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Moyamoya disease presenting with hemiballismus and parkinsonism

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Abstract

A young Nepali woman having characteristics angiographic findings of moyamoya disease and manifesting hemiballismus and parkinsonism is reported. To the best of our knowledge, this is the first case report of moyamoya disease from Nepal.

INTRODUCTION

Moyamoya, a Japanese word meaning "something hazy like puff of smoke drifting in air", was first used in 1963[1] to describe a rare occlusive cerebrovascular disease having angiographic findings of progressive occlusion of the terminal part of both internal carotid arteries and the proximal segments of the anterior and middle cerebral arteries with development of anastomotic channels at the base of brain.[1-3] These collaterals develop in an attempt to improve circulation across the occluded vessels and are believed to consist of pre-existing and new vessels arising from the circle of Willis and connecting with the distal portion of the middle cerebral arteries. These collaterals also enter brain substance in the region of lenticulostriate and thalamoperforate arteries.[4] These collaterals consist of dilated and fragile vessels having an attenuated media and fragmented internal elastic lamina.[5] How these vessels develop is not clear and the roles of angiogenic growth factors and adhesion molecules are under study.[6] Hemorrhagic stroke in adults and ischemic stroke in children are the common presentations of moyamoya disease.[1-3] Other manifestations are rare. Observation of moyamoya disease in a young Nepali woman presenting as involuntary movements and parkinsonism led to this case report and brief review of literature.

CASE REPORT

A 23-year-old Nepali woman presented with acute onset of involuntary movements affecting the left hand. The movements initially consisted of uncontrollable non-purposive movements at rest and wild flinging movements during active use of the affected limb which lasted for 2 weeks, followed by resting tremors which persisted. She had no risk factors for stroke and denied any history of fever, sore throat, arthralgia, rash or photosensitivity. Family history for similar illness was negative. She was not on any medications.

On examination she was afebrile with BP 110/70 mm Hg and Pulse 80/min. Neurological examination revealed normal higher mental functions. No motor, sensory or cerebellar deficits were detected. Deep tendon reflexes were normal with plantar responses flexor on both sides. She had rigidity, bradykinesia and a pill-rolling resting tremor, more on the left side. In addition, she had postural and kinetic tremors. The tremor increased markedly during walking however she was able to carry her daily activities independently. Her gait was hesitant and slow but she had no ataxia or freezing of gait. Other systems were normal.

Her haematological parameters, blood biochemistry, liver function tests, electrocardiogram, chest X-ray and echocardiography were normal. Workup for coagulation disorders, connective tissue disorders and Wilson's disease was negative. MRI FLAIR imaging revealed bilateral hyperintense signals over the cortical surface, leptomeninges and basal ganglia in the middle cerebral artery and anterior cerebral artery territories. The lesion showed gadolinium enhancement consistent with the "IVY" sign of moyamoya disease (Figures 1A and 1B). Angiography showed occlusion of both internal carotid arteries with prominent collaterals (Figures 2A and 2B). She was treated with physiotherapy, aspirin, trihexyphenidyl and amantadine. When reviewed after 3 months, she was free from transient ischaemic attacks and had fine resting

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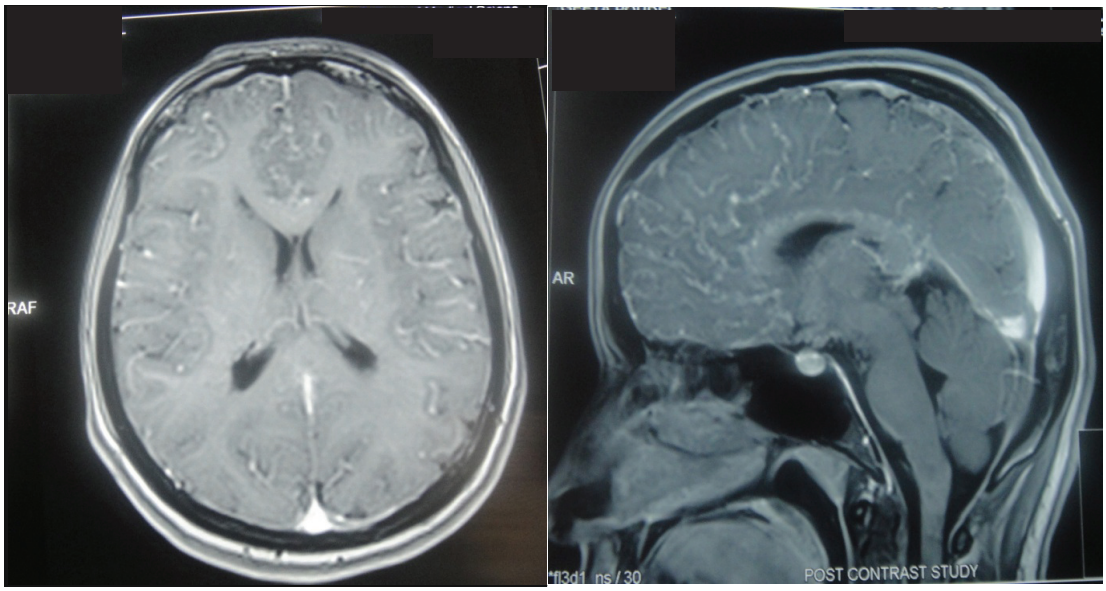

Figure 1. Post-contrast axial (A) and sagittal (B) T1-weighted images showing Gadolinium enhancement in a sulcal pattern due to the formation of collaterals from leptomeninges ("Ivy sign").

tremors only. Mild improvement in rigidity and bradykinesia was also noted.

