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MOYAMOYA DISEASE AND ALTERNATING HEMIPLEGIA
A REPORT OF TWO CASES*

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Moyamoya disease is one of the cerebrovascular disorders and is characterized by bilateral stenosis or occlusion of the distal internal carotid arteries and their main branches. Repeated ischemic episodes in children and intracranial hemorrhage in adults with moyamoya disease is usually noted. In this report, we describe two children with moyamoya disease who presented with alternating hemiplegia. The final diagnosis was made by cerebral angiography and cranial computed tomography in these patients. Other neurological and radiological features of the patients were described. *Key words: moyamoya disease, alternating hemiplegia.*

Moyamoya disease is the angiographic diagnosis of one of the cerebrovascular disorders. It is characterized by bilateral stenosis or occlusion of the distal internal carotid arteries and their major branches, resulting in development of an abnormal vascular network[1-3]. The disorder has a worldwide distribution. In children, the initial symptoms consist of repeated ischemic episodes leading to various degrees of permanent neurological deficit, whereas adults usually present with intracranial hemorrhage. Although the underlying cause has not been identified, a generalized immunological arteritis, especially in those arteries innervated from the superior cervical sympathetic ganglia, is under speculation[4].

In this report we present two children found to have moyamoya disease and discuss the nature of the disease as seen in childhood.

Case Reports

Case 1

A 5-year-old boy had developed left hemiparesis and facial asymmetry when admitted to the hospital. Two similar attacks of alternating hemiplegia, the first one a year ago and the second 16 days ago, had occurred and resolved without

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any neurological sequelae. He was the first child born to healthy parents and had an unremarkable past history except a mild head trauma at two years of age. On admission he was alert and cooperative but had nonfluent speech and left central facial paralysis. Motor examination showed left-sided hemiparesis, hyporeflexia and left plantar extensor response.

Laboratory studies including complete blood count, blood and urine amino acids, electrocardiography, chest x-ray, tests for collagen vascular disease; blood urea nitrogen, creatinine, serum transaminase activities, tests for infectious diseases and coagulation disorders, were all in normal range. Electroencephalography (EEG) showed diffuse slow wave activity in the right hemisphere with slow sharp wave activity in the right frontotemporal region. Cranial computed tomography (cranial CT) revealed multiple low-density areas in the right temporal, parietal and left parietal regions consistent with cerebral infarctions.

^{99m} THMPAO single photon emission computed tomography (SPECT) study showed irregular hypoperfusion areas in the bilateral frontal, right temporal, parietal and right cerebellar regions. On digital substruction angiography (DSA) the patient had bilateral stenotic lesions in the syphon and dilated collaterals in basal levels. Bilateral middle and anterior cerebral arteries were filled via the posterior communicating artery from the vertebro-basillary system. The right internal carotid artery was filled with the transdural collaterals from the external carotid circulation. During the hospital stay the hemiparesis resolved and his speech returned to normal and the patient was discharged on acetyl salicylic acid and diphenylhydantoin.

Case 2

An 11-year-old boy came to the hospital with right-sided paralysis and aphasia. Two days before he had complained about not being well and had had slurred speech. His symptoms progressed rapidly, and finally he was unable to walk and talk. His past and family history were unremarkable.

On physical examination, he was alert, moderately cooperative and mildly hypotonic. His ability to name objects and repeat words was impaired, thus giving the impression of mild mental retardation. He had right central facial paralysis, right sided hemiplegia, hypoactive tendon reflexes and a bilateral positive Babinski sign. The rest of the examination was normal.

Laboratory examination: The routine work-up done for the first patient was also carried out for this patient and revealed no abnormality except bilateral maxillary sinusitis. EEG showed disturbed basal activity and voltage suppression and asymmetry of the two hemispheres. Cranial CT revealed an area of hemorrhagic infarct in the left frontal lobe.

