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Moyamoya: Report of a Pediatric Case

Moyamoya (a Japanese term, meaning 'hazy things') was first described by Takeuchi in 1963. Two forms of this disease have been distinguished: 1-Primary moyamoya, or moyamoya disease, with a strong hereditary predisposition and girls are more frequently affected. 2-Secondary moyamoya, or moyamoya syndrome, which is caused by a variety of underlying diseases.

The Japanese scientists have classified moyamoya into four types: hemorrhagic, epileptic, infarct, and transient ischemic attack.

Herein, we introduce an 8-years-old girl with the chief complaint of speech disorder. In her physical examination, we detected expressive aphasia and right-sided central facial palsy. After a few days, right hemiplegia and cortical blindness appeared as well. Gradually she was totally unable to move and was transferred to the ICU because of loss of consciousness.

MRI showed diffuse hyper signal lesions in the left temporoparietal and bilateral occipital area. MRA showed narrowing of the internal carotid artery and abnormal collaterals (moyamoya vessels). After indirect bypass surgery (EDAS), she is now able to sit, walk, run and speak.

There are rare angiographically proven moyamoya cases. To our knowledge this was the first EDAS in Iran and a rare case of moyamoya with a dramatic response to operation.

Keywords: moyamoya syndrome, cerebral ischemic attack

Introduction

Moyamoya is a Japanese term, first used by Kudo, [1-3] and it refers to 'hazy things' such as smoke. This condition was first described by Takeuchi in 1963, and later more fully delineated by Suzuki.[1,2,4,5] Takeuchi distinguished two forms: Primary moyamoya, and Secondry moyamoya. Our case was an instance of primary moyamoya that is very rare out of Japan.

Case Report

An 8-year-old girl was referred to the children's neurology center of Mofid Children Hospital with the chief complaint of acute speech disorder on 15 May 2003. This symptom had appeared suddenly at 11 a.m. of the day before. Later at 1 p.m. left facial deviation appeared.

In the past history, she did not have a problematic prenatal, perinatal and postnatal period. Her parents were not relatives, and her motor and mental development was normal.

She weighed 40 kg. In physical examination, she was alert; and in neurological examination, right central facial palsy was detected. The force of her distal right upper extremity was less than that of her left side. Her aphasia was of the expressive type, as she understood speech but could not speak. She was admitted and appropriate investigations were performed. In the course of hospital stay, general total weakness of right upper extremitiy (proximal and distal) was noted.

The results of our evaluation include is summarized as follows:

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*1- **Physical exam**:*
Cardiologist consult: very mild MR
Blood pressure: normal
*2- **Laboratory:***
Chromatography of serum amino acids: normal
The evaluation of collagen vascular disease: negative
Antiphospholipid Antibody: negative
Serum Triglycerid & Cholesterol: normal
Leiden V factor: normal
Hgb electherophoresis : normal
CSF evaluation: normal
Serum Lactate and Ammonia: normal
PCR: negative
3-Neurological Evaluation:
Visual Evoked Potential(VEP) and Audiotory Brain stem Response(ABR) : normal
4- Imaging:
Brain CT: normal
MRI : Left temporoparietal lobe indicated of infarct, significantly hyposignallesion on T1 and hyper signal on T2 weighted(Figure 1)
Angiographic findings:
Evidence of severe stenosis of suprasellar portions of both ICAs.
Bilateral oblitration of MCA and ACA.
vertebro basiliar system: severe stenosis of Right posterior cerebral, Right superior cerebellar and left posterior cerebral arteries: with subsequent hyper trophy and hyperplasia of thalamo striate arteries. (Figure 2) Compensatory hypertrophy and hyperplasia of the lenticostrial arteries (smoke puff).(Figure 3,4)

After 2 weeks, acute visual loss was added to the previous symptoms. Ophthalmoscopy was normal and VEP showed latency: therefore, cortical blindness
was the cause of visual loss. Repeat MRI showed extensive hypersignal lesion in bilateral occipital area with the previous lesion in the left temporoparietal lobe.

