RADIOLOGICAL FINDINGS IN MOYAMOYA DISEASE

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ABSTRACT

The clinical, angiographic and computed tomographic features in eight children with Moyamoya disease were evaluated. The CT Scan findings were correlated with the angiographic features and the stage of the vascular disease. Stenosis/occlusion of the supraclinoid internal carotid artery (ICA) and the proximal parts of the anterior (ACA) and middle cerebral arteries (MCA) were the commonest angiographic findings". The cervical ICA was narrow in four patients, lnfarcts (100%), abnormal enhancement patterns (63%) and cerebral atrophy (88%) were the frequent CT scan findings. Although the CT scan findings did not correlate entirely with the angiographic and clinical findings, they were more frequently abnormal in later stages of the disease. The volume of Moyamoya increased progressively upto stage 3 *of the disease only to decrease with further progression. The etiology of Moyamoya disease in Indian children* is *not clear. However, the clinical and radiological features are similar to that in the Japanese.*

Key words: *Moyamoya disease, Computed tomography, Angiography, Brain infarction, Brain atrophy.*

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Moyamoya disease (MMD) has been reported frequently in the Japanese(l-6) with **a** few reports in non-Japanese populations(7-10). Moyamoya disease in children has been suggested to be distinct from that in adults(ll). The radiological features of MMD in children outside Japan are relatively undescribed(12). Moyamoya disease is an unknown cause of stroke in Indian children. Of a prospective study of 43 children with strokes from this institution, only 6 had Moyamoya disease(13). We report the angiographic and computed tomographic (CT) features in 8 Indian children with Moyamoya disease, seen at our centre over a period of six years.

Material and Methods

Angiograms and CT scans in 8 children with MMD were analyzed. Transfemoral selective serial external and' internal carotid angiography was done in 6 patients and two patients underwent direct antegrade puncture of the common carotid artery. Their ages ranged from 2 to 17 (mean = 7.5) years. The age at onset of the disease ranged from 1 to 11 (mean = 6.1) years. Detailed investigations including CSF analysis, blood sugar, VDRL, Lipid profile, rheumatoid factor and antinuclear factor were performed. Two patients underwent echocardiograms.

The angiographic data from a total of 14 sides of the anterior circulation was analyzed. Each side was classified into 5 stages depending upon the degree of stenosis of the internal carotid artery (TCA) and proximal portions of the anterior (ACA) and middle cerebral arteries (MCA) *(Table* J)(2). They were also clas-

sified into 6 stages of MMD depending upon the type and degree of collateral circulation *(Table* 7J)(12). The feeders to the basal Moyamoya disease were classified into those from the internal carotid artery (ICA) and the external carotid artery (ECA). The size of the Moyamoya blush was measured on the antero-posterior and lateral projections of the angiograms. The volume of the blush or abnormal network volume (ANV) was calculated as the product of the maximum dimensions of length, breadth and height.

The CT scans were done on CGR ND-8000 scanner (4 patients) and Seimen's Somatom DR3 (4 patients). Section thickness varied between 5 mm to 10 mm depending upon the region of interest. Contrast scan was done after intravenous bolus injection of 2 ml/kg body weight of meglumine iothalamate immediately prior to the scan. The scans were analyzed for infarcts, atrophy and abnormal enhancement. The CT and angiographic features were correlated with each other. The stage of ICA stenosis was correlated with the volume of abnormal vascular network on the same side.

TABLE *I*-*Progress of Stenosis of Moyamoya Disease (Suzuki and Takaku, 1989)*

Stage	Stenotic/Occlusive features
1.	Slight to moderate stenosis of TCA bifurcation
2.	Severe stenosis of ICA bifurcation
3.	Occlusion of ICA or MCA
4.	Occlusion of ICA or ACA and MCA, with partial retention of trunk of ACA/MCA
5.	Occlusion of ICA or ACA & MCA. Main trunk of ACA & MCA are not seen.

