Moyamoya disease: clinical manifestations and neuroimaging characteristics. A retrospective study of 4 patients

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Purpose

Moyamoya disease (MMD) is a rare occlusive cerebral vasculopathy of unknown etiology consisting of stenosis or occlusion of the distal portions of the internal carotid arteries and their major branches.

In rare cases, this process also involves the posterior circulation, including the basilar and posterior cerebral arteries.

It is characterized by a tendency toward multiple ischemic neurological events and small strokes in younger patients and in older populations. It can often be associated with further vascular degeneration and intracerebral hemorrhage.

The aim of this study is to discuss and illustrate the clinical and radiological manifestations of moyamoya disease over a retrospective study of four cases.
Methods and Materials

The clinical data of 4 patients with moyamoya disease were retrospectively studied. All our patients were explored with cranial CT scan, MRI performed in 3 patients and cerebral angiography done in 3 patients.
Results

Clinical findings

- The age of our patients was ranging from 18 months to 48 years. There were three pediatric cases (18 months -11 years) and one adult (48 years), with a sex ratio of 3 females/1 male.
- The main clinical symptoms at onset were: Transient ischemic attacks (1 case), ischemic strokes (3 cases), seizures (2 cases), recurrent headaches (3 cases) and hemorrhagic infarction (2 cases).
- The EEG showed various abnormalities at rest (3 cases) consisting in increased slow-wave activity mainly in anterior regions (2) and spike discharges (1 case).
- After hyperventilation, a delayed return to baseline pattern was observed; in 3 pediatric cases.
- The re-build-up phenomenon (polymorphous and high-voltage slow waves) was detected in 2 of pediatric cases.

Computed tomography (CT) findings

- Cerebral atrophy, ventricular dilatation and subarachnoid space widening were noted in 2 cases, (Fig 1, 2, 3).
- Low density areas (single in one case, multiple bilateral in 2 cases) were observed (Fig: 1, 2, 3).
- Cortical and subcortical hemorrhage seen in 2 cases: (Fig: 4, 5).
- After contrast injection enhancing areas were evident in 3 cases (Fig: 6, 7), and bright curved linear densities over the brain base and basal ganglia observed in 2 cases.

Magnetic resonance imaging (MRI) findings

- Cortical and/or subcortical atrophy was observed in 2 cases: Fig (8, 9, 10);
- Hyperintense areas (T2 and T2 Flair -weighted images) single in one case; or multiple bilateral in 2 cases: Fig (11, 12, 13, 14).
- Contrast cortical enhancement seen in 2 cases: Fig (15).
- Punctate signal void findings in the basal ganglia were observed in 2 patients: Fig (16, 17, 18).
- Steno-occlusion of internal carotid artery and/or middle cerebral artery was also evident by MRI and MRA imaging in all cases Fig (19, 20, 21, 22, 23).
- Involvement of anterior and middle cerebral arteries observed in all cases: Fig(19, 20, 21, 22, 23), and right posterior cerebral artery in one
Angiographic findings

- The selective cerebral angiographic study, performed in 3 patients, showed steno-occlusive lesions of main vessels at the skull base, with bilateral involvement in all patients (Fig: 24, 25, 26).
- Stenotic or occlusive disease prevailed on the carotid districts in all cases; the vertebro-basilar system was Affected only in one case (Fig:26).
- Collateral networks were evident in all cases, mainly in the basal ganglia.
- The correlation versus conventional angiographic patterns was satisfactory.
- The medical therapy was followed by all of cases and no surgical treatment was Performed.

DISCUSSION

Moyamoya disease is an uncommon cerebrovascular disease that is characterised by progressive stenosis of the terminal portion of the internal carotid artery and its main branches.

The disease is associated with the development of dilated, fragile collateral vessels at the base of the brain, which are termed moyamoya vessels.

The incidence of moyamoya disease is high in east Asia, and familial forms account for about 15% of patients with this disease.

Moyamoya disease has several unique clinical features, which include two peaks of age distribution at 5 years and at about 40 years. Most paediatric patients have ischaemic attacks, whereas adult patients can have hemorrhagic attacks.

This age distribution was found in our series as three of our patients were under the age of 10 years an only one patient was 40 years older.

Moyamoya should be considered in patients, particularly children, presenting with acute neurologic deficits or unexplained symptoms referable to cerebral ischemia.

Computed tomography (CT) in a patient with moyamoya disease may show small areas of hypodensity suggestive of ischemia or hemorrhage in the cortical watershed zones, basal ganglia, deep white matter, or periventricular regions. Focal or multifocal cerebral atrophy, ventricular dilatation and subarachnoid space
widening resulting from chronic ischemic lesions as seen in three of our patients. However, the CT scan can be normal, particularly in patients presenting solely with TIAs.

After contrast injection enhancing areas and bright curved linear densities over the brain base and basal ganglia are suggestive of the diagnosis as seen in three of our patients.

CT angiography can show the intracranial stenoses seen in moyamoya. Thus, CT angiography should be considered when magnetic resonance imaging (MRI) is not readily available and a diagnosis of cerebral occlusive vasculopathy is being considered.

The finding most suggestive of moyamoya on MRI is reduced flow voids in the internal, middle, and anterior cerebral arteries coupled with prominent flow voids through the basal ganglia and thalamus from moyamoya-associated collateral vessels. This sign was observed in 2 of our patients. These findings are virtually diagnostic of moyamoya.

An acute infarct is more likely to be detected with the use of diffusion-weighted imaging, whereas a chronic infarct is more likely to be seen with T1- and T2-weighted imaging. Diminished cortical blood flow due to moyamoya can be inferred from fluid-attenuated inversion recovery (FLAIR) sequences showing linear high signals that follow a sulcal pattern, which is called the "ivy sign" seen in 3 of our cases.

