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Case Report

Late-onset, first-ever involuntary movement after successful surgical revascularization for pediatric moyamoya disease – Report of two cases

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ABSTRACT

Background: A small number of children with Moyamoya disease develop involuntary movements as an initial presentation at the onset, which usually resolves after effective surgical revascularization. However, involuntary movements that did not occur at the onset first occur after surgery in very rare cases. In this report, we describe two pediatric cases that developed involuntary movements during the stable postoperative period after successful surgical revascularization.

Case Description: A 10-year-old boy developed an ischemic stroke and successfully underwent combined bypass surgery. However, he developed chorea six months later. Another 8-year-old boy developed a transient ischemic attack and successfully underwent combined bypass surgery. However, he developed chorea three years later. In both cases, temporary use of haloperidol was quite effective in resolving the symptoms, and involuntary movements did not recur without any medication during follow-up periods of up to 10 years.

Conclusion: Postoperative first-ever involuntary movement is very rare in pediatric moyamoya disease, and the underlying mechanisms are still unclear, but a temporary, reversible imbalance of excitatory and inhibitory circuits in the basal ganglia may trigger the occurrence of these rare symptoms. Careful follow-up would be mandatory.

Keywords: Bypass surgery, Haloperidol, Involuntary movement, Late-onset, Moyamoya disease

INTRODUCTION

In pediatric moyamoya disease, involuntary movements are known to occur very rarely as the initial presentation, but the underlying mechanisms are still obscure.[8,15] Surgical revascularization is accepted as useful to improve cerebral hemodynamics and resolve involuntary movement.[8] However, involuntary movements that did not develop before surgery may first appear postoperatively in very rare cases.[1,4,14]

We herein report two pediatric cases that developed chorea during the stable postoperative period after successful cerebral revascularization. Temporary use of haloperidol was quite effective in resolving the symptoms.

CASE PRESENTATION

Case 1

A 10-year-old boy suddenly developed visual field disturbance and cognitive dysfunction and was diagnosed with Moyamoya disease. Neurological examination on admission revealed right homonymous hemianopsia, sensory aphasia, apraxia, and agnosia. Brain magnetic resonance imaging (MRI) showed multiple infarctions in the bilateral deep frontal lobes and cortical infarction in the left occipital lobe and right temporoparietal lobes. Brain magnetic resonance angiography (MRA) demonstrated severe stenosis in the terminal portion of the internal carotid arteries (ICAs) and the posterior cerebral arteries on both sides. Single-photon emission computed tomography (SPECT) showed reduced cerebral blood flow (CBF) in the bilateral cerebral hemispheres [Figures 1a-c]. He underwent superficial temporal artery to middle cerebral artery (STA-MCA) anastomosis and encephalo-duro-myo-arterio-pericranial synangiosis (EDMAPS) on both sides.[7,9] Postoperative course was uneventful, and no further cerebrovascular events occurred. Postoperative radiological examinations at three months demonstrated well-developed surgical collaterals through direct and indirect bypass and a marked improvement of CBF except in the area of cerebral infarction [Figures 1d-f].

Six months after surgery, however, he developed involuntary movements in the tongue and left extremities. The involuntary movements were sustained and very similar to the chorea. Brain MRI/MRA showed no new abnormalities. He was treated with clonazepam for ten days, but his symptoms did not improve. Then, he was treated with haloperidol (1.5 mg/day); after that, his symptoms gradually improved and completely disappeared after one month. Although haloperidol was tapered and discontinued, involuntary movements have not recurred for ten years.

Case 2

An 8-year-old boy developed a headache attack on the right side and transient weakness of the left extremities and was diagnosed with Moyamoya disease. Neurological examination on admission revealed no abnormalities. Brain MRI showed no parenchymal lesions. Brain MRA showed severe stenosis of the terminal portion of the ICAs on both sides. Brain SPECT showed a marked CBF decrease in the territories of the bilateral cerebral hemispheres, especially in the frontal lobes [Figures 2a-c]. He underwent STA-MCA anastomosis and EDMAPS on both sides. The postoperative course was uneventful, and his attacks completely disappeared. Postoperative examination at four months later showed well-developed surgical collaterals and normalization of CBF [Figures 2d-f].

Three years after surgery, however, he developed chorea-like involuntary movements in the neck and left extremities. Brain MRI/MRA showed no new abnormalities. Blood flow measurement demonstrated a normal distribution of CBF [Figure 2g]. He was treated with haloperidol (1.5 mg/day). His symptoms gradually improved and completely disappeared after three months. Although haloperidol was tapered and discontinued, he has been free from involuntary movements for these two years.