ICA- Internal carotid artery.
ACA-Anterior cerebral artery.
MCA-Middle cerebral artery.
ICA Bifurcation--First portions of ICA, ACA and MCA

Results

The clinical and radiological findings are shown in *Table III.* All the 8 patients showed cortical infarcts; the infarcts were bilateral in 6 and unilateral in 2. Deep ganglionic or white matter infarcts were seen in 5 patients *(Fig. 1).* The frontal, parietal and temporal lobes were commonly affected, while the occipital lobe was spared. Considering the hemispheres individually, 11 showed atrophy, either cortical, subcortical or both and 4 were normal. In one patient with an acute infarct, the presence and degree of the atrophy could not be assessed on the infracted hemisphere (Patient 2). Infarction on CT scan and

TABLE II- *Classification of Basal Moyamoya (Modified from Suzuki and Kodama, 1983)*

Stage	Angiographic features
1.	Narrowing of bifurcation of the internal carotid artery
2.	Initiation of basal Moyamoya
3.	Increase of Moyamoya. Obvious Moyamoya vessels at base of brain
4.	Decrease of Moyamoya
5.	Cross education of Moyamoya. All the main cerebral arteries are missing
6.	Disappearance of Moyamoya. Cerebral blood flow is only from external carotid artery

TABLE *III* - *Clinical and Radiological Features in Mayamaya Disease*

Pt. No.	Age (Yr)	Sex	Duration of illness	Clinical features	Angio-graphic findings	Angio-graphic stage	CT findings
1	3	F	2 weeks	Right hemipa-resis	RCA – CI stenosis M2 occulusion LCA- stenosis	3 1	Infarcts both hem ipsheres; left cortical atrophy
2	16	F	5 yr	Quadri-paresis	RCA - C1 occlusion Al occlusion LCA – C1 stenosis M1 stenosis	5 3	Infarcts both hemispheres
3	5	M	2 yr	Left facio-brachial weakness	RCA- C1 occlusion M2 occlusion LCA- C1 occlusion M2 occlusion	4 4	Infarcts left hemisphere; bilateral subcortical atrophy
4	17	F	12 yr	Left hemi-plegia and aphasia	RCA- C1 stenosis LCA- C1 occlusion M2 occlusion	3 4	Infarcts both hemispheres; bilateral cerebral atrophy
5	2	F	6mo	Quadri-paresis	RCA - C1 occlusion M1 stenosis LCA- C1 stenosis Ml occlusion	4 4	Infarcts both hemispheres; right hem is-pheric atrophy
6	10	F	4 yr	Para-pMMis	RCA- C1 occlusion M1 stenosis LCA- C1 occlusion M2 occlusion	5 5	Infarcts both hem ispheres; bilateral cere-bral atrophy
7	6	M	2 days	Right hemi-paresis	RCA- Not done LCA- Cl stenosis M2 occlusion	– 3	Infarct left hem isphere; right cortical atrophy
8	7	M	6mo	Quadri-paresis	RCA- Cl stenosis LCA- Not done	2 -	Infarcts both hem ispheres; bilateral cerebral atrophy

RCA - Right carotid angiogram; LCA – Left carotid angiogram.

clinical deficits did not correlate in 2 patients (Patients 1 and 3). One patient had bilateral infarctions with unilateral deficits and the other had unilateral clinical deficits and ipsilateral infarction.

A total of fourteen angiographic sides were analyzed and all of them, were abnormal. Of them, four sides were in angiographic stage 3, three sides were in stage 4 and three were in stage five *(Figs. 2 &* 3). The cervical ICA was narrow in 4 patients (8 sides). The stenosis of the ICA extended from the cervical to the supraclinoid portion *(Fig. 4)*. Of the 8 patients with narrow ICAs, 7 patients had severe degree of basal Moyamoya. Collateral circulation was from both the ICA and ECA systems.

