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COMPUTED TOMOGRAPHY AND ANGIOGRAPHIC FINDINGS OF CHILDHOOD MOYAMOYA DISEASE*

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Moyamoya disease is a rare entity consisting of bilateral stenosis or occlusion of internal carotid arteries with abnormal collateral vessels at the base of the brain. We present five Turkish children with this disease, which is more common in Japan.

Focal neurologic deficits and/or epilepsy were the common presenting symptoms in two girls and three boys between the ages of 1.5 and 11 years. Multiple cerebral infarcts were diagnosed in all of the cases by Computed Tomography (CT). Abnormal net-like vessels at the base of the brain were detected in three patients. Cerebral angiography, which is necessary to confirm the diagnosis, showed moyamoya vessels and bilateral stenosis or occlusion of the internal carotid arteries in all cases. Although the angiographic staging was advanced in three patients, neither clinical status nor parenchymal abnormalities detected with CT were different from the other two cases. *Key words: carotid arteries, brain, moyamoya disease.*

Moyamoya is the descriptive term used for angiographic picture consisting of abnormal net-like vessels (ANV) at the base of the brain[1]. Bilateral stenosis or occlusion of the terminal parts of the internal carotid arteries (ICA) along with ANV specific for moyamoya disease. Although the disease has been described in Western literature, the Japanese are most frequently affected, with estimated cases numbering more than 2000[2].

Angiography is needed to confirm the diagnosis[1,3-7]. But other imaging modalities such as Computed Tomography (CT)[3] and Magnetic Resonance Imaging (MRI)[4,5] may suggest moyamoya disease.

Material and Methods

Between the years 1988 and 1990, two girls and three boys between the ages of 1.5 and 11 years were referred for cerebral angiography and were diagnosed with

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moyamoya disease. The cases are summarized in Table I. In two of our cases, upper respiratory tract infections appeared first, followed by hemiplegia, which necessitated evaluation of the patients.

Table I: Clinical Data of Patients With Childhood Moyamoya Disease.

	Age (Years)	Sex	Symptoms
Case 1	1.5	F	Focal motor epilepsy, right hemiparesis
Case 2	4	M	Epilepsy
Case 3	5	M	Alternating right and left hemiplegia and recovery within 10 months
Case 4	8	F	Focal motor epilepsy, right hemiplegia
Case 5	11	M	Mental and motor retardation, right hemiplegia

All of the patients had at least one CT examination before angiography. CT examinations were done at different centers. They were taken in the axial plane and the slice thickness was either 9 or 10 mm.

Cerebral angiography was performed by Digital Subtraction Angiography (DSA) technique using Digital Vascular Imaging (Philips, the Netherlands). All examinations were performed under general anesthesia. A transfemoral approach with a 5.0 F straight or 45 degree-curved visceral catheter was used. Selective cathetherization of both carotid and vertebral arteries was successfully performed except on the youngest patient, and anteroposterior, oblique and lateral projections were obtained in every patient. Iohexol (300 mgI/ml) was diluted by the same amount of saline and manually injected. The total dose of the contrast medium did not exceed 2 ml/kg.

Results

CT and DSA findings are listed in Table II. Suzuki and Kodama[7] gave two classifications of moyamoya disease: ethmoidal moyamoya and vault moyamoya, depending on the location of ANV. Satoh et al[6] suggested a new staging system based on the progress of vessel stenosis, which seems more accurate for therapy planning and follow-up: Stage 1: Slight to moderate stenosis of ICA bifurcation; Stage 2: Severe stenosis of ICA bifurcation; Stage 3: Occlusion of anterior or middle cerebral arteries; Stage: 4: Occlusion of ICA or anterior and middle cerebral arteries with partial retention of peripheral artery's trunk (Fig. 1); and

Table II: CT and DSA Results

	CT	DSA	Stage
Case 1	Bilateral parietal infarcts	R-ICA stenosis, L-ICA occlusion, ANV	2
Case 2	Cerebral atrophy Bilateral cortical infarcts ANV	Bilateral ICA stenosis, MCA, ACA, PCA stenosis, ANV	3
Case 3	Multiple frontal infarcts	Bilateral ICA occlusion, partial retention of MCA and ACA trunks, ANV	4
Case 4	Multiple cortical infarcts in left cerebral hemisphere ANV	Bilateral ICA occlusion, Partial retention of MCA and ACA trunks, ANV	4
Case 5	Cerebral atrophy Left frontal infarct ANV	Bilateral ICA occlusion, Partial retention of MCA and ACA trunks, ANV	4

(ICA: Internal carotid artery, ANV: Abnormal net-like vessels, MCA: Middle cerebral artery, ACA: Anterior cerebral artery, PCA: Posterior cerebral artery).

