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A Child with Moyamoya Disease: A Case Report

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ABSTRACT

Cerebral stroke is a rare disease in children. Moyamoya (MM) is one of the infrequent cerebrovascular diseases with unknown etiology. We report an 8 year-old-boy with chief complain of sudden onset bilateral parietal lobe headache. He mentioned that his headache was first started about three weeks ago and was associated with visual disturbance. His mother declared that the boy developed gait problems few days later and only could walk with assistance. He was diagnosed with MM disease. After, medical treatment his symptoms were mildly improved and because of his family disagreement cerebral revascularization surgery was not performed.

Keywords: Moyamoya; Stroke; Child

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INTRODUCTION

Progressive occlusion or stenosis of the intracranial part of the internal carotid artery and/or the proximal portion of the anterior and middle cerebral artery has been shown in Moyamoya (MM) cerebral angiopathy. "Moyamoya" in Japanese means appearing as a "puff of smoke". This title is due to steno-occlusive pattern and collateral vessels at the base of the brain on angiography [1]. Primarily, this pattern of angiopathy was described in Japan in 1957 and first "Moyamoya disease" was called by Suzuki and Takaku 12 years later [2]. Common presentation of the disease in children is stroke, while subarachnoid and intracranial hemorrhage commonly presents in adult with MM disease [3]. We present a rare case of Moyamoya out of the South Asia.

CASE PRESENTATION

An 8-year-old boy was referred to our hospital with complaining of sudden onset bilateral parietal lobe headache. He mentioned that his headache was first started about three weeks ago and was associated with visual disturbance. His mother declared that the boy developed gait problems few days later and only could walk with assistance. She didn't mention any changes in mental status or in his speech. His past medical history was negative for developmental delay, metabolic disorders, neurologic disease or other medical illness. His family didn't report history of head trauma. There was no history of seizure, bowel or bladder incontinency or loss of consciousness. His only medication was Acetaminophen tablet for headache. No history of medical illness or neurologic disorder was reported in his parents and other family members.

On admission, his vital signs were as follow: blood pressure of 115/70 mmhg, pulse rate of 80/min, respiratory rate of 17/min, oral temperature of 36.7° C and O_2 saturation of 97%. He was alert and his speech was fluent but didn't make eye contact. General physical examination revealed no abnormalities.

On cranial nerve examination, his pupil was reactive and other cranial nerves examination was in normal limits. Babinski sign on the left side was positive. Also, deep tendon reflexes were diminished in the left side. Because of the patient's visual loss, his gait was not evaluated.

In laboratory tests complete blood cell count revealed: white blood cells of 9600, hemoglobin of 13, platelet of 215000. Erythrocyte sedimentation rate was 15.

The patient PT and PTT was 12 and 30, respectively. Biochemistry consists of blood sugar, blood urea nitrogen, creatinine and electrolytes were all in normal limits. Immunology tests including C3, C4, CH50 and ANA were negative.

Spiral brain computerized tomography scan showed atrophy of right parieto-occipito-trmporal lobes associated with ventricular dilation suggestive of remote ischemic insult (Figure 1). Also, hypodense regions were seen in right parieto-occipital lobe. Hypodensity involved both gray and white matter with sulcal swelling in left occipital lobe (Figure 2).

On brain angiography, there was a "puff of smoke" appearance that was suggestive for MM disease (Figure 3).

We initiated treatment of this patient with Aspirin, Glycerol and the patient showed a little improvement on his power. After that, due to his family disagreement for performing cerebral revascularization surgery, we continued medical treatment and his symptoms were mildly improved.

DISCUSSION

Occlusion of the circle of Willis arteries with chronic and progressive pattern and developing collateral vessels seen on imaging are characteristics in MM disease. MM disease occurs predominantly in Japanese patients but has been found all over the world [2,4,5].

External stimuli, injuries, or genetic defects may leads to narrowing of cerebral vessels. Sickle cell anemia, Down's syndrome, and neurofibromatosis-1 are some the diseases associated with MM disease in the literature [6].

In the beginning of the disease narrowing of the carotid arteries were evident. This pattern followed by the major cerebral arteries dilatation and collateral arteries (MMvessels) [7].

To confirm the diagnosis magnetic resonance (MR) angiography are used among different imaging modalities.

