parenchymal lesions. However, MR angiography and intracranial angiography showed stenosis of the bilateral internal carotid and middle cerebral artery with prominent basal collaterals, which was compatible with moyamoya disease. Chorea or hemichorea is one of the rarer presenting features of moyamoya disease. Our findings suggest that moyamoya disease should be included in the differential diagnosis of pediatric onset chorea. ( Tw J Phys Med Rehabil 2010; 38(3): 169 - 174 )

**Key Words:** moyamoya disease, hemichorea, diffusion-weighted MR imaging

## INTRODUCTION

Moyamoya disease (MMD) is an uncommon cerebrovascular disorder which is characterized by bilateral progressive occlusion of the supraclinoid internal carotid artery (ICA) as well as its main branches within the circle of Willis. Diagnosis of MMD is achieved by angiography showing abnormal collateral circulation at the base of the brain triggered by stenosis or occlusion at the terminal portions of the ICA.[1] It usually presents with variable neurologic symptoms and recurrent ischemic attacks or seizures in children, whereas hemorrhagic stroke is more common in adults.[2] Interestingly, rare cases of MMD have been diagnosed with the involuntary movements of hemichorea, which are usually described as an initial presentation of the disease.[3] Most children with MMD experience transient ischemic attacks or cerebral infarctions, and diffusion-weighted magnetic resonance imaging (DWI) of the brain can be used to localize these infarcted lesions.

We describe a 13-year-old boy who initially presented with right hemichorea, which mimicked dyskinetic cerebral palsy, epilepsy or myopathy. Despite normal DWI of the brain, the patient was diagnosed as MMD by internal carotid artery (ICA) angiography and successfully treated surgically.

## CASE REPORT

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A 13-year-old right-handed boy who was a junior high school student had been suffering from insidious night headaches, fatigue and a tingling sensation in his right hand and right foot before his final term examinations. Two weeks later, he presented with a sudden onset of inertia in his right hand and forearm and subsequently involuntary movements in his right hand and right foot. Three weeks later, the involuntary movements progressed to involve the right limbs together with drooling, poor oral movement and right hemifacial spasms. Over a period of one week, around 10 hemichorea events were observed with a duration ranging from 20 minutes to one hour. The involuntary movements gradually increased in frequency and interfered with his activities of daily living. He was transferred to our rehabilitation department due to progressive right limb weakness three weeks after the first onset of the symptoms. The thin and weak boy presented with intermittent slow hemichoreic movements in his right limbs and right face and tongue fasciculation when sitting on a chair. No congenital malformations were found at birth, but he had developed sensory integration dysfunction during his childhood. On physical examination, he had an obviously weak level of muscle strength (4/5) in his right limbs. Bilateral winging scapula with negative Gower's sign was also observed. He had decreased pain, position sensation, vibration sensation, and proprioception in the right limbs. His tandem gait was normal, but other cerebellar function tests were difficult to investigate because of his involuntary movements. Deep tendon reflexes were symmetric and normoactive. Babinski's sign presented over his right side. Routine laboratory and blood tests were performed and yielded normal results (e.g. antistreptolysin O titers and ceruloplasmin levels, thyroid function tests, serum rheumatoid factor, lupus erythematosus cell test, antinuclear antibody, anti-DNA, anti-Ro, anti-La, anticardiolipin antibody, and lupus anticoagulant tests, protein C, protein S, and anti-thrombi III, Human Immunodeficiency Virus [HIV] testing), except that the creatine phosphokinase (CPK) test showed a high level of 554 ng/ml (normal range <500 ng/ml). From our observations, magnetic resonance (MR) imaging with T2-weighting, fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted imaging (DWI) of the brain showed no ischemic, hemorrhagic or mass lesions (Figure 1a, 1b). We assumed that dyskinetic cerebral palsy, epilepsy or myopathy could be contributing to this case. However, electroencephalography revealed bilateral hemispheric slow wave, which increased as a build-up phenomenon with the manifestation of delta wave induced by hyperventilation. Under the suspicion of a brain focal vascular lesion or encephalopathy, we performed bilateral ICA angiography which showed significant stenosis at bilateral distal internal carotid arteries, bilateral A1 segments, right M1 segment, and left middle cerebral artery narrowing with the presence of moyamoya vessels (Figure 1c, 1d). Single photon emission computerized tomography (SPECT) of the brain showed hypoperfused areas in the left frontoparietal lobes and putamen (Figure 2). Two weeks later, he was referred to our neurosurgery department and underwent left superficial temporal artery-middle cerebral artery bypass combined with encephalo-duro-arterio-myo-synangiosis. His hemichoreic movements resolved completely after the operation two weeks later. His right limb weakness gradually improved over a 2-month rehabilitation program. Brain SPECT revealed normal in contrast to previous hypoperfused areas in the left frontoparietal lobes and putamen. He was asymptomatic throughout a 6-month course of follow-up.