DISCUSSION

According to the previous reports, 3–6% of moyamoya children present with involuntary movements as an initial symptom. A majority of involuntary movements are chorea, but some patients develop dystonia or dyskinesia.[2] The pathogenesis is still unclear, but previous investigators have speculated that cerebral ischemia in the basal ganglia[3] or the basal ganglia–thalamocortical circuits may be responsible for involuntary movements in pediatric moyamoya disease.[5,10] Alternatively, the markedly dilated, pulsatile moyamoya vessels may exhibit some mechanical impact on the basal ganglia, causing involuntary movements.[1]

In this report, we describe two children who first developed involuntary movements six months or three years after successful surgical revascularization. In both cases, there were no episodes of involuntary movements before surgery, indicating that involuntary movements emerged for the 1st time several months or years after surgery. Chorea-like involuntary movements occurred in the tongue and unilateral limbs in one case and the neck and unilateral limbs in another. Their postoperative course was uneventful, and cerebrovascular events had completely resolved after surgery. Follow-up radiological examinations several months after surgery showed a marked improvement in cerebral hemodynamics in response to the well-developed surgical collaterals.

To the best of our knowledge, there are only four similar cases in the literature. Thus, the late-onset, first-ever involuntary movement in pediatric moyamoya disease would be quite rare.[1,4,14] Clinical data for the four previously reported cases and the two cases reported here are summarized in Table 1. All of them are pediatric cases. Their ages ranged from 4 to 12 years. The four cases reported previously were all girls, whereas the two cases reported here were boys. Initial symptoms at initial presentation included a transient ischemic attack in four cases, seizure in one, and ischemic stroke in one, suggesting that their gender, age, onset type and the presence of preoperative cerebral infarction were not related to the development of postoperative first-ever involuntary movements. They developed postoperative

