**Case Report**

**Moyamoya Disease Presenting as Alternating Hemiparesis with Relapsing Remitting Hemichorea: An Unusual Manifestation**

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**Case:** Moyamoya disease (MMD) is a neurological disease involving internal carotid artery (ICA) leading to its occlusion. Among the children, the disease presents as ischemic strokes, whereas in adults, it presents as hemorrhagic strokes. Movement disorder among the MMD is very rare with varied presentation. This article reports a case of 16-year-old girl presented with a history of alternating hemiparesis with recurrent hemichorea with self-remitting tendency. Magnetic resonance angiography brain showed marked-to-complete attenuation of supraclinoid ICA with multiple tortuous collateral vessels replacing the circle of Willis. **Conclusion:** MMD should be kept in the differential diagnosis of children presenting with alternating focal neurological deficit with recurrent movement disorder as movement disorder could be the initial presentation. MMD may present as waxing and waning features of the chorea and neurological deficit.

**KEYWORDS:** Cerebral arterial diseases, infarct, intracranial arterial diseases, movement disorder, Moyamoya disease

**INTRODUCTION**

Moyamoya disease (MMD) is a rare cerebrovascular disorder featured by the stenosis or the progressive occlusion of internal carotid artery (ICA).[1] Recurrent ischemic attack and seizure disorder are the frequent manifestations among the children, whereas hemorrhagic stroke is common among adults.[1-4] Movement disorder is an extremely rare presentation among the patients with MMD with 3%-6% of the cases with varied movement disorder.[1,2,4] Ischemic changes or manipulation in the inhibitory–excitatory circuits interconnecting the basal ganglia and cerebral cortex plays a crucial role for the movement disorder pathology among these patients. There are majority of case reports with hemihemichorea, anterior circulation involvement, alternating hemiparesis, and hemiparesis with hemichorea.[3] Here we report a unique case with alternating hemiparesis with recurrent episodes of hemichorea.

**CASE HISTORY**

A 16-year-old girl, with no significant perinatal, psychomotor developmental and family history, visited with 3-month history of left-sided motor weakness with progressive abnormal involuntary, irregular brief jerky movement of upper limb (UL) and lower limb (LL), accompanied by jerky hesitant speech. No history of dysphagia and nasal regurgitation, visual loss, sensory deficit, or any bladder and bowel disturbances was reported. She was unable to walk or sit without support at the time of presentation, but her orientation and comprehension were intact. History for accompanying seizure, headache, vomiting, or loss of consciousness was absent. These involuntary movements were present at rest but increased with activity and stress. These involuntary movements and weakness improved spontaneously without any treatment over few months. She had a history of generalized tonic–clonic seizure episodes at 4 years of age for which she took phenytoin 200 mg/day. At the age of 10 years, she developed right-sided motor weakness with abnormal brief, jerky irregular movement of right UL and LL with jerky resistant speech, which with 3-month history of left-sided motor weakness with progressive abnormal involuntary, irregular brief jerky movement of upper limb (UL) and lower limb (LL), accompanied by jerky hesitant speech. No history of dysphagia and nasal regurgitation, visual loss, sensory deficit, or any bladder and bowel disturbances was reported. She was unable to walk or sit without support at the time of presentation, but her orientation and comprehension were intact. History for accompanying seizure, headache, vomiting, or loss of consciousness was absent. These involuntary movements were present at rest but increased with activity and stress. These involuntary movements and weakness improved spontaneously without any treatment over few months. She had a history of generalized tonic–clonic seizure episodes at 4 years of age for which she took phenytoin 200 mg/day. At the age of 10 years, she developed right-sided motor weakness with abnormal brief, jerky irregular movement of right UL and LL with jerky resistant speech, which

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evolved over 3 months and subsided over 1.5 years spontaneously without treatment. According to family members, her condition in this episode deteriorated to such an extent that she was unable to perform her activities of daily living. At 12 years of age, she again developed abnormal jerky movements of left UL and LL, associated with motor weakness and speech involvement. It evolved over 3 months with spontaneous recovery. This was the time when the patient presented to our facility for consultation.