## DISCUSSION

Chorea is a movement disorder that presents as frequent, brief, sudden, twitch-like, involuntary movements arising from a continuous flow of random muscle contractions that progress from one part of the body to another in a chaotic manner. It may be either inherited owing to genetic mutations or acquired due to infections, autoimmune disease, neural degeneration, stroke, tumors, medication exposure, and metabolic diseases. MMD, an intracranial vascular lesion, is a very uncommon cause of chorea, and it must be included in the differential diagnosis when children developed sudden onset of involuntary movements.[4] We reported a case of a Taiwanese adolescent with MMD presenting with hemichorea induced by the stress of final examinations. Initially, he was misdiagnosed as myopathic, dyskinetic cerebral palsy or epileptic because of the high level of the CPK test and the normal brain MR imaging, including T2 FLAIR and DWI. Acute infarcts may be seen at an early stage on conventional

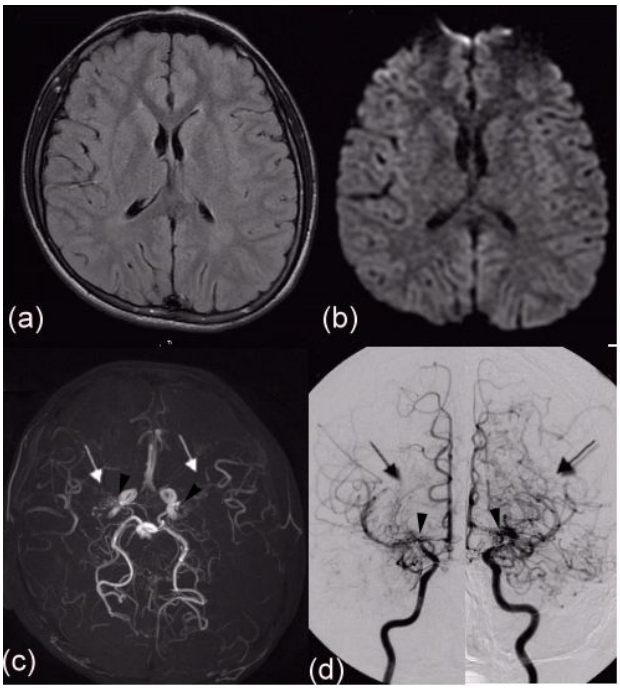


Figure 1. MR imaging with (a) Water suppressed proton (FLAIR) imaging. (b) Axial DWI of the brain showed no abnormal findings before the operation. (c) MR angiography showed significant narrowing of the supraclinoid portion of both distal internal carotid arteries (arrow) and abundant collaterals from the bilateral posterior cerebral arteries (arrow head) (d) Bilateral internal carotid angiography demonstrated both supraclinoid carotid narrowing (arrow) with a “puff of smoke” appearance of the collateral vessels (arrow head) in the neighborhood of the stenotic area.

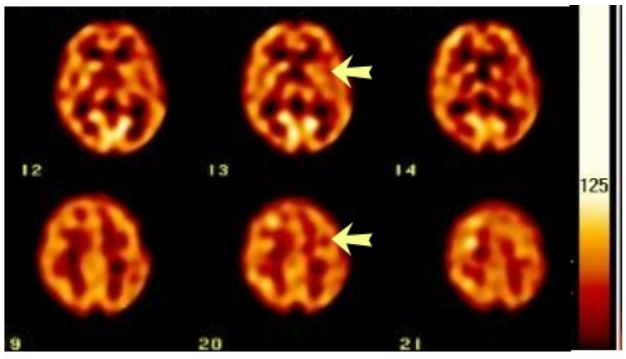


Figure 2. The SPECT brain perfusion images of our patient. Note the relatively decreased activity in the left putamen and frontoparietal area indicated by the open arrowhead.

DWI with a higher sensitivity of 88%~100% and specificity of 86%~100% by the diagnosis of hyperacute ischemic stroke.[5-6] Imaging studies of T2 FLAIR and DWI in our patient were normal. This may be due to abundant collateral circulation to the ischemic area in MMD. An article by Inoue et al provided an extensive discussion of negative DWI in patients with transient ischemic attack and reversible ischemic neurological deficit, especially in those with a rich collateral circulation. Ischemic stroke patients whose imaging studies of T2 FLAIR and DWI are normal always recover completely from their transient neurological deficits.[7] Therefore, it is still hard to explain the normal imaging studies of T2 FLAIR and DWI in our patient with the gradual progression of right hemichorea, hemianesthesia and hemiparesis.

