Brief Reports

Magnetic Resonance Angiography in Moyamoya Disease

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Moyamoya disease is a rare cerebrovascular condition of unknown etiology. It is common in Japanese, though few cases have been reported in Negroes, Indians and elsewhere(l-4). In adults, subarachnoid hemorrhage is the most common presentation, where as in children the clinical manifestations are the result of cerebral ischemia(2).

Contrast cerebral angiography is currently the means of diagnosing Moyamoya disease. The angiographic findings include stenosis of both internal carotid arteries at their supraclinoid portions extending to the proximal portions of the anterior cerebral artery and middle cerebral artery and presence of parenchymal, leptomeningeal and transdural collateral vessels(2-9).

CT scan and MR imaging finding are complementary to angiography. in the . diagnosis of Moyamoya disease. The CT scan shows cerebral atrophy, most prominent in the frontal lobes, with widening of the cerebral sulci, sylvian fissures and interhemispheric fissures. Focal areas of non-enhancing or mildly contrast enhancing hypodense areas are also present in the basal ganglia or cerebral cortices. The parenchymal, dural and leptomeningeal collaterals appear on contrast CT as curvilinear, tortuous, vascular\iensities(4,8-13).

Recently magnetic resonance angiography (MRA) has been introduced to noninvasively evaluate intracranial vascular lesions without the use of contrast media(4,6,7). MRA displays the signal from flowing blood. It can be done either in time-of-flight (TOF) or in phase contrast technique. Time-of Flight technique can be of either 3D types or 2D type(6,7,14,16). We report four such cases evaluated with 3D TOF MR angiography during the last two years.

Material and Methods

Four patients with Moyamoya disease were studied by 3 D TOF MRA. The patients were initially subjected to plain and contrast enhanced CT examination of the head. Following this, all patients underwent invasive conventional contrast angiography. These patients were 7 to 12 years old. The clinical profile of

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these patients is given in *Table I.* In 2 patients, the average interval from the appearance of the first symptoms to evaluation at this centre was 18 months and in the remaining 2 the internal was 4 years. All cases were treated symptomatically. Patients were subjected to MRA with 1.5 T MR imaging system (Magnetom, Siemens). MRA was performed using 3D fourier transformation rephrased gradient recalled echo sequences.

The scans were performed in axial, sagittal and coronal planes. A linearly polarized head coil was used. No intravenous contrast media were used. All 4 children were sedated before the study. The duration of study was about 40 minutes. Following date acquisition, the reconstruction was done using a computer ray tracing algorithm to yield maximum intensity projection images. Reconstructed MRA images, CT picture and

TABLE I—*Clinical Features CT Scan and MR Findings in Moyamoya Disease.*

Case No.	Age/Sex (Yr)	Clinical Features	CT Findings	MR Findings
1.	11/male	History of seizures, aphasia, right sided hemiplegia	Left temporo-parietal infarcts	Hyperintense areas in T2 weighted image & hypointense in T1 weighted image in the left temporo parietal region suggestive of infarcts.
2.	7/male	History of altered sensorium, aphasia left sided hemiplegia	Right temporo parietal infarct; mildly enhancing infarcts in the right basal ganglia ; curvilnear tortuous vascular densities in the right temporo parietal region.	Right temporo parietal infarct; infarct in the right basal ganglia
3.	9/male	History of recurrent focal seizures, recurrent hemiparesis, altered sensorium	Middle cerebral artery infarct	Infarct in the territory of middle cerebral artery
4.	12/male	History of seizures, alternating recurrent hemiparesis	Bifrontal infarcts and cerebral atrophy	Infarcts in the bifrontal region along with cerebral atrophy

conventional angiograms were reviewed to look for individual vessels, site of occlusion of the vessels, the presence of collaterals, the presence of blush and the extent of infarcts. The conventional invasive contrast angiograms were used as the standard reference in analysis of the MRA findings.

Results

Normal circle of Willis and basal cerebral portions of anterior, middle and posterior cerebral arteries and internal carotid arteries were visualized in MRA done on a normal volunteer *(Fig. 1)*.

In all the 4 patients, the occlusion of both internal carotid arteries in their supraclinoid portions was conclusively demonstated *(Fig. 2)*. The proximal portion of the anterior and middle cerebral artery were occluded. In 2 cases (Cases 2 and 4) the posterior cerebral artery could be visualized properly. In 1 case (Case 1), the full extent of the internal carotid artery till its supraclinoid portion could not be visualized on one side as the occlusion was localized in the cervical portion of the internal carotid artery. The internal carotid artery on the other side of the same patient was occluded at it's supraclinoid portion. Basal cerebral Moyamoya vessels were visualized in 3 patients *(Fig. 2)*. The findings were well correlated with those conventional angiograms. The results were compared with conventional angiograms. However, MRA could not detect basal Moyamoya vessels in one patient (Case 1). Leptomeningeal anastomoses from the posterior to anterior circulation were visualized well in 2 out of 4 patients (Cases 2 and 4) and poorly in 1 case (Case 3). Basal gang-lionic blushes were seen in all cases *(Fig. 2)*.

