

# CASE REPORT

## MOYAMOYA INDUCED ACUTE PARAPLEGIA IN A CHILD WITH EPILEPSY

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### Abstract

#### Objective

Moyamoya disease (MMD) is a chronic, occlusive, cerebrovascular disorder of unknown pathogenesis, characterized by progressive stenosis of the bilateral supraclinoid internal carotid arteries, with concomitant formation of tortuous arterial collateral vessels at the base of the brain, which reconstitute distal branches of the cerebral circulation. In Japanese, "Moyamoya" means "hazy puff of smoke" and refers to the angiographic appearance of the abnormal network of vessels that develop at the base of the brain and basal ganglia to supply a collateral route of blood flow.

We report here the case of Moyamoya disease in a 5 year-old girl with normal mentality with a one year history of epilepsy, with Todd's paralysis. This condition is rare and most patients are diagnosed in childhood. With this report we aim to underscore the possibility that a usual neurological sign could be associated with unusual neurological disorders.

**Keywords:** Moyamoya disease, Todd's paralysis, Epilepsy

### Introduction

Moyamoya disease(MMD) was initially described by Suzuki and Takaku in 1963 as a radiographic phenomenon relating to tiny collateral vessels, characteristic of the disease that resemble a cloud or puff of smoke; it consists of occlusive vascular disease at the events and small strokes. (1, 2) This disease is an idiopathic chronic progressive arterial occlusive disease that affects the distal intra cranial carotid arteries and the vessels arising from the circle of Willis (3). Secondary causes of distal intracranial artery occlusion such as atherosclerosis, vasculitis, and cardiogenic embolus may cause a similar syndrome (moyamoya syndrome). This syndrome also has been associated with neurofibromatosis, sickle cell disease, meningitis, and cranial radiation (4). The disease typically presents with cerebral ischemic symptoms in children and brain hemorrhage in adults (5), and can manifest as hemiparesies, monoparesis, sensory impairment, involuntary movement, headaches, dizziness, or seizures. Mental retardation or persistent neurology defects may be present (6). Etiology of the disease is unknown, and the condition is observed mainly in infants, children and adolescents, and seemingly more in females than in males (7,8). The prevalence of the disease is 0.86 per 100,000 in the United States and about 0.35-3.5 cases per 100,000 in Japan. There is strong evidence of hereditary factors in the disease, with familial cases reported especially among the Japanese, but also Europe and in identical twins (8, 9). Pathologically, MMD is characterized by intimal thickening in the walls of the terminal portions on internal carotid vessels

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bilaterally. The proliferating intima may contain lipid deposits (10,11). On clinical examination, children may have hemiparesies, monoparesis, sensory impairment, involuntary movement, headaches, dizziness, or seizures; mental retardation or persistent neurology defects may be present (6).

Cerebral angiography is the standard criterion for diagnosis, and the following findings support the diagnosis:

- 1- Stenosis or occlusion at the terminal portion of the internal carotid artery or the proximal portion of the anterior or middle cerebral arteries.
- 2- Abnormal vascular networks in the vicinity of the occlusive or stenotic areas.
- 3- Bilaterally of the described findings.

Magnetic resonance angiography (MRA) may preclude the need for conventional angiography (12, 13).

Treatment is directed primarily at complications of the disease. If intracranial hemorrhage has occurred, management of hypertension (if present) is imperative. In case of severe stroke, ICU monitoring is indicated until the patient's condition stabilizes and if the patient has had an ischemic stroke, anticoagulation or an antiplatelet agent should be considered (14).

Various surgical procedures have been used for these patients; those who present for treatment while symptoms are evolving have a better prognosis than those who present with static symptoms (which probably indicate a stroke). Procedures used include:

- 1- Middle cerebral artery (MCA) anastomosis.
- 2- Encephaloduroarteriosynangiosis (EDAS).
- 3- Encephaloduroarteriomyosynangiosis (EDAMS).
- 4- Pial synangiosis
- 5- Omental transplantation (15, 16).

The prognosis of the disease is not good and mortality rates are approximately 10% in adults and 4.3% in children. Death is usually from hemorrhage, and about 50-60% of affected individuals experience a gradual deterioration of cognitive function, presumably from recurrent strokes.

