Moyamoya Disease : A Case Report and Review of the Literature

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Moyamoya disease is characterized by the angiographic findings of bilateral occlusion of the distal portion of the internal carotid arteries with a vascular network at the base of the brain. The typical findings on conventional angiography and magnetic resonance imaging usually confirm the correct diagnosis of moyamoya disease. We experienced a 11-year-old girl with moyamoya disease showing repeated transient ischemic attacks and we report on this case with a review of the literature.

Key words : moyamoya disease, magnetic resonance imaging, magnetic resonance angiography, cerebral angiography

Introduction

Moyamoya disease is characterized by progressive stenosis or occlusions in the distal portion of the bilateral internal carotid arteries and the circle of Willis associated with an abnormal fine vascular network at the base of the brain ("moyamoya").

Moyamoya disease was first described by Takeuchi and Shimizu in 1957[1,2]. The incidence of moyamoya disease is predominantly found in East Asians, especially Japanese and Koreans[3]. The male-to-female ratio of moyamoya disease is generally known as 1:1.8[4].

Moyamoya disease most commonly presents in the first decade of life and also presents in the fourth decade of life. But clinical features are not uniform. Most children usually develop repeated transient ischemic attack or cerebral infarction, On the contrary, adults more frequently occur intracranial hemorrhage[5-8]. The etiology of moyamoya disease is not known. Cerebral angiography is a basic procedure to confirm the diagnosis of moyamoya disease.

Recently, magnetic resonance angiography has been accepted as a noninvasive diagnostic modality for moyamoya disease. We report a 11-year-old girl presenting with left hemiparesis and we review the literature on reported cases of moyamoya disease.

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Case

A 11-year-old girl had experienced repeated progressive left paraplegia and loss of consciousness during three months prior to admission. Three weeks before admission, she had suffered from loss of consciousness for two or three minutes after hyperventilation. She was then maintained on regular anticonvulsant medication. She was admitted to our hospital with the chief complaint of severe headache, sensory disturbance of left face, and left hemiparesis. She had no family history, no other history of disease and no past trauma. She did not have fever. A physical examination revealed no abnormalities. On neurological examination, she had grade II motor weakness of left upper arm. Electroencephalogram showed mild cerebral dysfunction. Laboratory studies including routine blood tests, and urine analysis were within normal limits.

Computed tomography performed at a local hospital demonstrated a low density lesion in the right frontal lobe, consistent with cerebral infarction(Fig. 1). Magnetic resonance imaging obtained 1 day after admission revealed a well defined triangular low signal intensity on the T1-weighted images and a high signal intensity on the T2-weighted images in the right frontal white matter(Fig. 2). Magnetic resonance angiography showed symmetrical stenosis of distal portion of bilateral internal carotid arteries, origin of the anterior and middle cerebral arteries, and proximal portion of posterior cranial arteries. She was diagnosed as moyamoya disease(Fig. 3). Conventional cerebral angiography obtained 4 days after admission demonstrated typical stenosis of the bilateral distal

portions of internal carotid artery, the bilateral M1 portions of middle cranial arteries, and the bilateral A1 portions of the anterior cranial arteries(Fig. 4). Consequently, the patient was confirmed with moyamoya disease.

Follow up single photon emission computed tomography showed decreased regional cerebral blood flow in the right frontal region(Fig. 5). Surgery was recommended but her parents refused the proposal and wanted to be discharged because her neurologic status had not been improved.

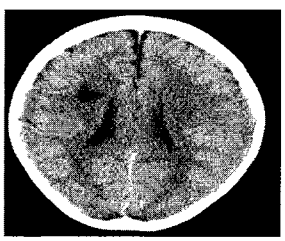

Fig. 1. CT shows a low density area in the right frontal lobe.

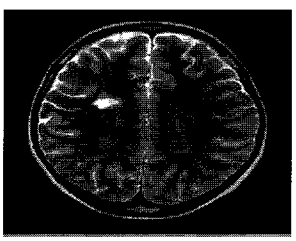

Fig. 2. T2-weighted magnetic resonance imaging reveals triangular shaped high signal intensity lesion in right frontal white matter.

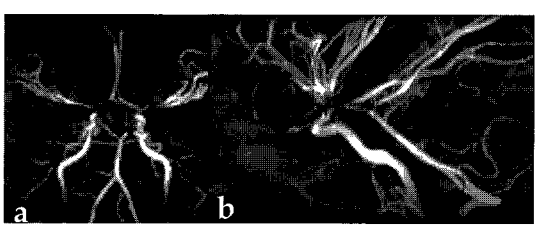

Fig. 3. Magnetic resonance angiography (a : A-P view, b : lateral view) shows symmetrical stenosis of distal portion of bilateral internal carotid arteries, origin of the anterior and middle cerebral arteries, and proximal portion of posterior cranial arteries.

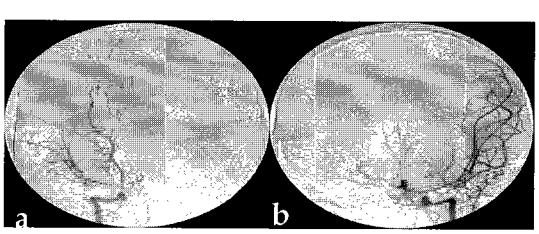

Fig. 4. Cerebral angiography (a : right carotid angiography b : left carotid angiography) shows stenosis of the bilateral distal portions of internal carotid artery, the bilateral M1 portions of middle cranial arteries, and the bilateral A1 portions of the anterior cranial arteries.