Collateral vessels from the dilated superficial temporal artery to the territory of the middle cerebral artery were visualized on both sides in only 2 out of 4 cases. Periorbital external carotid collaterals were shown in 2 out of 4 cases *(Fig.* 2). The routine T1 and T2 SE sequences images taken in axial planes from the base to vertex and findings were almost similar to the axial scans obtained from CT scan. *(Table II)*.

Discussion

Cerebral arteriography is still considered the gold standard for diagnosis of

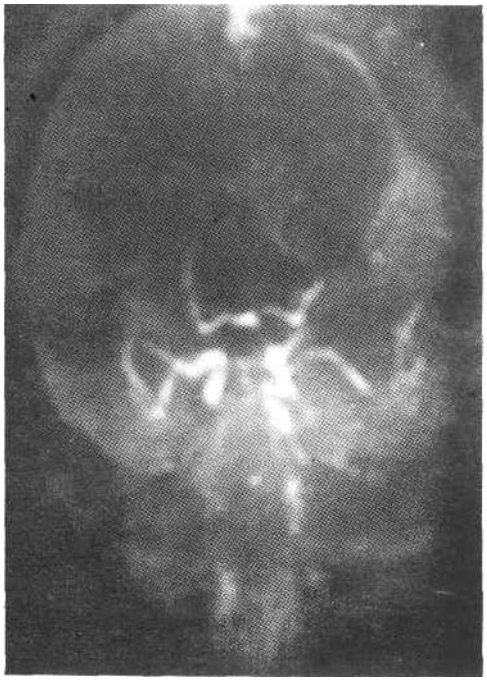

Fig. 1. *MR angiogram of a normal healthy volunteer in the axial plane shows the normal circle of willis and basal cerebral portions of anterior cerebral artery and internal carotid artery.*