### Case Report

A five year-old girl was admitted in our hospital with a chief complaint of seizures. The patient was the second child of unrelated Iranian parents, with history of seizures

since she was 15 months old after her DPT vaccination. She was delivered at term by normal vaginal delivery with a birth weight of 3000 gr; her mentality was normal. In her 4<sup>th</sup> year of life she experienced seizures with Todd's paralysis, and was treated with Carbamazepine. When the paralysis duration became prolonged, she was brought to the clinic following her last episode of seizure with paraplegia. Physical examination showed the cranial nerve was intact, without loss of consciousness, deep tendon reflexes were increased; Plantar reflex and was downward, and muscle forces were reduced; sensory and motor exams were normal, and the cerebellum was unremarkable.

Laboratory data included: WBC count of 7500 cell/mm<sup>3</sup>, PMNs 35%, lymphocytes 55%, Hb 12.5gr/dl, Hct 38% and platelets 35 x 10<sup>9</sup>/l. ESR 15 mm/hr, CRP(-), Ca 9.1 mg/dl, BS 80 mg/dl, Na 136 meq/l and K 4.1 meq/l.

The patient underwent a CT scan for primary evaluation, which showed cortical opacity in the bilateral frontal lobe parenchyma; an MRI imaging showed ischemic changes (small vessels disease) in the periventricular region and lacunars infarction in both brain hemispheres. With this evidence, suspicions of vascular anomaly increased and the patient underwent MRA/MRI and CT angiography (figures 1&2), which confirmed the diagnosis of moyamoya disease.

### Discussion

In this case the diagnosis was based on past medical history of seizure, clinical features of stroke (paraplegia) & imaging findings. MMD should be considered in any patient with acute symptoms of stroke. This disease usually shows various neurologic signs. In children, the disease occurs in early childhood, around the age of 5 years. The neurological manifestations could be varied, ranging from Transient Ischemic Attack (TIA) to extensive bilateral strokes, seizures, involuntary movements and speech disturbances. Some children may have mental retardation due to long-term ischemia or repeated infarctions. Much data is available on MMD in current literature, in particular the related neurologic problems. This patient presented with clinical features of seizures, signals of bilateral upper motor neuron lesion and paraplegia.

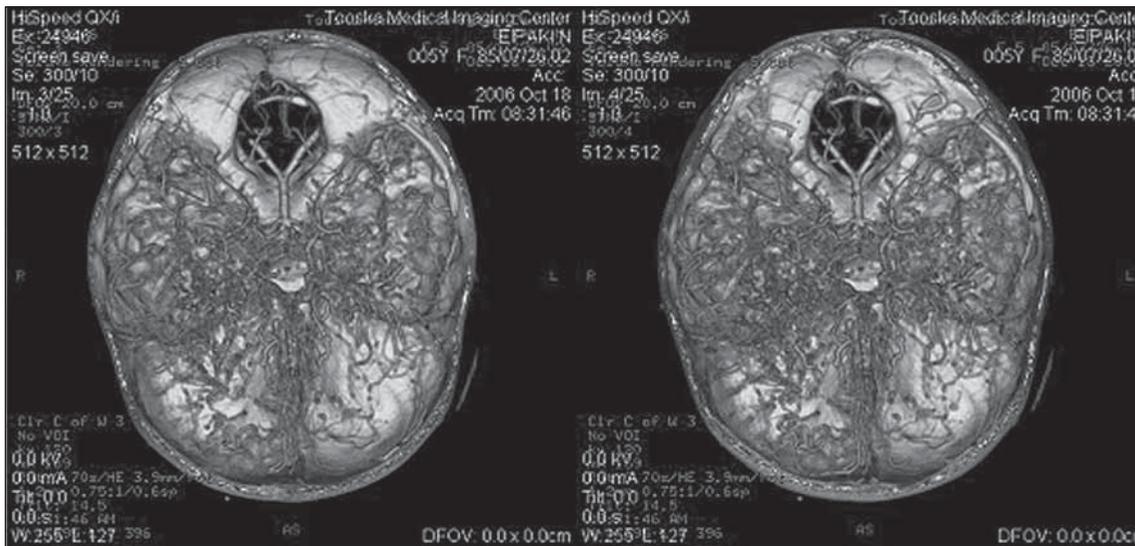


Figure 1: CT angiography of patient showing multiple collaterals of left carotid artery that resemble Moyamoya disease.