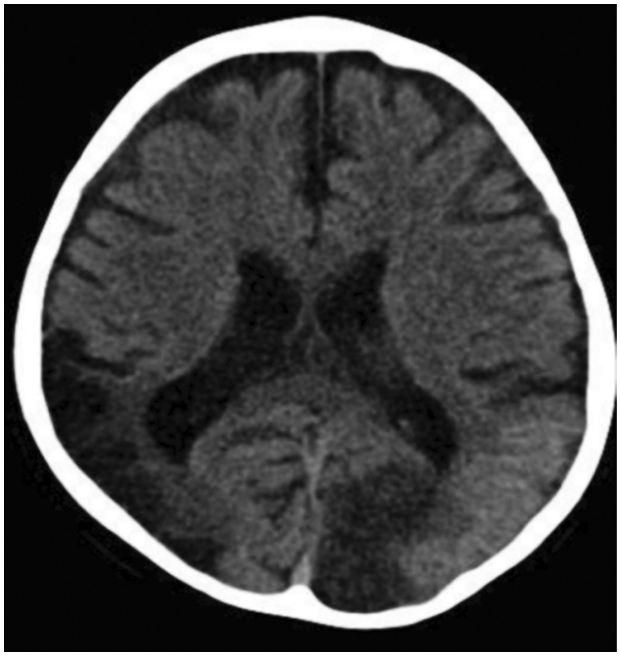

Figure 1. Atrophy of right parieto-occipito-trmporal lobes with ventricular dilation

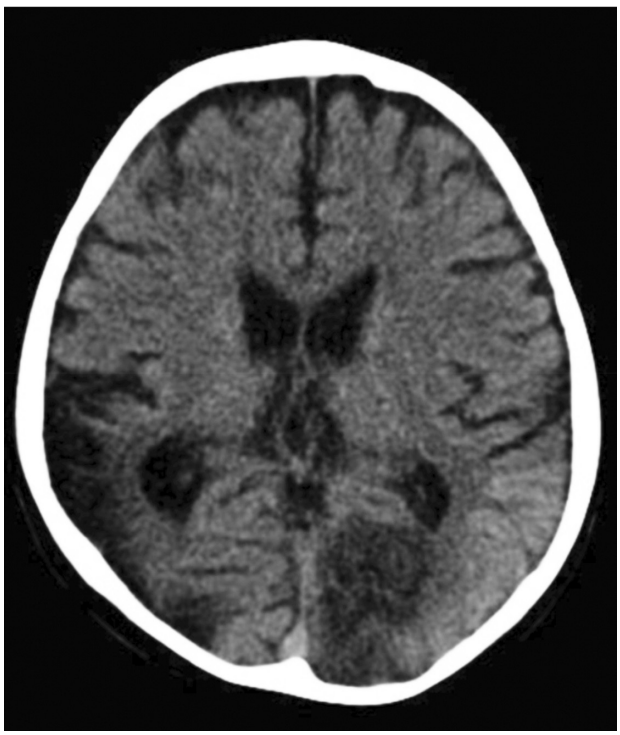

Figure 2. Hypo dense regions in right parieto-occipital lobe

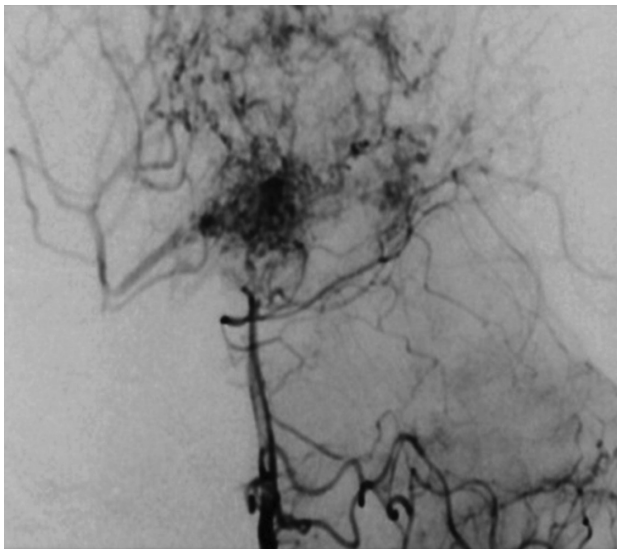

Figure 3. Puff of smoke appearance on brain angiography

On MR angiography, occlusion and narrowing of proximal part of cerebral vessels with collateral arteries are seen [8].

Ischemic MM angiopathy in all patients commonly is treated with medical therapy. For prevention of thrombosis and thromboembolism long-term oral antiplatelet therapy is used in children and adults. Decreased regional cerebral blood flow in the patient's angiography or clinical ischemic symptoms are some of the indications for revascularization surgery, according to Japanese and US guidelines [9,10].

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