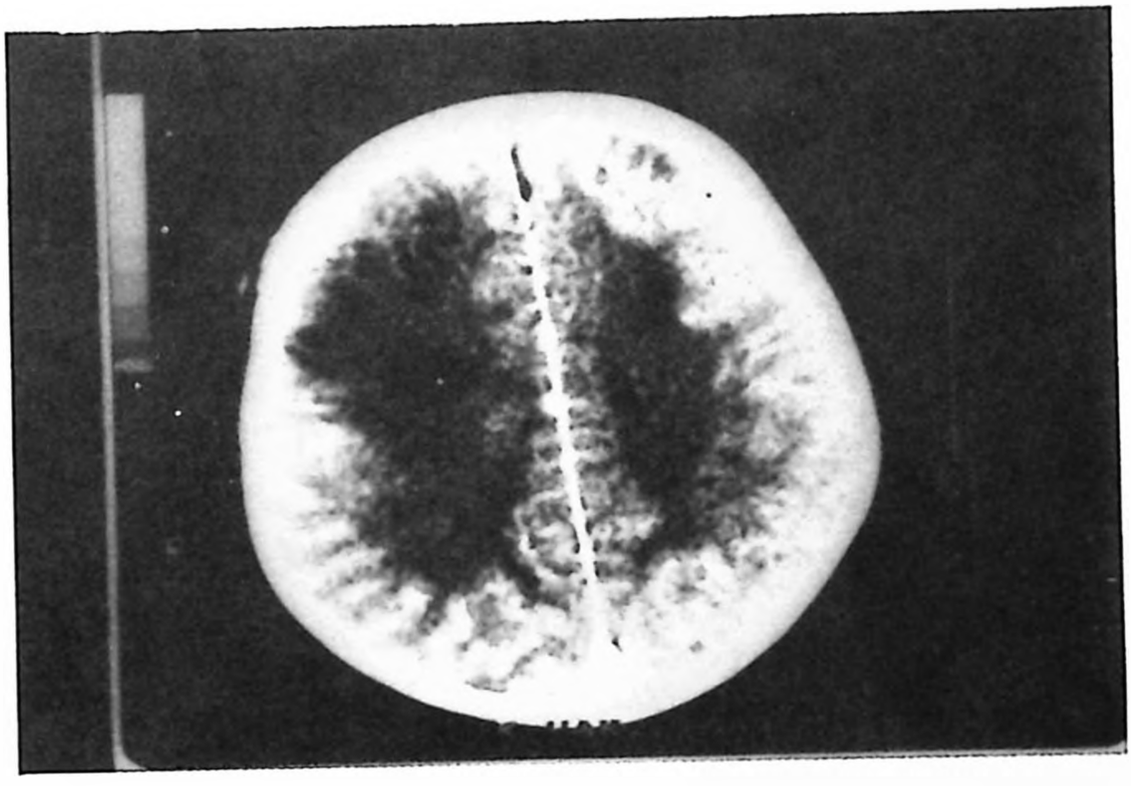

Fig. 1 a: CT of Case 1: Multiple low-density areas in the right temporal-parietal and left parietal regions.