After excluding other diagnosis including metabolic, toxic, or drug induced chorea, Sydenham’s chorea, systemic lupus erythematosus, protein C, protein S and plasminogen deficiency, antiphospholipid antibody syndrome, multiple sclerosis, HIV encephalopathy, and bacterial endocarditis, we arranged advanced neuroimaging studies of bilateral ICA angiography due to his abnormal EEG findings.[1] Bilateral ICA angiography in our patient showed stenosis of bilateral ICA and a "puff of smoke" appearance at the intracranial artery. To clarify the pathophysiology of chorea, we considered the abnormality within a complicated neuronal network communicating motor cortex, including the premotor cortex and supplementary motor area and a group of subcortical nuclei collectively termed the basal ganglia and thalamus.[8] Chorea is an uncommon complication of acute vascular lesions, reported in less than 1% of patients with acute stroke. Vascular hemichorea or hemiballism is typically associated with ischemic or hemorrhagic lesions of the basal ganglia and adjacent white matter in the territory of the middle or the posterior cerebral artery.[9] SPECT imaging of the brain revealed that the blood perfusion of the left putamen and frontoparietal area contralateral to his paroxysmal hemichorea was significantly decreased. This finding suggests that the interruption of the basal ganglia-cortical circuits may have been closely related to the development of chorea in this patient.

Early surgical intervention is the therapy of choice for pediatric MMD because of progressive occlusive

vasculopathy. Surgical intervention should be undertaken early, before irreversible damage occurs. Various surgical revascularization options, including direct bypasses such as superficial temporal artery-middle cerebral artery bypass (STA-MCA), indirect bypasses such as encephalo-duro-arterio-myo-synangiosis (EDAMS), encephalo-duro-arterio-synangiosis (EDAS), omentum transplantation and burr hole operations, and other techniques have all been proved effective in reducing the occlusion of intracranial vessels. Excellent results have been reported in a combination of surgeries (STA-MCA bypass with EDAMS) for pediatric MMD in reducing the risk of the postoperative ischemic attacks compared with other surgery.[10-11] In our patient, we preformed only left STA-MCA bypass combined with EDAMS because his involuntary movements were limited to his right hemichorea and weakness. In unilaterally symptomatic MMD, bypass surgery for the asymptomatic side can be progressed slowly until the development of ischemic symptoms, such as frequent transient ischemic attacks.[12] The involuntary movements disappeared two weeks after the operation and brain SPECT revealed that the cerebral blood flow had normalized two months after surgical revascularization. His hemichorea did not recur during the follow-up period.

The causes of hemichorea can be variable and complicated. Hemichorea or chorea occurs only in 3 to 6% of children with MMD. Previous studies have attributed hemichorea or chorea in MMD to ischemic lesions in the basal ganglia-thalamocortical circuit.[13-17] Lyoo et al reported no ischemic change on brain MRI in one case with MMD and chorea, but they did not mention DWI studies.[14] In our patient, T2 FLAIR and DWI studies revealed no specific findings.

In conclusion, our case report suggests that MMD should be included in the differential diagnosis of childhood and adolescent chorea or hemichorea. Imaging studies of the cerebral arteries should be performed to recognize this uncommon form of occlusive carotid disease for the early diagnosis and proper intervention of neurological deficits.

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# 以舞蹈症爲表現的毛毛樣腦血管病變：病例報告

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我們報導一個不常見的右側偏側舞蹈症的 13 歲男孩，剛開始以右側肢體以及臉部的肌張力異常來表現。起初，水分子擴散磁振造影影像檢查並沒有發現到有腦部梗塞或是其他腦實質病兆。然而，磁振影像血管攝影術以及顱內血管攝影術顯示兩側內頸動脈與中大腦動脈狹窄，且合併明顯的基底動脈側枝循環，我們確定診斷病人爲毛毛樣腦血管病。舞蹈症或是偏側舞蹈症在毛毛樣腦血管病是少見的臨床表現。我們建議毛毛樣腦血管病應該列入兒童時期發生的舞蹈症的鑑別診斷。( 台灣復健醫誌 2010；38(3)：169 - 174 )

**關鍵詞：**毛毛樣腦血管病(moyamoya disease)，偏側舞蹈症(hemichorea)，水分子擴散磁振影像(diffusion-weighted MR imaging)

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