In our case, after a few days, the patient lost all of her movement abilities (walking, sitting, neck holding) and gradually a severe dystonia with restlessness appeared.

We transferred the patient to the radiological center for angiography and arteriography, which showed stenotic internal carotid artery and abnormal collaterals or moyamoya vessels. MRA showed the narrow-

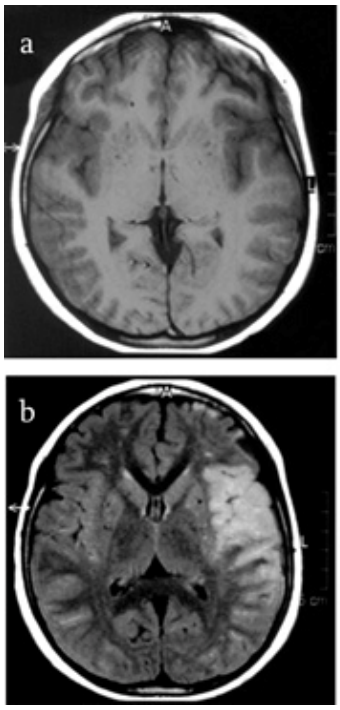

Fig 1. a: axial T1WI, b: axial flair sequence. High signal area in the cortical and subcortical areas of the left temporo-parietal lobes

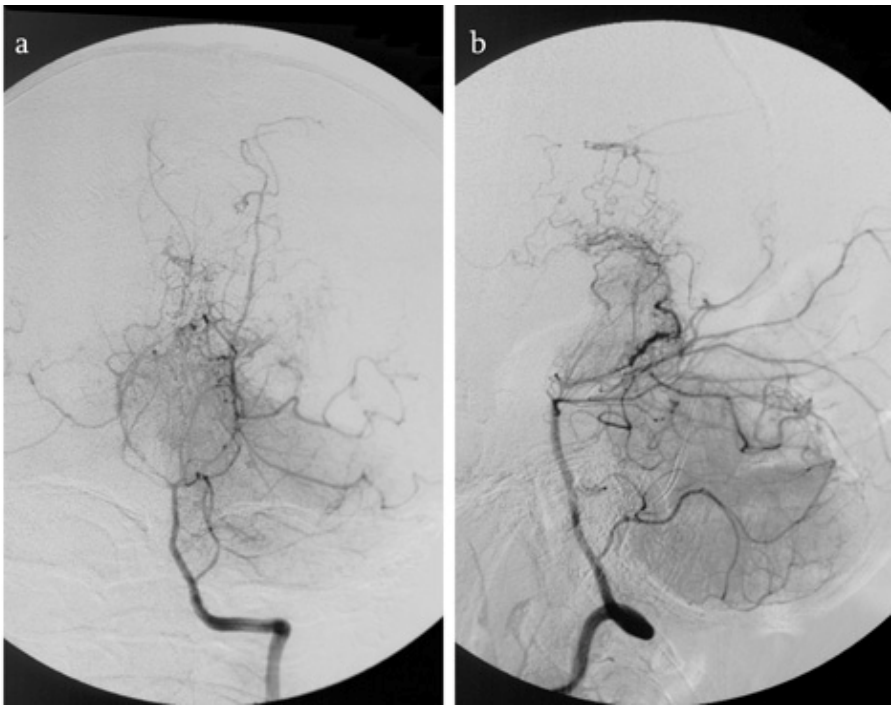

Fig 2. a: AP left vertebral artery, b: lateral left vertebral artery. Compensatory hypertrophy and hyperplasia of the thalamo striate arteries.

ing of internal carotid artery and abnormal collaterals (suspected moyamoya vessels).

Then the patient was admitted to the ICU because of loss of consciousness.

Indirect bypass, encephalo-duro-arterio synangiosis (EDAS), was carried out. Twenty days after the surgery, the patient was alert, and one month later, she had non-verbal communication.