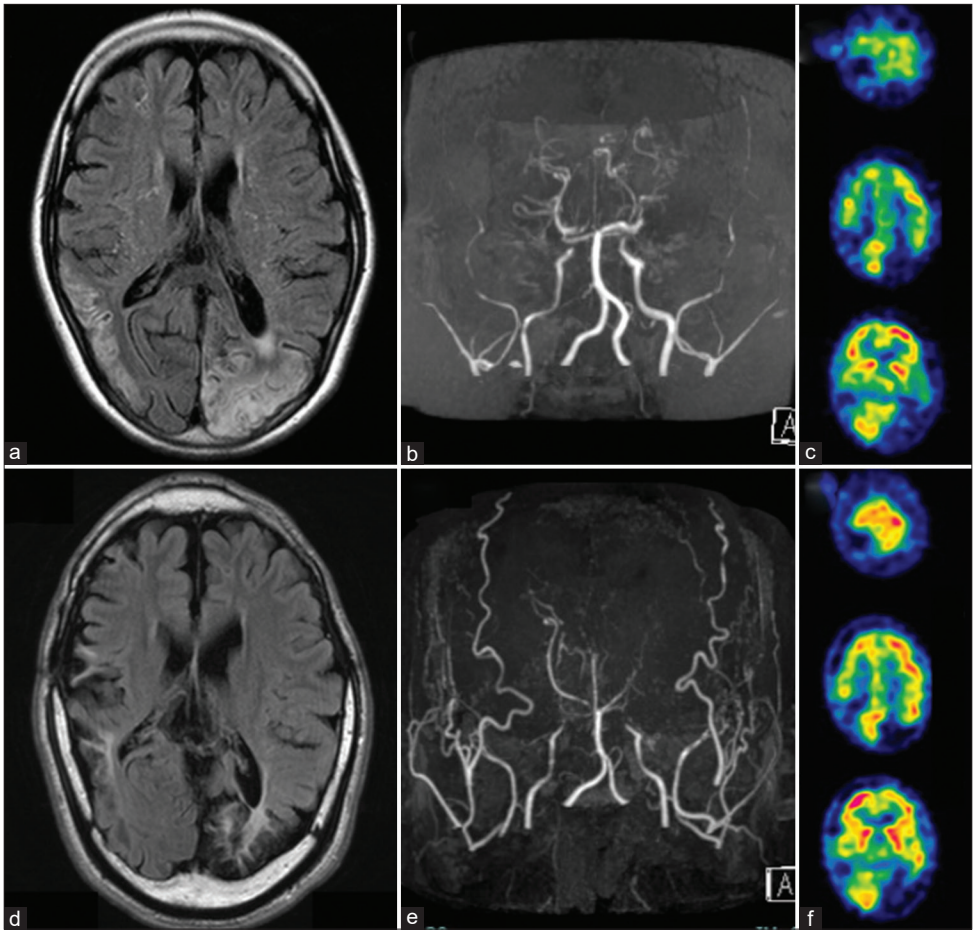

Figure 1: Pre- (a-c) and postoperative (d-f) radiological findings of case 1. (a) Fluid-attenuated inversion recovery (FLAIR) image demonstrated small cerebral infarction in the bilateral deep frontal lobes, the left occipital lobe, and the right temporoparietal lobes. (b) Magnetic resonance angiography (MRA) showed severe stenosis of the terminal portion of the internal carotid arteries and the posterior cerebral arteries on both sides. (c) Blood flow study demonstrated a marked decrease in cerebral blood flow (CBF) on both sides. (d) Postoperative FLAIR image showed no newly developed cerebral infarction. (e) Postoperative MRA showed a marked increase in the caliber of superficial temporal arteries, middle meningeal arteries, and deep temporal arteries on both sides, indicating the sufficient development of surgical collaterals through direct and indirect bypass procedures. (f) Postoperative single-photon emission computed tomography demonstrated sufficient improvement of CBF in the vital tissues except for cerebral infarction.

involuntary movements three months–5 years after surgical revascularization. Their symptoms simulated chorea, and the locations of symptoms were unilateral extremities in the majority of cases. In our two cases, however, the involuntary movements also involved the tongue or neck, so the symptoms of involuntary movements should be carefully observed in such cases. In fact, we have reported two children with Moyamoya disease who presented with oromandibular dyskinesia as an initial symptoms.[6]

In five of the six cases, radiological findings were described at the onset of postoperative involuntary movements, and no new lesions were not found on MRI. Therefore, the newly developed cerebral infarction or hemorrhage is unlikely to be the cause of these rare symptoms. Sugita *et al*. found reduced CBF in the unilateral basal ganglia,[14] but CBF was within normal limits in our two cases, so cerebral ischemia in the basal ganglia would not be the common factor, at least among patients with the first-ever involuntary movements after surgery. More interestingly, Sugita *et al*. reported a transient, increased uptake of ^{18}F-fluorodeoxyglucose (^{18}F-FDG) in the unilateral striatum on ^{18}F-FDG positron emission tomography.[14] It is obscure whether this finding is the cause or result of involuntary movements. Still, it may be the clue to clarify the underlying mechanism of involuntary movements that develop after surgery. The information would also be