DISCUSSION

The clinical diagnosis of moyamoya disease is based on the demonstration of typical angiographic abnormalities either with conventional angiography (Criterion "A") or by CT or MRI angiography (Criterion "B"), and exclusion of other causes of cerebrovascular disease (Criterion "C").[7] In the present case, conventional angiography showed typical findings (Figures 1A and 1B) and she had no other systemic illness, thus fulfilling diagnostic criteria "A" and "C". In addition, MRI FLAIR sequences showed linear high signal following a sulcal pattern and marked enhancement of cortex and leptomeninges with gadolinium (Figures 1A & 1B), termed "IVY sign" due to its resemblance to creeping poison ivy on stones.[8]

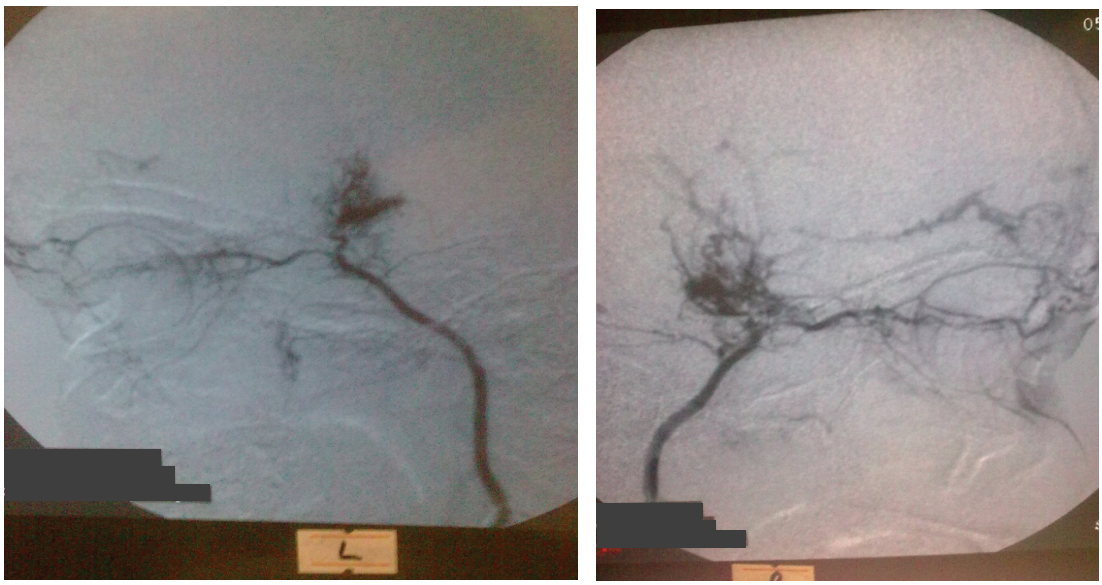

Figure 2. Carotid angiogram showing bilateral (A - left, B – right) terminal narrowing and complete block of the internal carotid arteries with characteristic collateral formation at the base of the brain (Suzuki Grade III-IV).

Although initially thought to affect the Japanese population only, moyamoya disease is now reported from other countries as well.[9-14] Unlike moyamoya disease cases from Japan, a lack of bimodal distribution and a better response to surgical treatment was noted in cases from the USA.[14] Commonly, moyamoya disease has four types of manifestations, i.e., ischemic stroke, mainly in children (63.4%), hemorrhagic stroke, mainly in adults (21.6%), seizure disorder (7.6%), and various other presentations (7.5%).[15] In another review[16], patients experienced ischemic stroke in 50-75%, transient ischaemic attacks (including drop attacks) in 50-75%, and intracerebral haemorrhage in 10-40% of cases. Presentation with choreiform movements is reported in children[10,17] and is considered a rare presentation in adult cases of moyamoya disease.[16,18] To our knowledge, moyamoya disease manifesting as hemiballismus has not been reported, and vascular parkinsonism has been reported only once.[19] Dilated moyamoya-associated collaterals in the basal ganglia are implicated in development of choreiform movements.[10,17] A similar pathology involving basal ganglia may explain the presentation in our case.

Treatment of moyamoya disease is usually conservative in mild cases and includes antiplatelet agents, control of seizures and other supportive measures. Vasodilators and fibrinolytic agents are not beneficial.[6] Severe cases are treated by direct (anastomosis of superficial temporal artery to middle cerebral artery) or indirect (encephaloduroarteriosynangiosis and encephalomyosynangiosis) bypass surgery or both.[6] The course of choreiform movements is variable, with a majority resolving after revascularization surgery, likely due to reduction in moyamoya-associated collaterals in the basal ganglia.[17] Our patient showed marked improvement in her involuntary movements with mild residual resting tremors. However, her bradykinesia and rigidity showed little improvement.

In conclusion, we report the first case of moyamoya disease from Nepal. The patient's presentation with abnormal movements (hemiballismus and chorea) and parkinsonism was a rare presentation of a rare disease.

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