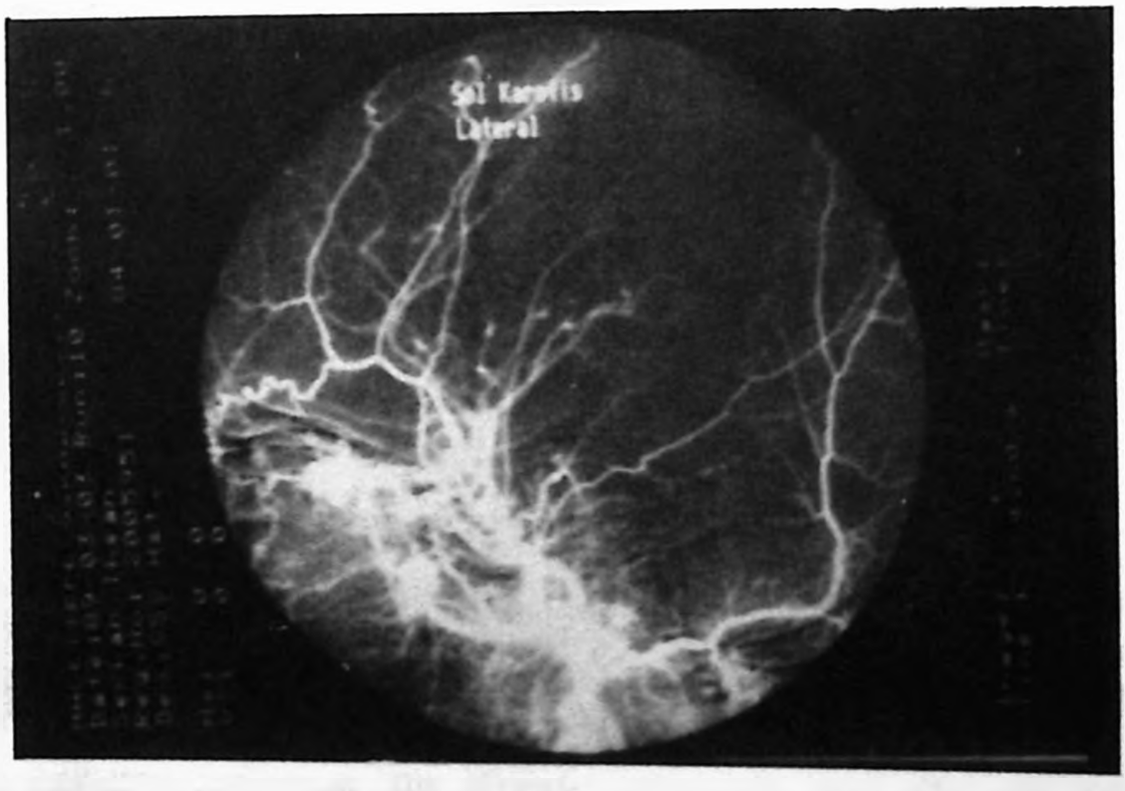

Fig. 1 b: Angiography of Case 1: Total occlusion at the cavernous portion of internal carotid artery.