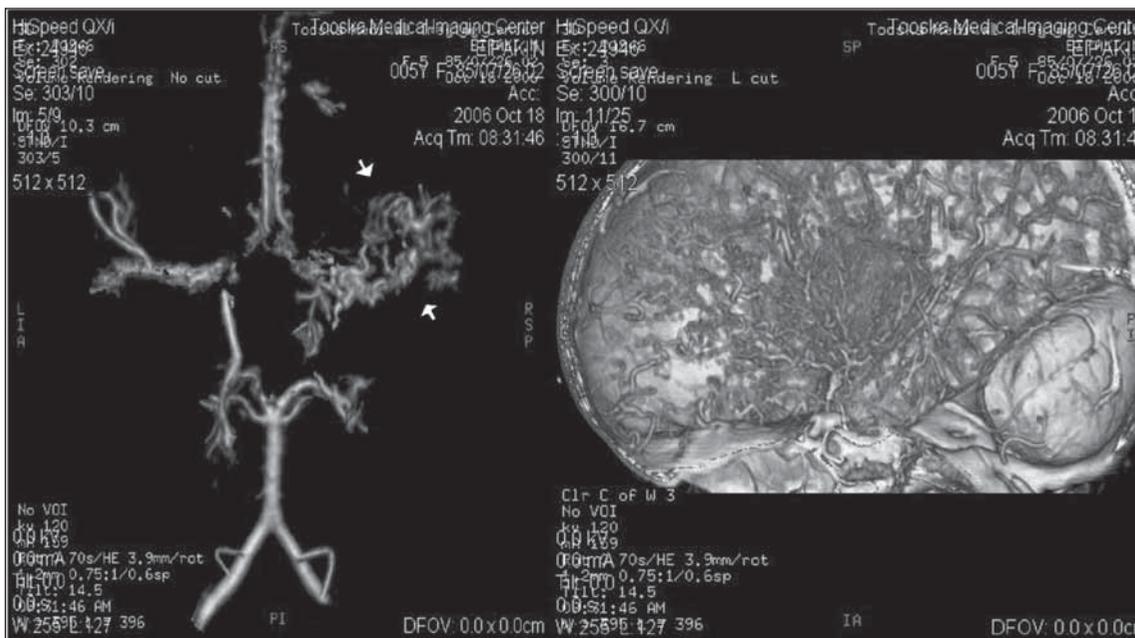


Figure 2: The puff of smoke view, characteristic of Moyamoya disease.

## References

1. Knierim DS, Woodward MW. Moyamoya disease in Hispanic child. *Surgical Neurology* 2002 Jan; 57(1):55-62.
2. Roy Sucholeiki, MD, Jasvinder Chawla, MD. Moyamoya disease. *Emedicine* last update 2006 Nov.
3. Fuki M. Moyamoya disease. *Clinical neurological neurosurgery* 1997;18:232-234.
4. Andrew R, Gregory W, Gary K. Moyamoya syndrome in children with Alagille syndrome. *Pediatrics* 1999; 103: 505-508.
5. Ueki K, Meyer FB, Mellinger JF. Moyamoya disorder and surgical treatment. *Mayo clinical procedure* 1994; 69: 749-757.
6. Hilal SK, Solomon GE, Gold AP, Carter S. primary cerebral arterial occlusive disease in children. *Radiology* 1971 Apr; 81-94.
7. Adams RD. *Cerebrovascular disease. Principles of neurology*, New York. 1993.P. 669-748.
8. Piepgras DG. Moyamoya disease. *Neurosurgery*.

McGraw-Hill Book Company. 1987P.1254-1258.

9. Houlikin K, Nishizaki T, Tshua M. Familial occurrence of moyamoya disease. *Child nerve System* 1994; 10: 421-425.
10. Bruno A, Adams HP, Biller J, Rezai K, Cornell S. Cerebral infarction due to moyamoya disease in young adult. *Stroke* 1988; 19: 826-833.
11. Yamamoto M, Aoyagi M Tajima S. Increase in elastin gene expression and protein synthesis in arterial smooth muscle cells derived from patient with moyamoya disease. *Stroke* 1997; 28: 1733-1738.
12. Tomura N, Ingami A, Higano S. Moyamoya syndrome in young children. *Shinkei* 1988 Oct; 40: 905-912.
13. Yamada I, Suzuki S, Matsushima Y. Moyamoya disease. *Radiology* 1995 Jul;1: 211-218.
14. Momse KJ, New PF. Non-atheromatous stenosis and occlusion of the internal carotid artery and its main branches. *Nuclear medicine* 1973 Jul; 118: 550-566.
15. Matsushima, Inaba Y. moyamoya disease in children and its surgical treatment. *Child brain* 1984; 11: 155-170.
16. Suzuki J, Kodama N. moyamoya disease: the disorder and surgical treatment. *Mayo clinical procedure* 1983; 69: 749-757.