Of the ICA collaterals, anterior falcine artery, ethmoidal branches and recurrent branches of the ophthalmic artery were the most frequent and seen in 65 to 75% of the patients. Predominent ECA collaterals were the anterior and posterior branches of the

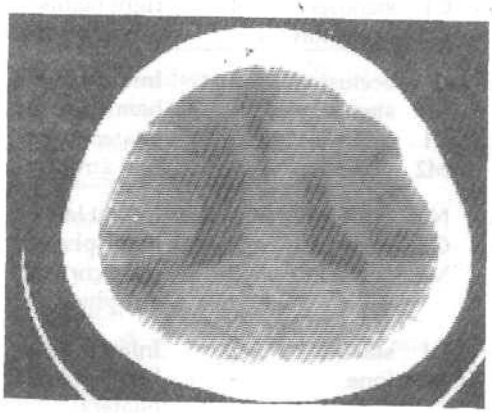

Fig. 1. Baseline scan. Axial slice at the level of lateral ventricles showing multiple cortical infarcts in both the frontal and parietal lobes and ventricular dilatation.

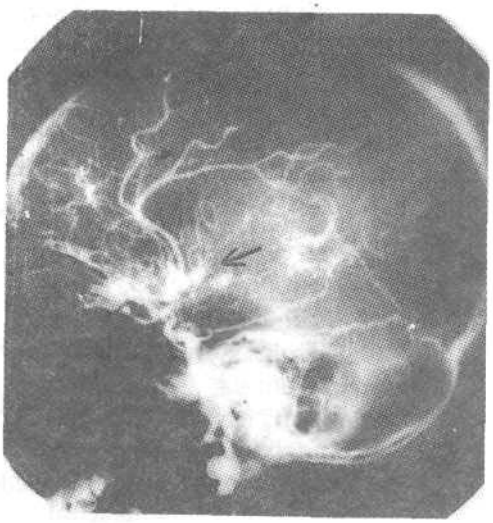

Fig. 2. Left carotid angiogram: Arterial phase, lateral view. There is basal and orbital Moyamoya (arrow) with severe narrowing of the supraclinoid internal carotid artery and total occlusion of the middle cerebral artery.

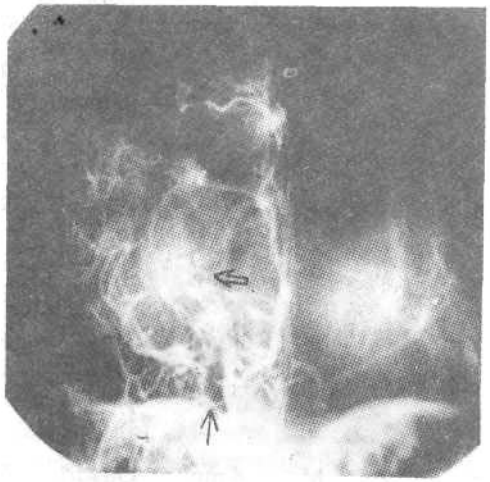

Fig. 3. Right carotid angiogram: Arterial phase, anteroposterior view. There is severe narrowing of the M1 segment of the middle cerebral artery (arrow) with good distal run off through the basal Moyamoya.

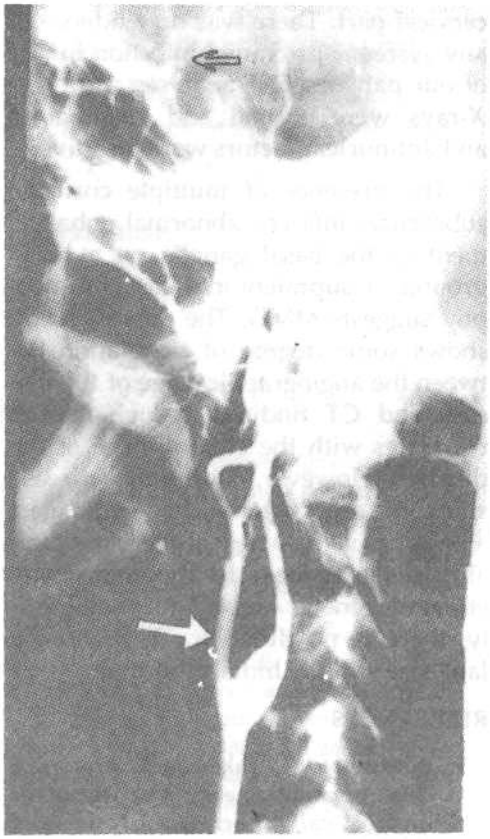

Fig. 4. Left carotid angiogram. Arterial phase. There is diffuse narrowing of the cervical internal carotid artery (arrow) with severe stenosis of the supraclinoid part (arrow head).