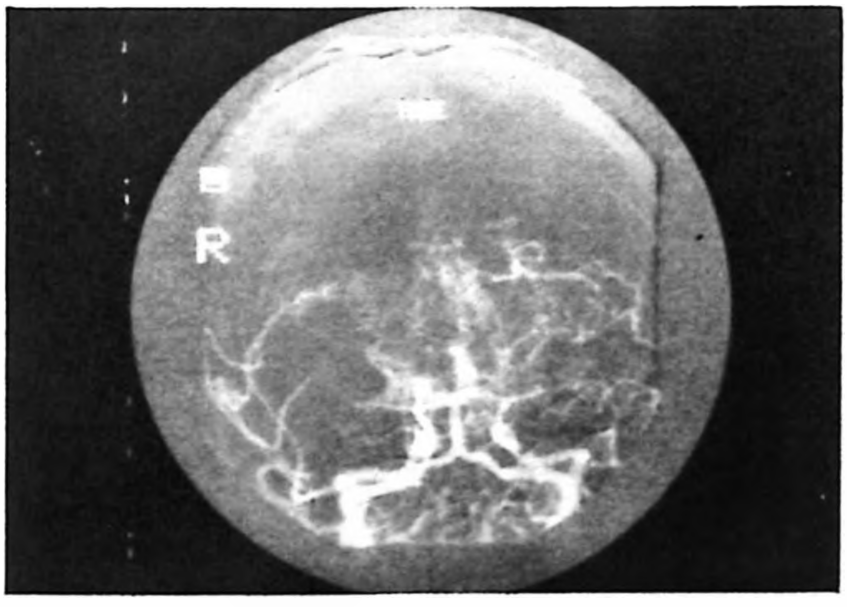

Fig. 1: Case 3, A-P cerebral DSA showing bilateral stenosis at ICA bifurcation, stenosis and occlusion of ACA and MCA along with abnormal net-like vessels at the base of the brain.

Stage 5: Occlusion of ICA or anterior and middle cerebral arteries with no main trunk. We used Satoh's staging system in our cases, as shown in Table II.

ANV were present in all our cases. ANV at the base of the brain fed by ICA and/or the posterior cerebral artery was the most common location (Fig. 2). Ethmoidal and ophthalmic collaterals and collaterals arising from the external carotid artery were less common in our patients.

Aneurysm and subarachnoid hemorrhage incidence is high in the adult form of moyamoya disease[2,7]. No aneurysm was found in our cases.

