

Moyamoya Disease Presenting as a Seizure Disorder

A Case Report

Bruce S. Schoenberg, MD, MPH; James F. Mellinger, MD; Devera G. Schoenberg, MS; Floyd S. Barringer, MD

• This report of moyamoya disease presenting with vascular headaches and a seizure disorder in a young girl is, to our knowledge, unique in the world literature. The apparent rarity of this condition may reflect the infrequent use of cerebral angiography in investigating such symptoms.

(*Arch Neurol* 34:511-512, 1977)

Moyamoya disease, characterized by stenosis of vessels in and around the Circle of Willis in association with a telangiectatic network of vessels emanating from the base of the brain, was first observed among the Japanese. The name of the condition is derived from "a Japanese expression for something hazy just like a puff of cigarette smoke drifting in the air," which describes the angiographic appearance of the abnormal network of vessels. Childhood moyamoya disease presenting with seizures as the initial symptom is a rare phenomenon. Only three such

cases could be found in a review of published papers.²⁻⁴ The child described in this report, in whom a seizure disorder and vascular-type headaches were the only neurologic manifestations of proved moyamoya disease, is, to our knowledge, unique in the world literature.

REPORT OF A CASE

This 7½-year-old white girl was evaluated for recurrent stereotyped spells of fright and staring, followed by right hemiparesis and aphasia.

Past medical history showed that the pregnancy and delivery were normal, although the patient appeared to be slightly premature at birth. She had no serious illnesses, but suffered from frequent nasal discharge and sore throats, necessitating a tonsillectomy at the age of 3½ years. She had a history of mild headaches, particularly when riding in a car. Other than these minor medical problems, the patient enjoyed good health until August 1974. At that time, after awakening from a nap, she appeared to be glassy-eyed and was unable to speak. She was also noted to have a right hemiparesis. Her speech returned to normal in a few minutes, and she complained of severe left-sided head pain. Both the headache and hemiparesis resolved in a few hours. Studies done at this time were negative except for an EEG that showed left-sided occipital slowing and the funduscopic finding of tortuous and dilated retinal vessels. Her physician prescribed ethosuximide, 250 mg orally, twice daily. However, the patient continued to have brief episodes (lasting one to two minutes) of fright, staring, slight right-sided hemiparesis, and a peculiar distant feeling, occurring mostly at night. In January 1976,

she began experiencing vascular-type headaches. These were most severe over the left eye, commonly occurred either early in the morning or after awakening from a nap, and were often associated with nausea and vomiting.

On March 9, 1976, the patient, after having a few minor spells, underwent a second EEG. It was during this EEG that she experienced a right-sided focal seizure, with both sensory and motor symptoms, immediately followed by a flaccid paralysis of the right upper extremity and aphasia. The EEG showed generalized severe slowing, greatest in the left hemisphere, but no spike activity.

The patient continued having seizures, characterized by severe headache at the onset, followed by a feeling of strangeness, crying, hemiparesis of the right upper extremity, and slight right facial droop. Such episodes lasted about three minutes. No abnormal motor movements, incontinence, or complete loss of consciousness were associated with these spells. Phen妥in (50 mg orally, three times daily) was prescribed.

A computer-assisted transaxial tomogram and a lumbar puncture performed in late March 1976 were negative. However, bilateral carotid angiograms showed multiple segmental vascular occlusions in the upper carotid siphon bilaterally, with telangiectatic anastomoses. There was also severe stenosis and probable occlusion of the supraclinoid portion of the right internal carotid artery. Florid collateral circulation in the region of the basal ganglia, with partial distal filling of branches of the middle cerebral arteries was also apparent. These findings were compatible with moyamoya disease. Further review of the angiograms showed an anastomotic network of vessels in the orbit with an enlarged right ophthalmic artery. This

Accepted for publication March 31, 1977.

From the National Institutes of Neurological and Communicative Disorders and Stroke, Bethesda, Md (Dr Schoenberg), the Department of Neurology (Dr Schoenberg), Georgetown University School of Medicine, Washington, DC, and the Department of Neurology (Drs Schoenberg and Mellinger), Mayo Clinic, Rochester, Minn.

Reprint requests to Section on Epidemiology, Office of Biometry and Epidemiology, Office of the Director, National Institute of Neurological and Communicative Disorders and Stroke—National Institutes of Health, Room 7C10, Federal Bldg, 7550 Wisconsin Ave, Bethesda, MD 20014 (Dr Schoenberg).