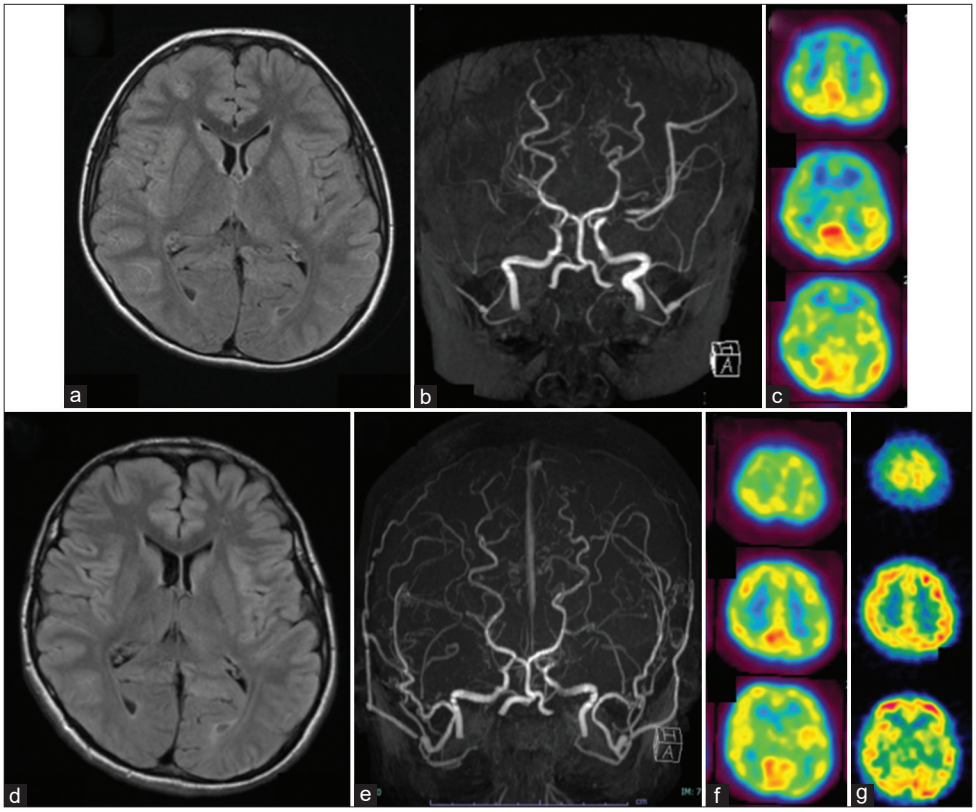

Figure 2: Pre- (a-c) and postoperative (d-f) radiological findings of case 2. (a) Fluid-attenuated inversion recovery (FLAIR) image demonstrated no parenchymal lesions. (b) Magnetic resonance angiography (MRA) showed severe stenosis of the terminal portion of the internal carotid arteries (ICAs) on both sides. (c) A blood flow study demonstrated a marked decrease in cerebral blood flow (CBF) in the territory of the ICA, especially in the frontal lobes on both sides. (d) Postoperative FLAIR image showed no newly developed cerebral infarction. (e) Postoperative MRA showed a marked increase in the caliber of superficial temporal arteries, middle meningeal arteries, and deep temporal arteries on both sides, indicating the sufficient development of surgical collaterals through direct and indirect bypass procedures. (f) Postoperative single-photon emission computed tomography demonstrated sufficient improvement of CBF on both sides. (g) A blood flow study at the onset of involuntary movements revealed no abnormal distribution of CBF.

useful to evaluate the mechanisms of involuntary movements as the initial symptom of Moyamoya disease.

In one of the six patients, the involuntary movements resolved spontaneously, whereas haloperidol was effective in completely resolving it in the other five cases.[1,4,14] On the other hand, clonazepam was not effective in our first case. It is known that a certain subgroup of adult patients with stroke may develop involuntary movements. Still, their prognosis is not so favorable, being quite different from that of postoperative first-ever involuntary movements in pediatric moyamoya disease.[11,13] Thus, Ristic *et al*. (2002) reported that haloperidol could completely resolve the symptoms in only 15 (56%) of 27 patients with stroke-related chorea.[12] Haloperidol has a high affinity for the dopamine D2 receptor and is widely used in psychiatric disorders. Therefore, haloperidol may resolve this rare symptom by largely modulating the imbalance between the dopaminergic and GABAergic circuits in the basal ganglia.

In the four cases reported previously, there is no description about whether the involuntary movements recurred during follow-up periods or not. In our two cases, however, the symptoms did not recur when they were followed for 2–10 years, although haloperidol was discontinued. The fact strongly suggests that the late-onset, first-ever involuntary movements after surgery for moyamoya disease may result from a temporary, reversible dysfunction of the extrapyramidal system. Since this extremely rare symptom has only occurred in children but not in adults, it may involve some kinds of pediatric-specific pathophysiology.

Table 1: Summary of reported cases that presented with first-ever involuntary movement after successful surgical revascularization for pediatric moyamoya disease.

Case No.	Authors	Year	Age	Sex	Diagnosis at onset	Surgical Treatment	First-ever involuntary movement after surgery							
							Timing after surgery	Locations	Symptoms	Radiological findings	Treatments	Outcome	Follow-up	Recurrence
1	Ahn *et al.*	2013	11	F	TIA	Indirect bypass	3 years	Unspecified	Chorea	Unknown	None	Resolved (2 days)	None	Unknown
2	Sugita *et al.*	2016	10	F	TIA	STA-MCA +EMS (bilat.)	3 months	Left face and arm	Chorea	Glucose hypermetabolism and dilated MMV in the right striatum	Haloperidol	Resolved (2 months)	None	Unknown
3	Sugita *et al.*	2016	12	F	TIA	STA-MCA +EMS (bilat.)	5 years	Right arm and leg	Chorea	Hypoperfusion, glucose hyper-metabolism, and dilated MMV in the left striatum	Haloperidol	Improved (unknown)	None	Unknown
4	Enriquez-Ruano *et al.*	2021	4	F	Seizure	EDAS (Lt)	2 years	Bilateral arms and legs	Chorea	Dilated MMV in the left striatum	Haloperidol	Resolved (unknown)	None	Unknown
5	Case 1	2023	10	M	Ischemic stroke	STA-MCA +EDMAPS (bilat.)	6 months	Toungue and left arm	Chorea	No new lesions	Haloperidol	Resolved (one month)	10 years	No recurrence
6	Case 2	2023	8	M	TIA	STA-MCA +EDMAPS (bilat.)	3 years	Neck, left arm and leg	Chorea	No new lesion and normal CBF	Haloperidol	Resolved (3 months)	2 years	No recurrence