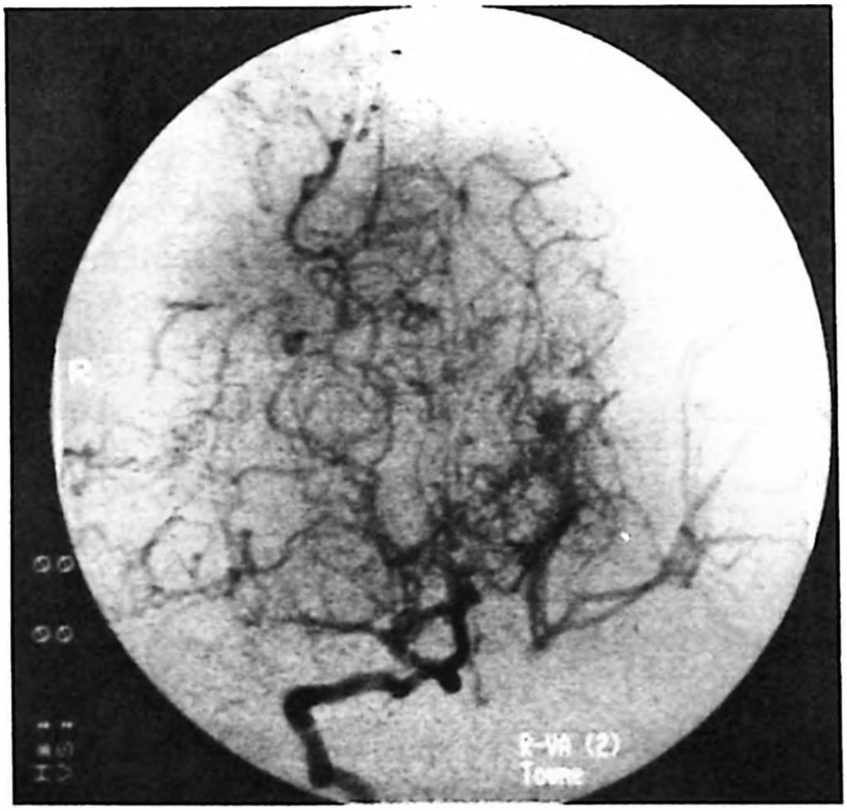

Fig. 2: Case 4, R-Vertebral artery arteriography showing abnormal net-like vessels fed by posterior cerebral arteries. On the right side, vault moyamoya vessels could also be seen.

Discussion

Moyamoya, a Japanese word meaning "something hazy like a puff of cigarette smoke drifting in the air" is the descriptive term applied to abnormal net-like collaterals seen in cerebral angiography[7]. In addition to ANV, stenosis or occlusion of the ICA at the level of its terminal bifurcation, with bilateral but asymmetric stenosis of anterior and middle cerebral arteries is a common finding. Its etiology is still unknown. Most authors consider the carotid stenosis to be the primary condition and ANV to be the result of it[7]. Basal meningitis, tumor, radiation therapy, neurofibromatosis, atherosclerosis, connective tissue abnormalities, sickle cell disease, Fanconi's anemia and congenital spherocytosis can cause ICA

stenosis and development of ANV[7,8]. But familial incidence (7%)[9], a report of monovular twins[10] and other infantile cases[10] suggest a genetic factor. Exclusion of the above-mentioned possible causes of "acquired" moyamoya disease is necessary for the final diagnosis.

Children with moyamoya disease usually present with repeated cerebral ischemic attacks (hemiplegia, paresthesia, convulsions). Hemiplegia may be reversible and alternating. If left untreated, cerebral atrophy and mental motor retardation develop, as in our oldest patient.

Usually CT is performed first when evaluating a child with cerebral ischemic symptoms. The major CT feature of moyamoya disease is cerebral atrophy and focal areas of low density in the basal ganglia or cerebral cortices representing infacrcts (Fig. 3). The curvilinear tortuous vascular densities correspond to the parenchymal, dural and leptomeningeal collaterals seen on cerebral angiography[3]. In three of our cases, these ANV could be seen around basal ganglia (Fig. 4). MRI has the great advantage of contrast medium[5]. Although MRI is expensive and not readily available, it must be used in patient follow-up because angiography has a high incidence of morbidity, especially in children[4].

Superficial temporal artery-middle cerebral artery by-pass, which was introduced by Yaşargil, has become an established mode of treatment for both childhood and adult moyamoya disease[11]. Although by-pass surgery helps to maintain cerebral blood flow, childhood moyamoya disease usually progresses, and the incidence of intracranial hemorrhage increases with age[6].