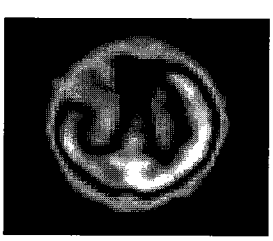

Fig. 5. Single photon emission computed tomography shows decreased regional cerebral blood flow in the right frontal region.

Discussion

Moyamoya disease is a rare cerebrovascular disorder characterized by bilateral stenosis or occlusion of the distal portions of the internal carotid arteries, proximal portion of anterior cranial arteries and middle cranial arteries with development of net-like collateral vessels at the base of brain[1].

Moyamoya disease was initially described by Takeuchi and Shimizu in 1957[2]. The abnormal vascular network is found on angiography[1,2]. Collateral vessels consist of leptomeningeal vessels, perforating basal ganglionic vessels, and transdural anastomoses from external carotid artery. Angiographic appearance of these abnormal fine collateral vessels, named "moyamoya" or "something hazy just like a puff of cigarette smoke in the air" in Japanese, gives the disease its name[1,6]. Moyamoya disease reveals regional and racial differences. The incidence of the moyamoya disease is a predominantly higher in Asian populations, such as Japanese and Korean. Among Japanese, the annual incidence of 0.35 per 100,000 populations has been reported[4]. Moyamoya disease has a bimodal age distribution, there is the first peak in the first decade of life and a second smaller peak in the fourth decade of life[6,9,10]. The clinical manifestations of moyamoya disease differ considerably between pediatric and adult patients. Cerebral ischemic attacks are seen mainly in pediatric patients, whereas cerebral and subarachnoid hemorrhages are more commonly seen in adults[11,12]. Right cerebral infarction was observed from our study.

Although the exact pathogenesis of moyamoya disease remains unclear, several pathological theories have been proposed including vascular growth factors, cytokines, basic-fibroblast growth factor or genetic abnormality[13-15]. The histopathological findings of autopsy studies reveal that distal internal cranial arteries, and proximal anterior and middle cerebral arteries are severely narrowed or occluded by an extensive fibrocellular thickening of the intima. The intima is typically a laminated structure with duplication or triplication of the internal elastic lamina. Other associated findings include frequent mural thrombi and occasional lipid deposits, but inflammatory cells are conspicuously absent. Accumulating evidence suggests that smooth muscle cell proliferation and phenotypic modulation may underlies these observed vessel changes[16]. The basal moyamoya collaterals have unique pathological characteristics. Some are thin-walled and dilated, while others are thick walled and stenotic[17].

Once the diagnosis of moyamoya disease is suspected on clinical evidences, noninvasive imaging modalities play a major role in suggesting the diagnosis of moyamoya disease.

Although abnormalities of brain can be detected on computed tomography, magnetic resonance imaging provides important additional information including excellent ability to delineate cerebral vasculature. Therefore, when magnetic resonance imaging and magnetic resonance angiography show the typical findings of stenosis or occlusion of a major intracranial arteries in association with abnormal fine vessels at the basal brain, cerebral angiography is not essential to confirm the diagnosis of moyamoya disease[18-21].

Other imaging modalities, such as xenon-enhanced computed tomography, single photon emission tomography, and positron emission tomography, can be evaluated to measure regional cerebral blood flow, the clinical severity, and the effect of surgical revascularization[22-24].

Moyamoya disease must be considered in the differential diagnosis of other causes of progressive cerebrovascular occlusive disease including neurofibromatosis, arteriosclerosis, radiation therapy and sickle-cell disease[25].

Although many medical treatments of moyamoya disease have been performed, the efficacy of medical treatments has not been well satisfied. Surgical treatments of moyamoya disease are divided into two bypass procedures. Direct surgical treatments include superficial temporal artery-middle cerebral artery anastomosis, occipital artery-middle cerebral artery anastomosis, and vein grafts. Indirect surgical treatments include encephalo-duro-arterio-synangiosis(EDAS), encephalo-myo-arterio-synangiosis(EMAS), encephalo-myo-synangiosis(EMS), and transplantation of the omentum[26-29]. Surgical revascularizations for moyamoya disease have been performed to improve regional cerebral blood flow and may change the course of cerebral hemodynamic ischemic brain[30-33]. Many authors have reported that prognosis of untreated moyamoya disease is poor, more than 70 % rate of major neurological deficit or death in 2-3 years after diagnosis in children[34,35]. Our case plays an important role in the early diagnosis and treatment of moyamoya disease.

Conclusion

Moyamoya disease, the most common pediatric cerebrovascular disease in Asian populations, is characterized by distinct bilateral progressive steno-occlusive cerebrovascular disease. Children with cerebral ischemic features are more likely to have offensive examination with neuroradiological modalities such as magnetic resonance imaging and magnetic resonance angiography, which can be used as the primary diagnostic imaging modality than conventional cerebral angiography.

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