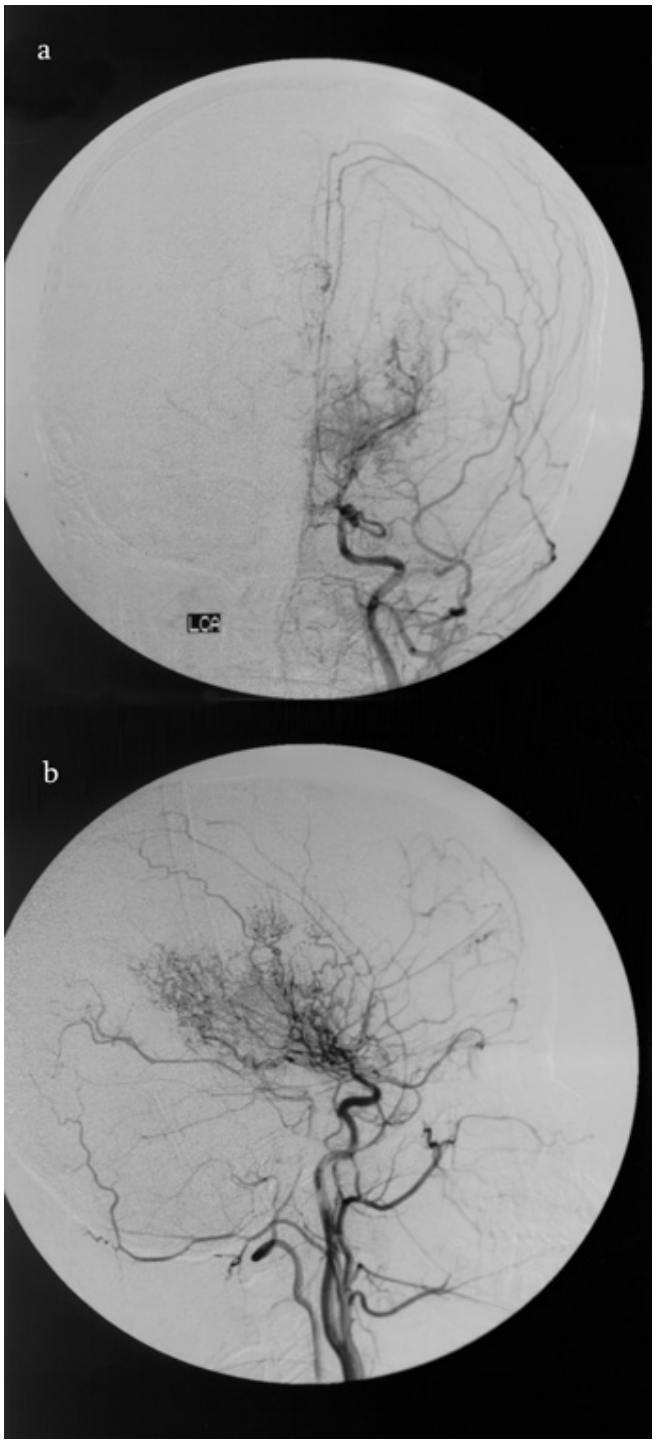

Fig 3. a: Left A.P carotid angiography, b: Left lateral carotid angiography.

Compensatory hypertrophy and hyperplasia of the lenticolo striate arteries (smoke puff).

After 2 months, she could sit and after two courses of surgery (indirect bypass) she could walk and run normally.

Now, she has the ability "of activity of daily living (ADl) ,and she can speak words but she has some behavior disorder that is being treated with behavior therapy. Neurofibromatosis I, sickle cell disease, tuberous sclerosis, Down syndrome, immuno-osseous dysplasia, hypomelanosis Ito, various infections such as tuberculous meningitis or Varicella and AV malformations were ruled out.

Discussion

As noted earlier, there is two types of moyamoya disease: primary and secondary.