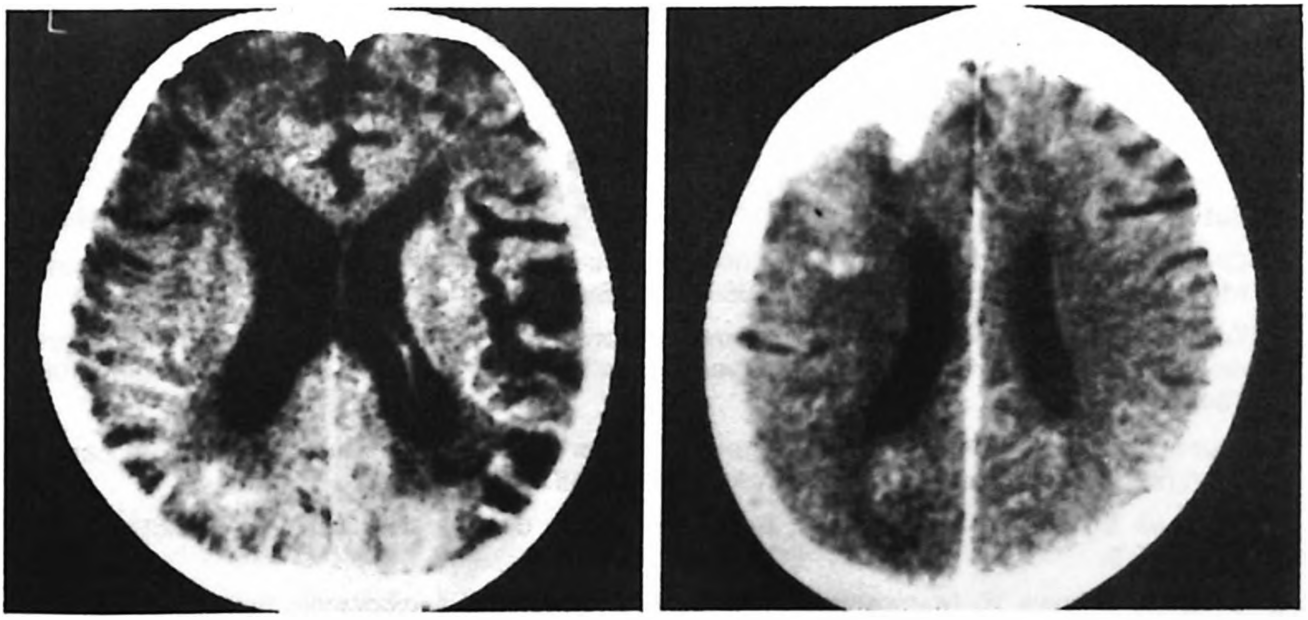

Fig. 3: a) CT of Case 2: Cerebral atrophy and multiple cortical infarcts. b) CT of Case 3: Right frontal sub-acute cortical infarcts enhanced with contrast medium.

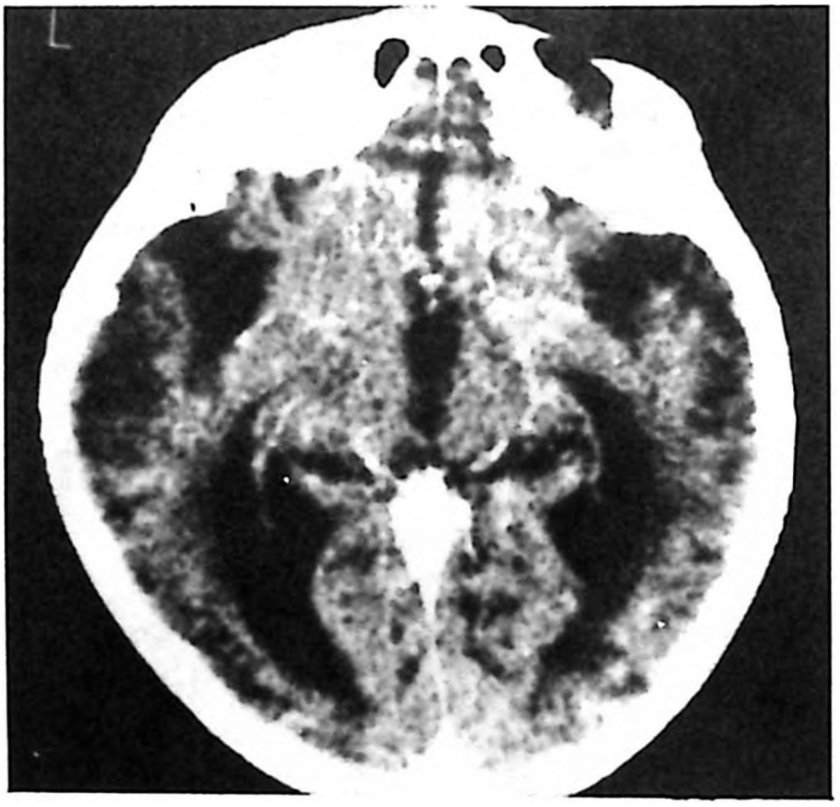

Fig. 4: Case 5: CT showed linear vessels in basal ganglia representing abnormal net-like vessels. Note dilated temporal horns and subarachnoid space secondary to cerebral atrophy.

Recurrent and alternating hemiplegia in a child must raise the suspicion of moyamoya disease. Cerebral atrophy, multiple infarcts and collateral vessels seen on CT are highly suggestive of the disease. Selective cerebral angiography is needed to confirm the diagnosis and planning for surgical intervention. Once the diagnosis is established, MRI is the best method for follow-up.

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