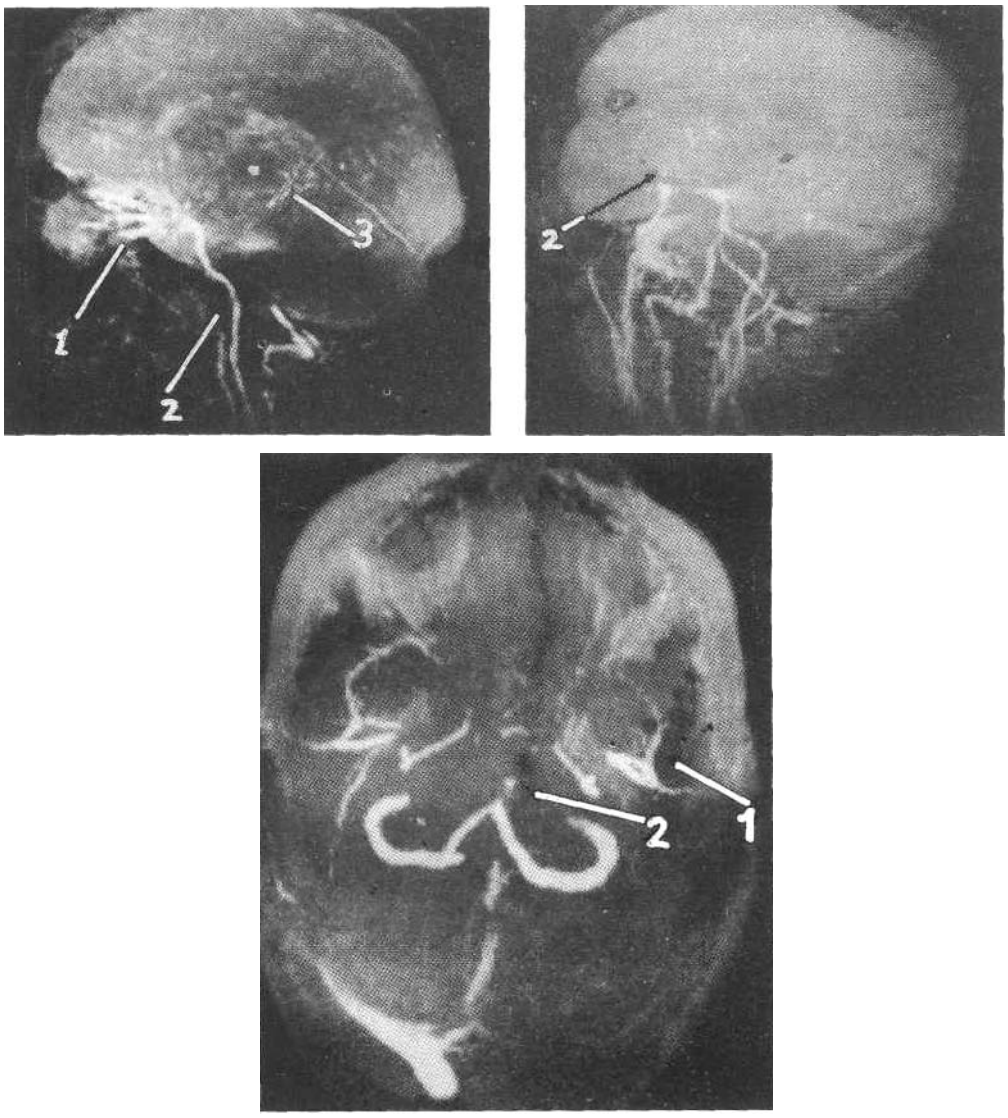

Fig. 2. MR angiogram in sagittal plane (a), coronal plane (b) and axial plane (c) shows the evidence of block of both internal carotid arteries at their supmclinoid portions with external carotid collaterals (1) in (c): basal ganglial collaterals (2) in (c) and (b); leptomeningeal collaterals (3) in (a). The internal carotid artery (2) in (a) is also shown.

the Moyamoya disease-although the usefulness of computed tomography and conventional MR imaging in diagnosis of this disease has already been establised(9,13). In Moyamoya disease, the supraclinoid portions of both inter-

TABLE II–*Conventional Angiogram And MRA Findings In Moyamoya Disease*

Case No.	Conventional angiogram	MRA
1.	Occlusion of right internal carotid ortery at it's supraclinoid portion. Nonvisualization of the supraclinoid portion of the left internal carotid artery. Basal Moyamoya vessels were seen.	Occlusion of right internal carotid artery at it's supraclinoid portion. Nonvisualization of the supraclinoid portion of the left internal carotid artery. Basal Moyamoya vessels could not be visualized.
2.	Supraclinoid block of both internal carotid arteries. Leptomeningeal collaterals and basal ganglionic blush were seen.	Supraclinoid block of both internal carotid arteries. Leptomeningeal collaterals and basal ganglionic blush were seen.
3.	Supraclinoid block of both internal carotid arteries. Basal Moyamoya vessels were seen. Leptomeningeal collaterals were visualized poorly.	Supraclinoid block of both internal carotid arteries. Basal Moyamoya vessels were seen. Leptomeningeal collaterals were visualized poorly.
4.	Supraclinoid block of both internal carotid arteries. Basal Moyamoya vessels, leptomeningeal collateral and transdural collaterals were seen.	Supraclinoid block of both internal carotid arteries. Basal Moyamoya vessels, leptomeningeal collateral and transdural collaterals were seen.

nal carotid arteries are occluded or narrowed along with the most proximal portions of the anterior and middle cerebral arteries. The enlarged lenticulo-striate and thalamoperforate arteries form the extensive parenchymal collaterals. The name "Moyamoya" which means "Hazy" or "Misty" in Japan has been coined by these parenchymal collaterals.

Ischemic complications, including death have been reported follwing the invasive cerebral arteriography in patients with Moyamoya disease(8,9). An alternative safer and earlier procedure like MRA, therefore, has tremendous potential in the diagnosis of this condition.

In our study all the 4 patients showed neurological deficits concordant with the site of focal deficit demonstrated on MRI scans. MRA was a useful screening test for evaluating the extent of Moyamoya disease. Almost all the relevant cerebral vessels and collaterals could be visualized by MRA. In one case, the internal carotid on one side could not be visualized upto the supraclinoid region, due to a proximal block resulting in loss of signal arising from the occluded vessels (Case 1).

The advantage of MRA lies with it's ability in multiplanar imaging and also the maximum intensity projection reconstruction which allows the visualization of the vessels at any angle. However,

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MR angiography has certain limitations like intravascular signal loss in the presence of rapid or turbulent blood flow(6). Spatial resolution of MR angiography is markedly lower than that of conventional angiography(6,7). With the rapid advances in MR imaging techniques, noninvasive MRA may prove to be an alternative rather than complementary to invasive contrast angiography in the future.

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