M: Male, F: Female, TIA: Transient ischemic attack, STA-MCA: Superficial temporal artery to middle cerebral artery, EMS: Encephalo-myo-synangiosis, EDAS: Encephalo-duro-arterio-synangiosis, EDMAPS: Encephalo-duro-myo-arterio-pericranial synangiosis, BILAT: Bilateral, CBF: Cerebral blood flow, MMV: Moyamoya vessels

CONCLUSION

Late-onset, first-ever involuntary movements after surgery are quite rare in pediatric moyamoya disease. The underlying mechanisms are still unclear, but a temporary, reversible imbalance of excitatory and inhibitory circuits in the basal ganglia may trigger the occurrence of these rare symptoms. It is quite important to carefully follow up with pediatric patients for a long time, even after successful surgical revascularization.

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Ethical approval

Not applicable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript, and no images were manipulated using AI.

REFERENCES

1. Ahn ES, Scott RM, Robertson RL Jr., Smith ER. Chorea in the clinical presentation of moyamoya disease: Results of surgical revascularization and a proposed clinicopathological correlation. J Neurosurg Pediatr 2013;11:313-9.
2. Baik JS, Lee MS. Movement disorders associated with moyamoya disease: A report of 4 new cases and a review of literatures. Mov Disord 2010;25:1482-6.
3. Bhatia KP, Marsden CD. The behavioural and motor consequences of focal lesions of the basal ganglia in man. Brain 1994;117:859-76.
4. Enriquez-Ruano P, Navarro CE, Penagos N, Espitia OM. Late-onset chorea after cerebral revascularization as a clinical manifestation of moyamoya disease. Neurol Sci 2021;42:3027-30.
5. Gonzalez-Alegre P, Ammache Z, Davis PH, Rodnitzky RL. Moyamoya-induced paroxysmal dyskinesia. Mov Disord 2003;18:1051-6.
6. Koga Y, Kashiwazaki D, Hori E, Akioka N, Kuroda S. Oro-mandibular dystonia in pediatric moyamoya disease: Two cases report. Surg Neurol Int 2021;12:449.
7. Kuroda S, Houkin K, Ishikawa T, Nakayama N, Iwasaki Y. Novel bypass surgery for moyamoya disease using pericranial flap: Its impacts on cerebral hemodynamics and long-term outcome. Neurosurgery 2010;66:1093-101; discussion 1101.
8. Kuroda S, Houkin K. Moyamoya disease: Current concepts and future perspectives. Lancet Neurol 2008;7:1056-66.
9. Kuroda S, Nakayama N, Yamamoto S, Kashiwazaki D, Uchino H, Saito H, *et al*. Late (5-20 years) outcomes after STA-MCA anastomosis and encephalo-duro-myo-arterio-pericranial synangiosis in patients with moyamoya disease. J Neurosurg 2020;134:909-16.
10. Lee JY, Kim SK, Wang KC, Chae JH, Cheon JE, Choi JW, *et al*. Involuntary movement in pediatric moyamoya disease patients: Consideration of pathogenetic mechanism using neuroimaging studies. Childs Nerv Syst 2014;30:885-90.
11. Park J. Movement disorders following cerebrovascular lesion in the basal ganglia circuit. J Mov Disord 2016;9:71-9.
12. Ristic A, Marinkovic J, Dragasevic N, Stanisavljevic D, Kostic V. Long-term prognosis of vascular hemiballismus. Stroke 2002;33:2109-11.
13. Siniscalchi A, Gallelli L, Labate A, Malferrari G, Palleria C, Sarro GD. Post-stroke movement disorders: Clinical manifestations and pharmacological management. Curr Neuropharmacol 2012;10:254-62.
14. Sugita Y, Funaki T, Takahashi JC, Takagi Y, Fushimi Y, Kikuchi T, *et al*. Reversible striatal hypermetabolism in chorea associated with moyamoya disease: A report of two cases. Childs Nerv Syst 2016;32:2243-7.
15. Suzuki J, Takaku A. Cerebrovascular "moyamoya" disease. Disease showing abnormal net-like vessels in base of brain. Arch Neurol 1969;20:288-99.

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