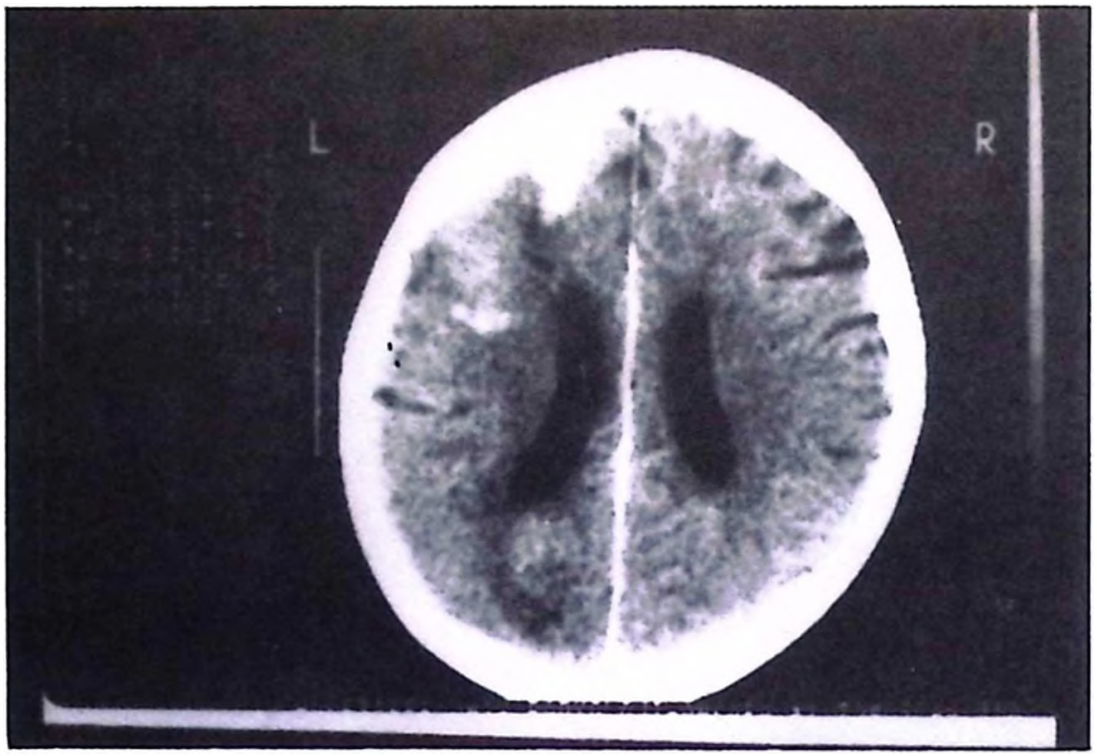

Fig. 1 c: CT of Case 2: Hemorrhagic infarct in the left frontal lobe.

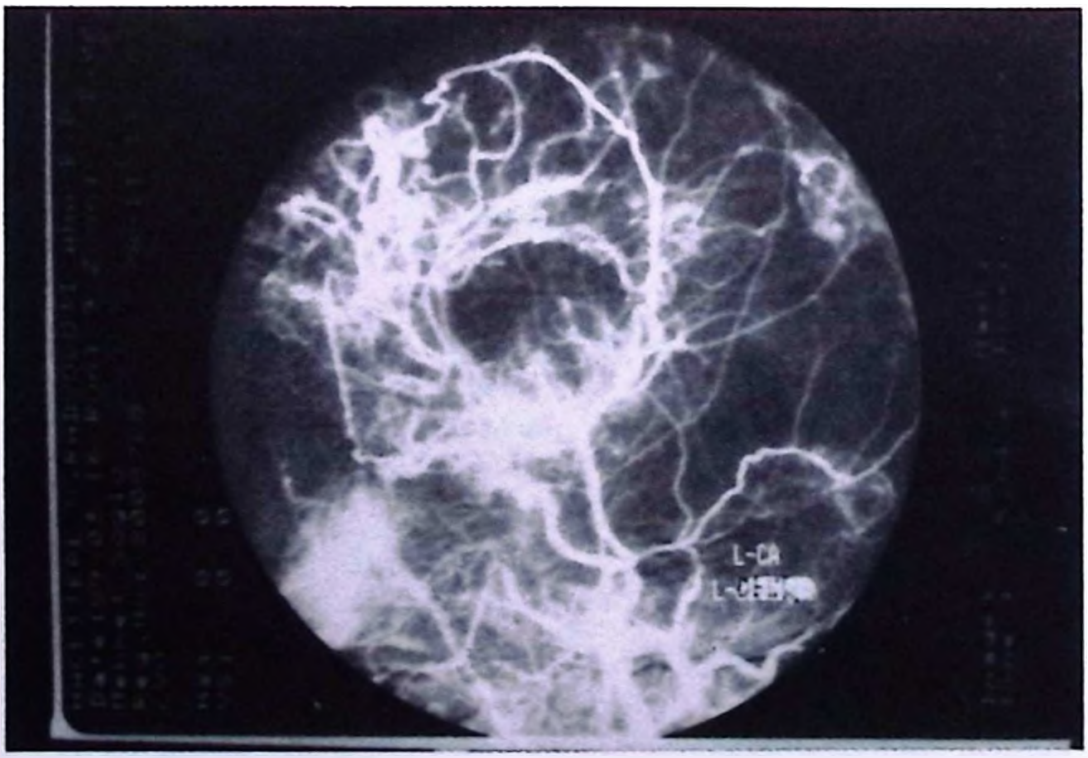

Fig. 1 d: Angiography of Case 2: Total occlusion at the bifurcation of internal carotid artery and basal collaterals.

On cerebral angiography, bilateral obliteration of the internal carotid arteries after the syphon and typical dilated collateral basal vessels were shown. Perfusion was maintained by the orbital and transdural collaterals from the external carotid circulation. In the vertebro-basillar system, the right middle cerebral artery was filled by the enlarged right posterior communicant artery, and the blood supply to the cerebellum was maintained by the collaterals developed from the posterior cerebral arteries. In the follow-up period, hemiparesis markedly improved.

