

# The 'Ivy Sign' of Moyamoya disease

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## ABSTRACT

Stroke is uncommon in pediatric age group. Moyamoya is a rare idiopathic disease characterized by progressive vaso-occlusion of the main blood vessels to the brain. We report a case of 13 year old female who presented with recurrent headache and transient left hemiparesis and was diagnosed to have Moyamoya disease. The 'ivy sign' is a characteristic magnetic resonance image finding seen in patients with Moyamoya disease.

**Key words:** Moyamoya; Ivy sign; Magnetic resonance image

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## INTRODUCTION

Moyamoya disease is a rare idiopathic cerebrovascular disorder. There is stenosis of distal internal carotid arteries and anterior and middle cerebral arteries; as a result of which several collaterals develop to compensate for the occlusion. The incidence is 0.086 per 100,000 individuals.<sup>1</sup> It is usually seen in association with conditions like Down's syndrome, sickle cell disease and neurofibromatosis-1.<sup>2-5</sup> The disease commonly presents as stroke in children, and as subarachnoid or intracranial haemorrhage in adults.<sup>6</sup>

## CASE REPORT

A 13 year old female presented with history of recurrent headache since 1 year, which was mainly confined to both temporal regions. Her last headache episode was 2 days ago and she noticed weakness of her left upper (difficulty in combing her hair and lifting tea cup) and lower limb (difficulty in walking). However, her weakness resolved in about 6 to 8 hours along with her headache. She did not have similar episodes of weakness in the past. Her headache was not associated with nausea or vomiting. There was no facial deviation, visual disturbance, loss of consciousness or seizures. She was initially taken to a local practitioner and was told to have hemiplegic migraine.

On presentation, she was conscious, oriented and afebrile, with stable vitals. Her systemic examinations were normal,

with no neurological deficits. There were no signs of meningeal irritation. Her blood investigations like complete blood counts, peripheral blood smear, renal and liver functions, electrolytes, calcium, antinuclear antibody profile and TSH were normal. ECG and echocardiogram were also normal. Her magnetic resonance imaging (MRI) of brain showed linear and curvilinear hyperintense signals in the sulcal spaces and bilateral basal ganglia (Figure 1) and MR angiography revealed severe narrowing of bilateral supracavernous internal carotid arteries with highly attenuated flow signal intensities in bilateral anterior and middle cerebral arteries. Collateralisation was evident in bilateral cerebral hemispheres, suggestive of Moyamoya disease.

She was started on antiplatelet medication, and was advised the need for revascularisation procedure; but due to financial issues she had to be referred to Government Medical College for further neurosurgical management.

## DISCUSSION

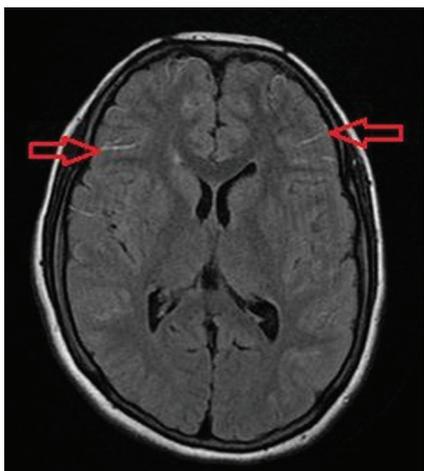
Moyamoya disease was first described by Takeuchi and Shimizu from Japan in the year 1963. The disease is characterized by chronic, progressive occlusion of the arteries of circle of Willis leading to the development of collateral vessels, which can be seen on MRI, particularly cerebral angiography.<sup>7</sup> The process of narrowing of cerebral vessels is believed to be a reaction of the blood

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**Figure 1:** Ivy sign on T2 FLAIR image

vessels to several external stimuli, injuries, or genetic defects. The term Moyamoya, which means “puff of smoke” in Japanese, came from the characteristic appearance of small, multiple vessels near the apex of the carotids on angiography. It was initially believed to affect mainly Asian population, but has now been observed worldwide. The condition is more common among females. The peak incidence is seen in children who are 5 years old and adults in their fourth decade.<sup>8</sup> Familial transmission has been observed in 15% of these patients.<sup>9</sup> The term Moyamoya syndrome is used when the disease is found in association with systemic lupus erythematosus, Down syndrome, neurofibromatosis-1, glycogen storage disease type-1a, hereditary spherocytosis, sickle cell anaemia, radiation treatment and tuberculosis meningitides. Epstein Barr virus infection has also been associated with Moyamoya disease.<sup>10-12</sup> Congenital heart defects, antiphospholipid syndrome, renal artery stenosis and thyroiditis have been noticed with Moyamoya.<sup>13</sup>

The clinical features differ in children and adults. Transient ischemic attack with infarcts involving the internal carotid artery territory in the frontal lobe is seen in children. They present with headache, seizures, involuntary movements like hemichorea, monoparesis, hemiparesis, aphasia and dysarthria.<sup>10</sup> Slowly progressive mental impairment has also been noticed in untreated cases. Adults, on the other hand, present with subarachnoid or intracranial haemorrhage.

The disease is characterized by the development of collateral in order to compensate the stenosis. The first collateral, known as the basal moyamoya, includes abnormal dilatation of the lenticulostriate and perforating arteries in the thalamus and basal ganglia. The dilatation of the anterior choroidal and posterior pericallosal arteries forms the second collaterals. Ethmoidal moyamoya, the third collateral, includes the abnormal dilatation of the

anterior and posterior ethmoidal arteries which supply the anterior cerebral artery branches from the ophthalmic arteries. The last collateral path, the vault moyamoya, is between the dural and the pial arteries.<sup>10</sup>

Cerebral angiography is the gold standard method for diagnosing Moyamoya disease. However, since it is invasive, MRI and MR angiography are preferred, especially in children. The ‘ivy sign’ is a MRI finding of Moyamoya disease, seen in post-contrast and fluid attenuated inversion recovery (FLAIR) images. It was first described in post contrast MRI as a gadolinium accumulation in the pial vasculature appearing as leptomeningeal and cortical contrasting images; and is seen in 70% of the patients. Cortical and leptomeningeal signal increase have also been reported in FLAIR images. The sign derives its name due to leptomeningeal contrasting on the cortical surface and FLAIR signal increase which resembles ivy growing on a rock.<sup>13,14</sup>

There is no effective medical therapy; and acute management is mainly symptomatic and aims at reducing the elevated intracranial pressure, improving cerebral blood flow and controlling seizures.<sup>15</sup> Surgical revascularization improves cerebral perfusion, thereby reducing the risk of stroke in both paediatric and adult patients. Surgical options are either direct bypass extending from the superficial temporal artery to the middle cerebral artery or encephaloduroarteriosynangiosis (EDAS) or encephalomyosynangiosis (EMS) including indirect bypass and combined bypass techniques. The prognosis depends on the age of the patient and the nature of presentation. All age groups have demonstrated similar hemodynamic improvement after surgical procedures. The patients with infarctions hold a poor outcome when compared to those presenting with transient ischemic attack and seizures.<sup>16</sup>

## CONCLUSION

The diagnosis of Moyamoya disease can be difficult mainly due to the rarity. The condition can mimic hemiplegic migraine. Imaging techniques like MRI and MR angiogram aid in the diagnosis. Knowledge of the clinical manifestations and MR findings is essential, as early diagnosis can improve neurological prognosis, especially in children.

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