Moyamoya disease and migraine-like headaches

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Summary

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Rarely, adult moyamoya disease presents with recurrent migraine-like headaches during a long course. We report the case of a 31-year-old woman with moyamoya disease and migraine-like headaches, with a 13-year history. Physical examination showed no neurologic deficits. Magnetic resonance angiography (MRA) and digital subtraction angiography (DSA) showed occlusion of the right internal carotid artery (ICA), severe stenosis of the left ICA, and abnormal vascular network at the base of the brain. The calcium channel blocker, nimodipine, was introduced in order to prevent further symptoms. After the introduction of nimodipine, no further headache, weakness and numbness of the left hand occurred in this patient. This suggests that migraine-like headaches may be caused by moyamoya disease and that nimodipine may have a beneficial effect on this condition.

Keywords: migraine; moyamoya disease; nimodipine

Introduction

Moyamoya disease is an obstructive cerebrovascular disease characterised by peculiar stenosis or occlusion of the internal carotid arteries or its terminal branches associated with teleangiectatic vessels at the base of the brain [1]. The clinical manifestation depends on age: Headache, seizure, transient ischemic attacks (TIAs) and ischemic strokes often with progressive intellectual deterioration are the predominant presentation in childhood, whereas subarachnoid and intracerebral hemorrhage occur more frequently in adults [2]. Although headache is one of the common symptoms of moyamoya disease, it is often associated with other residual symptoms such as weakness, paresis or paralysis of the extremities [2, 3]. It is very rare that the adult patient with moyamoya only presents with recurrent migraine-like headaches in the absence of other neurologic deficits during a long duration of illness. We report the case of an adult woman with recurrent migraine-like headaches associated with moyamoya disease with a history of more than 13 years.

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Case report

A 31-year-old right-handed woman was admitted to the Department of Neurology of Jinling Hospital in October 1998 with a 13-year history of recurrent migraine attacks with aura. In October of 1985, she developed for the first time headache in both temples and behind the right eye associated with nausea, vomiting and photophobia during three days with a frequency of one to three times a day. Each episode lasted about 30 minutes to 2 hours. In November 1989, she had girdle headache episodes in the right frontal temple and the retro-orbital region accompanied again by anorexia, nausea and vomiting. About 10–30 minutes before the headache, there were prodromi including bright sparkles and scintillating zigzag lines in front of her eyes. The episodic headache continued for 10 days, and anodyne could not relieve the pain. Later the headaches occurred for about two years with almost the same symptoms as mentioned above, especially during the change of autumn to winter and at times of fatigue. These episodes lasted about three days to one week and disappeared after a few days of rest and good sleep. Since the end of 1997, the episodic headaches had become severe and frequent, sometimes associated with paresis of the left hand and numbness of the left finger tips which persisted five to ten

Figure 1 Axial MRA showing rarefaction of the right internal carotid artery (R-ICA), compensatory coarsening of the basilar artery (BA), and formation of fine collateral vessels in the base of brain.

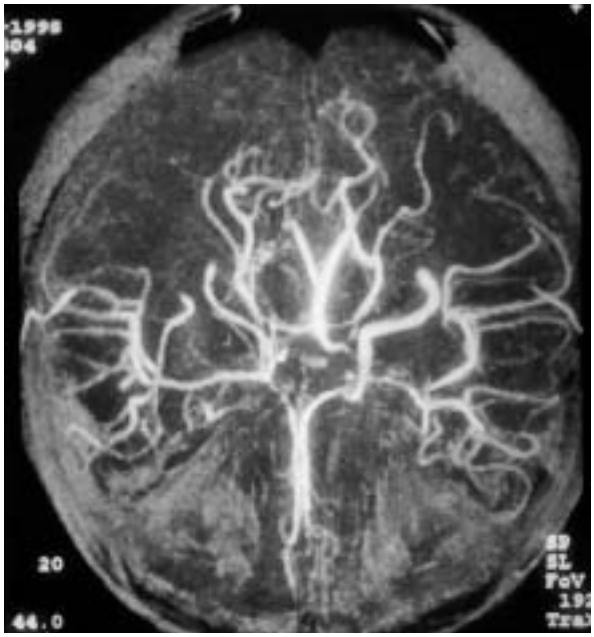


Figure 2 DSA of R-ICA showing occlusion of the supraclinod R-ICA and coarsening of the right ophthalmic artery.