Discussion

Moyamoya disease is a chronic occlusive cerebrovascular disorder characterized by the angiographic features of:

1. Stenosis primarily involving the region of the internal carotid artery bifurcation.
2. Presence of dilated basal collaterals.
3. Bilateral abnormalities[2,5].

If these features are found in association with any of the conditions listed in Table I a diagnosis of "moyamoya syndrome" is appropriate. When the angiographic pattern is limited to one side only without the above conditions a "probable moyamoya disease" should be considered[5]. This situation is known to be rare and is mainly caused by spontaneous occlusion of middle cerebral and internal carotid arteries.

Table I: Conditions Associated with Moyamoya Disease

Neurofibromatosis	Fanconi's anemia
Tuberosclerosis	Down's syndrome
Encephalotrigeminal angiomatosis	Cerebral glioma
Fibromuscular dysplasia	Congenital heart disease
Vasculitis	Hypertension
Tuberculous meningitis	Type I glycogenosis
Radiation - induced arteritis	Myopathy
Sickle cell anemia	

Although moyamoya disease has been described most often in the Japanese poulation, with an estimated incidence of 1:100.000[2], many sporadic cases have been reported in other parts of the world[6-9], with a female predominance. There is no definite evidence of genetic determination[10].

In the pediatric age-group, the disease commonly presents with repeated ischemic episodes and is one of the important causes of acute hemiplegia and focal neurological deficits[2,4,7]. The hemiplegia may alternate between the right

and left sides. Mental and sensory disturbances may occur. Headache and seizures are also described during and between the ischemic episodes. In children, by serial angiographic studies, the disease is shown to be progressive with respect to the extent of the occlusion[1,2,10]. When the occlusive process has stopped, the maximum number of collateral vessels have been developed and the clnical course is stabilized. In a study of 39 cases of pediatric patients, the fatality rate was 13% and 77% of the patients had developed significant neurological deficits. Only 23% of the living patients had remained neurologically intact despite repeated ischemic attacks[10]. If the onset is before the age of four, rapid deterioration and intellectual regression with irreversible neurological deficits develop[4,7,10]. In adults, intracranial hemorrhage is the prevailing presentation[6,10,11] but cerebral infarctions may also be seen[5,8].

A generalized immunological arteritis manifesting in the arteries which are innervated by the superior cervical sympathetic ganglia has been suggested as an etiologic factor. The high incidence of tonsillitis, otitis media and other infections above the neck supports the theory that there may be a connection between such infections and the arteritis[7]. One of our patients had bilateral maxillary sinusitis. The pathological studies have demonstrated endothelial hyperplasia and fibrosis with no associated inflammatory reaction while the tunica media and adventitia have remained intact[1,4,10]. The association of moyamoya disease with cerebral aneurysms has been defined[12] but the combination with arterio-venous malformations is rare and coincidental[13,14]. Cerebral angiography is needed to confirm the diagnosis of moyamoya disease and six stages in the angiographic course of the disease in children have been defined[1,2]. Our patients' angiographic appearances were consistent with the disease.

Cranial CT studies demonstrated multiple and bilateral infarctions in our patients as reported in the literature[5,7,15,14]. By SPECT, in one of our patients we noted frontal hypoperfusion which could be highly suggestive of moyamoya disease. As in our patients, EEG may show increased slow wave activity and focal abnormalities without any significance. Magnetic Resonance Imaging studies have shown evidence of large and small infarcts and can also detect slow blood flow within the middle cerebral artery in severe cases[16]. In a recent study, a significant correlation between the neurological deficit and abnormalities in evoked potentials was shown[17].

The medical treatment of moyamoya disease has been unsuccessful[7]. However, a number of surgical procedures such as by-pass of the stenotic arteries and cervical perivascular ganglionectomy in order to improve perfusion and prevent neurological deterioration have been recommended[7,10].

In any child presenting with a transient ischemic episode, moyamoya disease should be considered in the differential diagnosis.

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