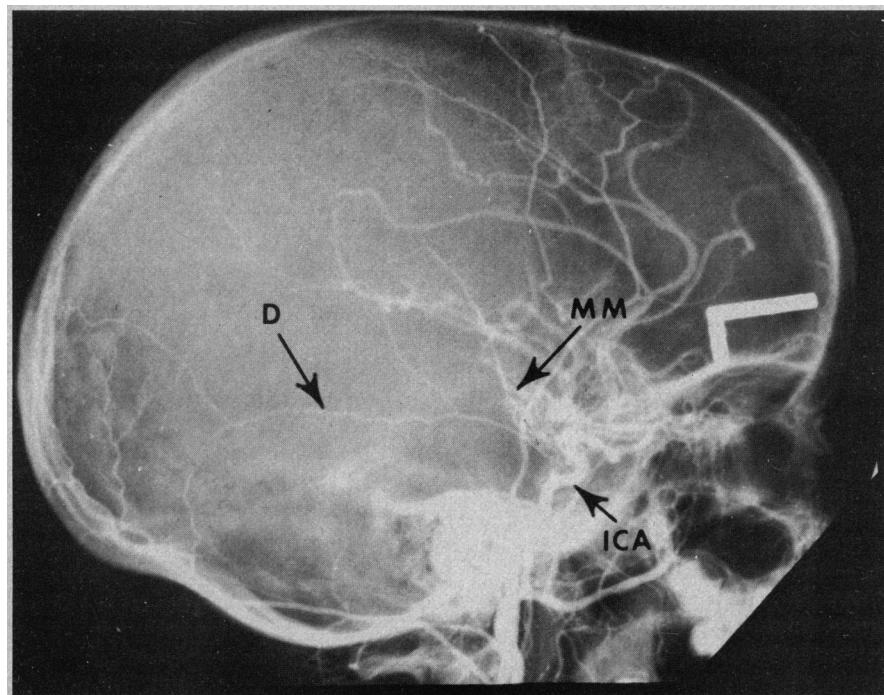
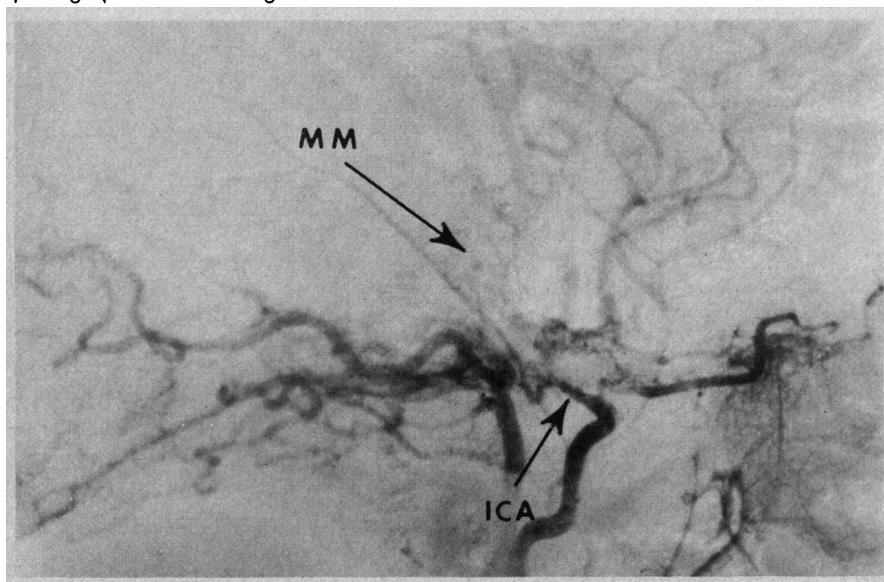


Fig 1.—Lateral projection of right carotid arteriogram. ICA indicates internal carotid artery; MM, telangiectatic network of vessels characteristic of moyamoya disease; and D, dural vessel filling from external carotid system.

Fig 2.—Lateral projection (subtraction view) of right carotid arteriogram. Note stenosis of internal carotid artery (ICA) and fine telangiectatic network of vessels (MM). Portion of photograph to reader's right is anterior.



artery was probably supplying a small vascular network in the region of the cribriform plate. Details of the right carotid angiogram are illustrated in Fig 1 and 2. It is unusual to see the dural vessel noted in Fig 1 this early in the sequence of a normal arteriogram. The early appearance of such a vessel is compatible with the high grade of stenosis of the internal carotid artery shown in Fig 2.

Findings of the general physical and neurological examination performed in mid-April 1976 were normal except for marked dilation and tortuosity of the retinal blood vessels detected by funduscopy. Moreover, the patient has had no further seizures since phenytoin therapy was initiated.

COMMENT

Moyamoya disease has only recently been recognized as a clinical entity. It may manifest itself in a variety of ways, with symptoms of either ischemic or hemorrhagic cerebrovascular disease. The case reported here is unique in that the only neurologic manifestations are a seizure disorder and vascular headaches. It may be that the seizures have some origin other than the cerebrovascular disease, but this possibility appears to be unlikely.

Cerebral angiography is the only currently available method of diagnosing moyamoya disease in the living patient. The apparent rarity of this condition as suggested by the paucity of case reports may simply reflect the relatively infrequent use of angiography to investigate cerebrovascular disease in children.

On the basis of this report, one must consider moyamoya disease as a possible cause of seizures presenting in childhood. The probability of this phenomenon must await further clinical experience with this disorder.

References

1. Suzuki J, Takaku A: Cerebrovascular "moyamoya" disease: Disease showing abnormal net-like vessels in base of brain. *Arch Neurol* 20:288-299, 1969.
2. Hately W, Shapiro R: Carotid rete mirabile: An unusual example associated with diffuse bilateral cerebral telangiectasia. *Clin Radiol* 20:32-35, 1969.
3. Solomon GE, Hilal SK, Gold AP, et al: Natural history of acute hemiplegia of childhood. *Brain* 93:102-120, 1970.
4. Hilal SK, Solomon GE, Gold AP, et al: Primary cerebral arterial occlusive disease in children: I. Acute acquired hemiplegia. *Radiology* 99:71-86, 93-94, 1971.