Examination suggested brief, involuntary, irregular purposeless, nonrhythmic hyperkinesia of left UL/LL involving distal parts of upper extremity, with repeated twitching and grimacing movements of face, tongue, and lips. Power in the left UL and LL was negative 4/5 with asymmetry of reflexes and left plantar extensor. Rest of the physical examination and higher mental function was normal with no Kayser–Fleischer ring. No derangement in hemogram, kidney function test, liver function test, Antistreptolysin O titer, C-reactive protein, serum ceruloplasmin, and antineutrophil cytoplasmic antibody was reported. No abnormality in electrocardiography, electroencephalography, echocardiography, X-ray chest, and slit lamp examination of the eye was observed.

Non-contrast computerized tomography scan of head was normal. Magnetic resonance imaging (MRI) brain revealed prominences of bilateral Virchow–Robin spaces with narrowing of cavernosal part of ICA with non-visualization of supraclinoid ICA with multiple tiny flow voids in the basal cisterns [Figure 1A and B]. Magnetic resonance angiography (MRA) brain showed marked-to-complete attenuation of supraclinoid ICA with multiple tortuous collateral vessels replacing the circle of Willis and showing the puff of smoke appearance [Figure 2A and B]. There is marked attenuation to non-visualization of the basilar and bilateral posterior cerebral arteries [Figure 3]. The patient was started on low-dose haloperidol 2.5 mg bd (dose) with symptomatic management and she improved completely this time also in a couple of months.

**DISCUSSION**

MMD is a rare disease with higher incidence among the females and Asians. Also familial linkage of the disease is noted in 15% of the population, but was absent in our case. MMD causes ischemia in the territory of ICA with common manifestations of speech difficulty and hemiparesis. Although rare, it commonly leads to stroke among the children. Its low incidence always makes it prone to be missed during the presentation as in our case where the diagnosis was made during the third attack, similar to the findings suggested by Borah et al. Epilepsy is another important presentation of MMD; however, it is reported in very few cases. Movement disorder, an important clinical presentation especially among the pediatric patients, could be the initial manifestation or could be observed during the disease course with varied picture ranging from dystonia, chorea, or dyskinesia. Irregular jerks were noted to be the most common involuntary movements by Kraemer et al., which were similar to our case. Baik et al. reported chorea as the most common movement disorder presentation. Sudden onset of hemichorea most frequently occur because of a lacunar infarction or neoplasm in the contralateral basal ganglia or subthalamic region. In patients with MMD, dilated Moyamoya collateral vessels compress the basal ganglia

![Figure 1: (A) and (B) MRI brain showing prominences of bilateral Virchow–Robin spaces and narrowing of cavernosal part of ICA with non-visualization of supraclinoid ICA. There were multiple tiny flow voids in the basal cisterns](image-url)
or cause local ischemia and can cause choreiform dyskinesia.[2] In our case, the purposeless movements were increased on activity or stress. Baik et al.[2] found that movement disorder increased on emotional stress and exercise in many of their reported cases. The jerky movements at the time of presentation could be misdiagnosed as epilepsy. Presentation of transient movement disorder among the patient with MMD can be explained by the fact that ischemic damage increases membrane excitability, release of excitatory neurotransmitters and hyperexcitability leads to the abnormal body movement.[8]

**CONCLUSION**

MMD should be kept in the differential diagnosis of children presenting with the alternating focal neurological deficit with recurrent episodes of movement disorder as movement disorder could be the initial presentation. This case provides a glimpse that MMD may present as waxing and waning features of the chorea and neurological deficit. Early disease diagnosis is crucial for timely intervention as recurrent strokes may cause permanent neurological deficit.

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**Conflicts of interest**

There are no conflicts of interest.

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