Primary moyamoya disease, which has a strong hereditary predisposition, is common among Japanese patients. The incidence is 0.1 in 100000 per year. The gene responsible for this disorder is located on the short arm of chromosome 3. In the pediatric age range, girls more frequently affected, with the peak age of onset being before 5 years of age. [1-3,5,6]

The initial symptoms include motor disturbance, an alternating hemiparesis, transient ischemic attack, speech disturbance, and seizure. Mental deterioration seen in approximately one third of children, and involuntary movements appear in approximately 5%. Unlike in adults, intracranial hemorrhage is unusual in children. Transient ischemic attacks are seen in 20% of children, which can be readily brought on by crying or hyperventilation. Electroencephalograms taken during such events reveal a rapid and marked buildup and a rebuilt of slow waves 20 to 60 seconds after cessation of hyperventilation.[1-3,6]

The Japanese have classified moyamoya disease into four types:

1- The hemorrhagic type characterized by subarachnoid bleeding.

2- The epileptic type with repeated seizures.

3- The infarct type with permanent paresis.

4- The transient ischemic attack type marked by recurrent transient ischemic attacks.

The last type is the most common form seen in Ja-

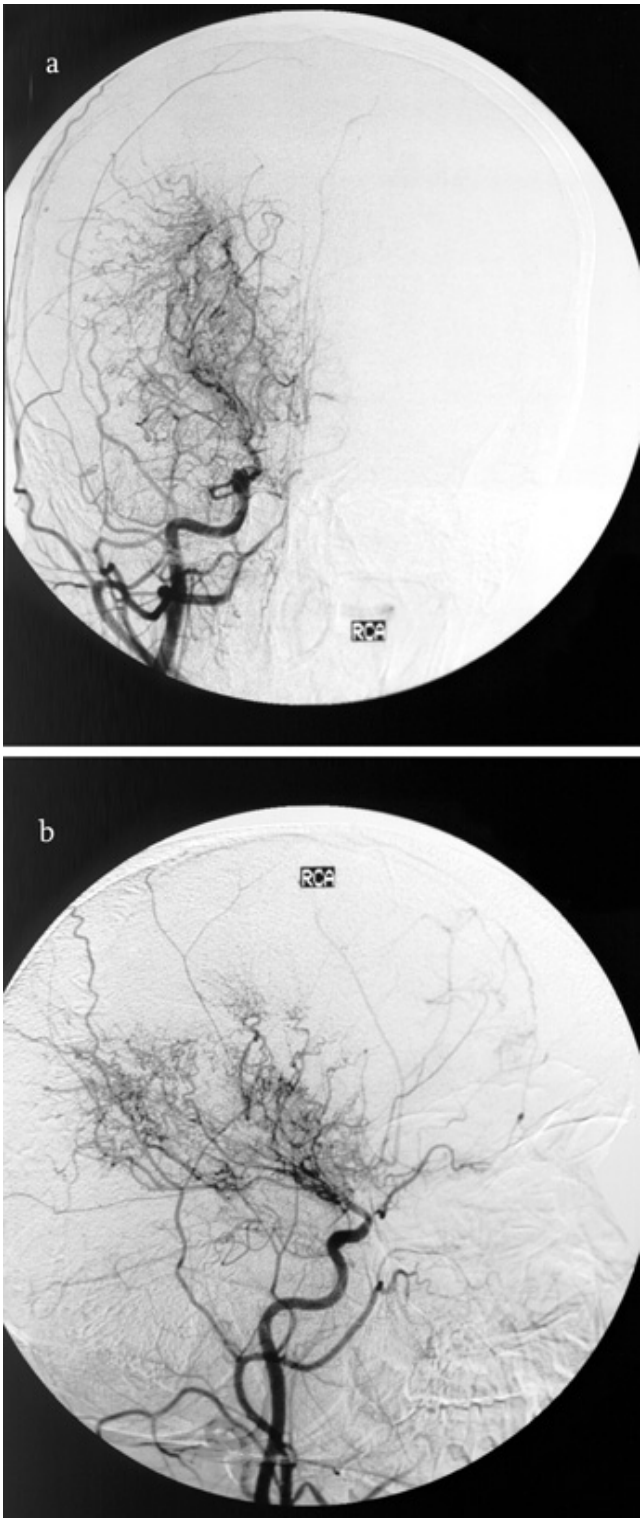

Fig 4. a: Right A.P carotid angiograghy, b: Right lateral carotid angiography.