Magnetic resonance angiography (MRA) is also useful to diagnose moyamoya disease in a non-invasive way. MRA can be used to identify stenotic lesions in the ends of the carotid artery; thus, MRA has enabled easier detection of asymptomatic patients with familial moyamoya disease. However, the possibility of overestimation of accuracy should always be taken into account because of the imaging quality.

MRA can also be used to identify moyamoya vessels around the basal ganglia and thalamus.

The definitive diagnosis is based on a distinct arteriographic appearance characterized by stenosis of the distal intracranial internal carotid artery, extending to the proximal anterior and middle arteries.

Three of our patients presented multiple proximal stenosis of the carotid system. Only one pediatric patient presented abnormalities of the vertebro basilar system as occlusion of the right posterior cerebral artery which is unusual with moyamoya disease.
Development of an extensive collateral network at the base of the brain along with the classic "puff of smoke" appearance on angiography is characteristic as seen in all of our patients. Imaging of the external carotid arteries is essential to identify any preexisting collateral vessels so that surgery, if performed, will not disrupt them.

Aneurysms, as well as the rare arteriovenous malformation known to be associated with certain cases of moyamoya, are also best detected by means of conventional angiography.
Images for this section:

Fig. 0: Brain Ct scan showing cerebral atrophy, ventricular dilatation and subarachnoid space widening with multiple bilateral low density areas.

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**Fig. 0:** Brain Ct scan showing cerebral atrophy, and subarachnoid space widening with multiple bilateral low density areas in fronto parietal regions.

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Fig. 0: Enhanced brain Ct scan showing single low density area in left parietal lobe with cerebral atrophy, ventricular dilatation and subarachnoid space widening.

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**Fig. 0**: Unenhanced brain Ct scan showing left parieto occipital cortical and subcortical hemorrhage.

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Fig. 0: Unenhanced brain Ct scan showing left parieto occipital cortical and subcortical hemorrhage.

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Fig. 0: Brain CT scan After contrast injection showing enhancing areas with bright curved linear densities over basal ganglia and left parieto occipital region.

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**Fig. 0:** Brain CT scan after contrast injection showing enhancing areas with bright curved linear densities over left parieto occipital region.

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Fig. 0: Brain MRI, axial T2 weighted images showing cortical and subcortical atrophy over the fronto parietal areas.

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**Fig. 0:** Brain MRI, axial T2 weighted images showing cortical and subcortical atrophy over the fronto parietal areas and left occipital region.

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Fig. 0: Brain MRI, axial T2 Flair weighted image showing hyperintense cortical areas (the ivy sign) in frontal areas and in basal ganglia (caudate nucleus head bilaterally).

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**Fig. 0:** Brain MRI, axial T2 Flair weighted image showing hyperintense cortical areas in fronto parietal regions: the ivy sign.

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Fig. 0: Brain MRI, axial T2 Flair weighted image showing hyperintense cortical and subcortical areas in right occipital region: the ivy sign.

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**Fig. 0:** Brain MRI, axial T2, weighted image showing hyperintense cortical and subcortical areas in right parieto frontal regions.

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Fig. 0: Brain MRI: Axial post contrast T1 weighted image showing bilateral curvilinear cortical enhancement in fronto parietal areas.

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**Fig. 0:** Brain MRI, axial post contrast T1 weighted image showing Punctate signal void findings in the basal ganglia.

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Fig. 0: Brain MRI, coronal post contrast T1 weighted image showing Punctate signal void findings in the basal ganglia.

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**Fig. 0:** Brain MRI, axial T2 weighted image showing Punctate signal void findings in the basal ganglia.

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Fig. 0: Brain MRA:axial 3D TOF weighted image showing Steno-occlusion of middle and anterior cerebral arteries with extensive collateral network at the base of the brain.

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**Fig. 0:** Brain MRA: 3D TOF MRA weighted image showing Steno-occlusion of distal internal carotid artery, middle and anterior cerebral arteries with extensive collateral network at the base of the brain.

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**Fig. 0**: Brain MRI: axial T2 weighted image showing Steno-occlusion of distal internal carotid artery, middle and anterior cerebral arteries with extensive collateral network at the base of the brain.

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Fig. 0: Brain MRA: coronal enhanced contrast MRA weighted image showing Stenoo-
occlusion of distal internal carotid artery, middle and anterior cerebral arteries with
extensive collateral network at the base of the brain.

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**Fig. 0:** Selective Angiogram of right internal carotid arteries showing stenosis of the distal intracranial internal carotid artery, extending to the proximal anterior and middle cerebral arteries. Development of an extensive collateral network at the base of the brain along with the classic "puff of smoke" appearance on angiography

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Fig. 0: Selective Angiogram of left internal carotid artery showing stenosis of the distal intracranial internal carotid artery, extending to the proximal left anterior cerebral artery. Extensive collateral network at the base of the brain along with the classic "puff of smoke" appearance on angiography.

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Fig. 0: Selective Angiogram of left vertebral artery, showing occlusion of the right posterior cerebral artery with development of an extensive collateral network at the base of the brain.

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Conclusion

Moyamoya disease should be considered in all young patients, especially children, presenting with stroke.

The radiological investigations play a crucial role in confirming the diagnosis. Cerebral angiography has occupied a prominent place in the diagnosis of moyamoya disease. Currently MRI and MRI angiography play an increasingly important role in the diagnostic confirmation and may even replace cerebral angiography.
References