middle meningeal artery and the superficial temporal artery and seen in 50 to 65% of the patients. The number and extent of collateral participation increased progressively with the severity of the disease. Correlating the scan abnormalities with the angiographic stage of the disease, the frequency of abnormalities on scan increased progressively with the stage of the disease. All the cerebral brain hemispheres with corresponding angiographic stenosis Stages of IV and V showed infarction.

Enhancement of the superficial cortex was seen across various stages of the disease (Stages II to V). However, abnormal intense enhancement of the basal ganglia was seen only in 2 patients in later stages of the disease (Stages IV and V). There was a distinct difference in the type of cerebral atrophy seen in different stages of the disease. While cortical atrophy (6 patients) was seen in patients from Stages I to V, subcortical atrophy (8 patients) was overwhelmingly seen in patients in Stages IV and V (75%). Involvement of the posterior circulation was seen in only 3 patients and in none of them was the CT scan demonstrative of any infarctions in the vertebrobasilar territory.

Discussion

The angiographic features which are previously described in children with MMD were corraborated in this study. They include narrowing/occlusions of the supraclinoid ICA and MCA with development of Moyamoya vessels in the base of the brain(2). Some of the features as yet unexplained include bilateral involvement of ICA, unpredictable rate of progression of the disease and spontaneous regression in later life(12). Though all the patients in the present study had evidence of bilateral disease on angiogram, the stage/severity of disease did not correlate with the scan in many patients. This disparity is probably due to the slow, asymmetric progression of the stenotic processes with development of extensive collaterals via the posterior and external carotid circulations.

Reports of abnormal enhancement in the region of the basal ganglia are variable(4,14,15). Although differences in the technique of administration of contrast medium could be 3 factor(16), the severity of the disease is perhaps an important determinent. The fact that all of our patients with enhancement of the basal ganglia were in angiographic Stages IV and V, is suggestive. The volume of the Moyamoya (ANV) when correlated with the degree of stenosis of the ICA, showed progressive increase of both (upto Stage III). With further progression of the stenosis of the TCA (Stages IV and V), the volume of the Moyamoya dropped.

ANV has been reported to be more extensive in children as compared to the adults(2). Results of the present as well *as* previous studies have shown ANV to be maximal in Stage III and to decrease with further progression of the disease(2,7,11,12,15).

An interesting feature of our study was the presence of narrowing of both cervical ICAs in 4 patients. All these patients were in angiographic Stages IV and V. Since involvement of cervical ICA has not been described easier as « feature of childhood MMD, it is possible that MMD as it occurs in the native Indian population may be different from that described in the Japanese(12). It is not clear from the present study but likely that the disease starts in the terminal part of the ICA and subsequently progresses distally to involve, the ACA and MCA and proximally to involve the cervical ICA. This is partly substantiated by the angiographic evidence of frequent and earlier involvement of the terminal part of ICA as compared to its cervical part. There was no evidence of any systemic or cranial infection in any of our patients. CSF analyses and chest X-rays were normal and rheumatoid and antinuclear factors were negative.

The presence of multiple cortical/ subcortical infarcts, abnormal enhancement of the basal ganglia on a background of supratentorial cerebral atrophy suggests MMD. The present study shows some degree of correlation between the angiographic stage of the disease and CT findings though neither correlates with the clinical stage of the disease. However, once the disease starts, the gross arterial changes suggest a progressive response of the intracranial vascular system to the underlying etiopathogenetic cause. Morphologically, there is no difference between the Japanese and the Indian children.

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