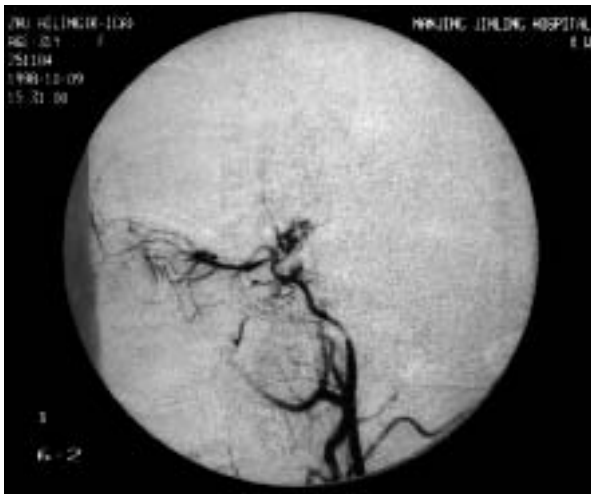


Figure 3 DSA of Left ICA showing severe stenosis in the siphon of L-ICA, light imaging of L-MCA and L-ACA.

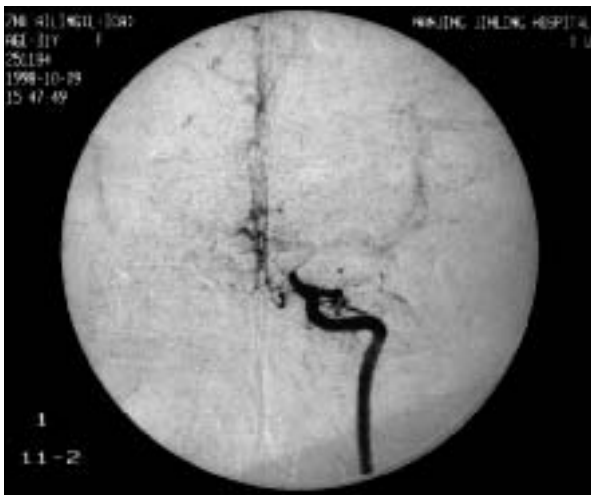
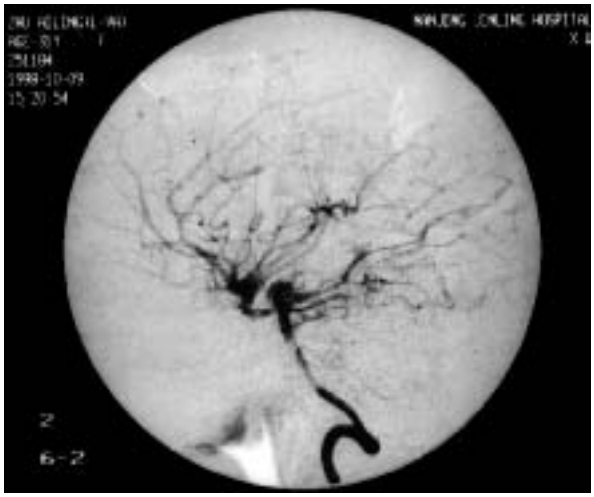


Figure 4
DSA of L-VA, blood of bilateral anterior cerebral arteries (ACA), middle cerebral arteries (MCA) were provided by left VA via posterior communicating artery (PCA).



minutes for each time, before, following or coexisting with the onset of headaches. The patient had a family history of migraine with her father, no history of seizures or trauma, and no risk factors for stroke.