Compensatory hypertrophy and hyperplasia of the lenticolo striate arteries (smoke puff).

pan. Suzuki has postulated an underlying autoimmune vasculitis as the etiology for primary moyamoya disease.[5]

Angiography or MRA can show several stages of primary moyamoya disease. Narrowing of the carotid arteries is seen in the beginning, followed by dilatation of the major cerebral arteries, and the appearance of collateral circulation (moyamoya vessels). Extensive collaterals can exist between meningeal branches of the external carotid artery and leptomeningeal vessels on the cerebral surface. A prominent collateral network is seen within the basal ganglia.[3]

The reason for the underlying arterial occlusion leading to the development of these extensive collaterals is obscure. Intimal fibrous thickening of the arterial walls of intracranial vessels are common, with similar changes in extracranial vessels.[3]

MRI (magnetic resonance imaging) and MRA (magnetic resonance angiography) are equally informative and permit visualization of the stenotic internal carotid artery and the moyamoya vessels in the basal ganglia.[3] In addition, the areas of infarction are demonstrable by MRI, as early as 2 to 3 hours after the vascular occlusion. Diffusion weighted MRI can be used to evaluate ischemic lesions within minutes after the onset of stroke.[3]

Intellectual deterioration is noted in 65% of children with moyamoya disease of longer than 5 years.[7]

Early onset of symptoms and hypertension are sign of poor prognosis, whereas the presence of seizures is not.

Secondary moyamoya or (moyamoya syndrome) is caused by a variety of underlying conditions. In the of patients seen at the Hospital for Sick Children, Toronto, neurofibromatosis was reported to be the most common cause of moyamoya syndrome (54%). Other causes metioned in literature include sickle cell disease, tuberous sclerosis, Down syndrome, immuneosseous dysplasia, hypomelanosis Ito, various infections such as tuberculous meningitis and varicella. A rare association of AVM in the cerebral hemisphere with moyamoya syndrome may be more than coincidental; the ischemia due to AVM may stimulate the neovascular formation of moyamoya.[8-10]

A variety of extra and intracranial bypass procedures have been proposed for the treatment of moyamoya disease. These procedures produce direct, indirect, or combined anastamotic revascularization.

Direct revascularization includes anastomosis of the superficial temporal artery or the occipital artery to

the middle cerebral artery.

Indirect bypasses, which are more or less effective, include the placement of a dural graft, encephalo-duro-arteriosynangiosis (EDAS) and encephalo-arteriosynangiosis, in which branches of the scalp arteries are used as donor arteries. Other indirect bypasses include encephalo-myosynangiosis (EMS), in which a pedunculated temporalis muscle flap is placed over the temporoparietal lobe. Indirect revascularization is less difficult and is used as the first step in most Japanese centers, and direct anastomosis is reserved for patients whose symptoms persist.

Indirect revascularization surgery for moyamoya disease results in the development of collaterals from the external carotid arterial system into the middle cerebral artery.[14-16] This is associated with a decrease in the abnormal moyamoya vessels, and a significant improvement in children with transient ischemic attacks and involuntary movements. In the exclusively pediatric series of Hoffman, 70% of children treated by EDAS, and sometimes by EMS, had an excellent outcome. In addition, 17% had a generally good outcome but significant neurologic deficits remained.

There had been reports of moyamoya in Iran before, but the diagnosis was never confirmed on angiography. This is the first case of moyamoya in children in Iran, in which indirect operation was carried out for the patient and a dramatic response was seen.

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