Physical examination was entirely normal and showed no neurologic deficits. The related blood examinations such as erythrocyte sedimentation, somatic antibody and leptospiral antibody etc. were all negative. Electroencephalography and computerized tomography were normal. Doppler ultrasonography showed severe stenosis of the right supraclinoid internal carotid artery and middle cerebral artery, vasospasm on the right anterior cerebral artery, left MCA, and slow flow in the vertebrobasilar artery. Magnetic resonance imaging (MRI) of the brain was normal. Axial magnetic resonance angiogram (MRA) showed rarefaction of the right internal carotid artery (ICA), compensatory coarsening of the basilar artery (BA), and formation of fine collateral vessels in the base of brain (fig. 1). Digital subtraction angiography showed occlusion of the right supraclinod ICA (fig. 2), severe stenosis of the left siphon ICA (fig. 3), occlusion of the distal right vertebral artery. Blood flow of bilateral ACA, MCA were provided by left VA via posterior communicating artery (PCA) (fig. 4). Collateral vessels from the right external carotid artery (ECA) to the intracranial region were also visualized by DSA.

This patient was treated with Nimotop® (nimodipine), with a dose of 10 mg by intravenous transfusion, one time a day during 14 days. After then, she was maintained on a dose of 30 mg three times a day per os, at the follow-up five months later, there was no further headache, no weakness on the left hand and numbness of the finger tips.

Discussion

Only a few previous reports indicate that headache is one of the common symptoms in pediatric patients with moyamoya disease, but it is always associated with other residual symptoms. Yamashiro et al. [3] reported the clinical features of 10 Japanese children with moyamoya disease. The most common initial manifestations were headache in four cases (40%), motor deficit and convulsion in three cases (30%). As residual symptoms, 8 children (80%) developed motor deficits with hemiplegia in 5 cases, and paresis or weakness of the extremities in 3 cases (30%), and 4 (40%) had headaches. Battistella and Carollo [4] described 34 Italian patients (15 males and 19 females) suffering from moyamoya disease. The early clinical symptoms consisted of TIAs and/or stroke (20 cases, 44%), recurrent migraine-like headaches (7 cases, 20%), seizures (6 cases, 18%) and hemorrhage (one case, 3%).

The clinical presentation of this case fulfils the diagnostic criteria of classic migraine [5]. It is rare that in the adult patient the only presentation is recurrent migraine-like headaches, in the absence of other residual symptoms such as weakness of the extremities, seizures and sensory disturbance. During such a long duration of illness, a diagnosis of migraine was made in several hospitals and moyamoya disease was not diagnosed until October 1998. Bernstein [6] reported the case of a 6-year-old girl with hemiplegic migraine in association with moyamoya disease. This child presented with a 6-month history of headaches that occurred every 7 to 10 days and were associated with nausea, vomiting, and photophobia. When symptoms occurred, 2 to 3 hours of sleep alleviated them. On three occasions, she awoke with weakness on the right side, numbness, and garbled speech followed by severe headache, nausea, and vomiting; again all symptoms resolved after 2 hours of sleep. This child had a relatively short history, and headache was associated with transient local neurologic deficits on three occasions. However, the case we report presented with a 13-year history of migraine-like headaches that was not associated with transient weakness and numbness until the end of 1997.

Moyamoya disease is rare, and migraine is common. However migraine-like headaches might be caused by moyamoya disease. It was difficult to differentiate the episodes of headache between migraine or TIAs, only according to the initial clinical presentations in the case reported herein. Because the recurrent headaches were associated with local deficits as numbness, weakness, paresis or paralysis of extremities, it might be a strong indication of cerebral ischemic attack.

The efficacy of the medical and surgical treatment of moyamoya disease is still controversial [8]. Surgical revascularization procedures have occasionally produced some benefit. Pharmacotherapeutic agents, such as aspirin, steroids and vasodilators, have been tried in the past without success [9]. There are a few reports that mention treatment of moyamoya disease with calcium channel blockers, which might be effective in preventing further symptoms [4, 9, 10]. Nimodipine seemed to be also efficacious in our case. Nimodipine may have some effects on the remission of ischemic symptoms, but further studies are needed to determine the effectiveness of nimodipine in the treatment of this condition.

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