Clinical features of hemichorea-hemiballism: A stroke-related movement disorder

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Abstract

We examined pathogenesis and clinical features of three hemichorea-hemiballism (HCHB) cases. We studied their age, magnetic resonance imaging results, vascular risk factors, management, and outcomes. One man and two women (aged 74-86 years) demonstrated acute onset of HCHB, lasting for at least several months. Patients had one or more vascular risk factors, including hypertension and diabetes. All patients presented subacute or old infarction in the basal ganglia with contralateral symptoms. We administered clonazepam (0.5-1 mg/day), haloperidol (0.375-0.75 mg/day), or both as necessary and observed symptom-control. Vascular lesions in the basal ganglia were a contributing factor. Symptoms were controlled using pharmacotherapy with gamma-aminobutyric acid-agonist (clonazepam) or anti-dopaminergic (haloperidol) medication.

Introduction

Strokes and movement disorders are relatively common and demand the attention of neurologists and primary care physicians. However, stroke-related movement disorders, such as hemichorea-hemiballism (HCHB), occur only in a small percentage of cases.[1-3] Although its pathogenesis remains unclear, research previously conducted in the field suggests that the mechanisms involve the motor circuitry of the basal ganglia, despite the absence of a specific and reliably predictive anatomical location. We examined the clinical course of three HCHB cases contralateral to the vascular insult.

Materials

We examined the age, sex, MRI results, vascular risk factors, management, and outcomes for three cases of HCHB.

Ballism is a condition that causes a violent, irregular, large-amplitude, involuntary movement that primarily involves the proximal extremities. It can be categorized as a fast form of chorea.[1-2]

Chorea presents as a condition that causes rapid, irregular, and involuntary movement that typically involves both proximal and distal muscles.[1-2]

Case Report #1

A 74-year-old man presented with continuous hemichoreic movements on his left arm, left leg, and tongue for ten to fourteen days before visiting our clinic. His blood sugar (BS) level on the first visit was 151 mg/dl. Brain MRI showed an old cerebral infarct in the right globus pallidus, which has been present for the past four years (Figure 1A,B). In addition, he had a transient cerebral ischemic attack that affected his left arm eight years ago. His vascular risk factors included hypertension and diabetes (BS=143 mg/dl, HbA1c=7.0%). Clonazepam (0.5 mg/day) administration was ineffective. His dyskinesia ceased three and a half months after onset once the clonazepam dose was increased up to 1.0 mg/day and haloperidol (0.75 mg/day) was added (Figure 2A).

Case Report #2

An 81-year-old woman suddenly presented with continuous hemiballism/choreic movements on her left arm and leg for one week before visiting our clinic. Brain MRI taken seven days after the onset showed subacute cerebral infarcts in the right globus pallidus (Figure 1C-E). Her vascular risk factors included hypertension but no diabetes. Her symptoms ameliorated after Haloperidol (0.75 mg/day) administration. Eighteen months later, she presented symptoms of oral dyskinesia. We attempted to eliminate haloperidol to treat her condition but could not prevent her symptoms from relapsing. Three years after its onset, we reduced the dose without relapsing the previous symptoms; however, she continued to demonstrate oral dyskinesia (Figure 2B).

Case Report #3

An 86-year-old woman presented with continuous hemiballism/choreic movement on her left leg and her left arm for one to two months before visiting our clinic. Brain MRI showed periventricular hyperdensity, along with old infarcts in the right globus pallidus and

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putamen (Figure 1F-H). She had hypertension and vascular dementia but no diabetes. Her symptoms completely disappeared after receiving clonazepam (0.5 mg/day) administration. We aimed to reduce its dose from daily to every other day (0.25 mg/day) for several months after the onset of her condition due to her drowsiness (Figure 2C).

Results

Among the three HCHB cases, all cases demonstrated a subacute or old infarction in the basal ganglia contralateral to the symptoms. Due to the severity of daily life disruptions caused by HCHB, which lasted at least several months in one case, we treated them with clonazepam (0.5-1 mg/day), haloperidol (0.375-0.75 mg/day), or both as necessary to control the observed symptoms. Symptoms were controlled using pharmacotherapy with gamma-aminobutyric acid (GABA)-agonist (clonazepam) or anti-dopaminergic (haloperidol) activity. We attempted withdrawal when the symptoms were brought under control due to the observed side effects, such as oral dyskinesia and drowsiness, but encountered difficulties due to recurrence.

Discussion

HCHB is a hyperkinetic movement disorder characterized by unilateral involuntary movement. Apart from cerebral

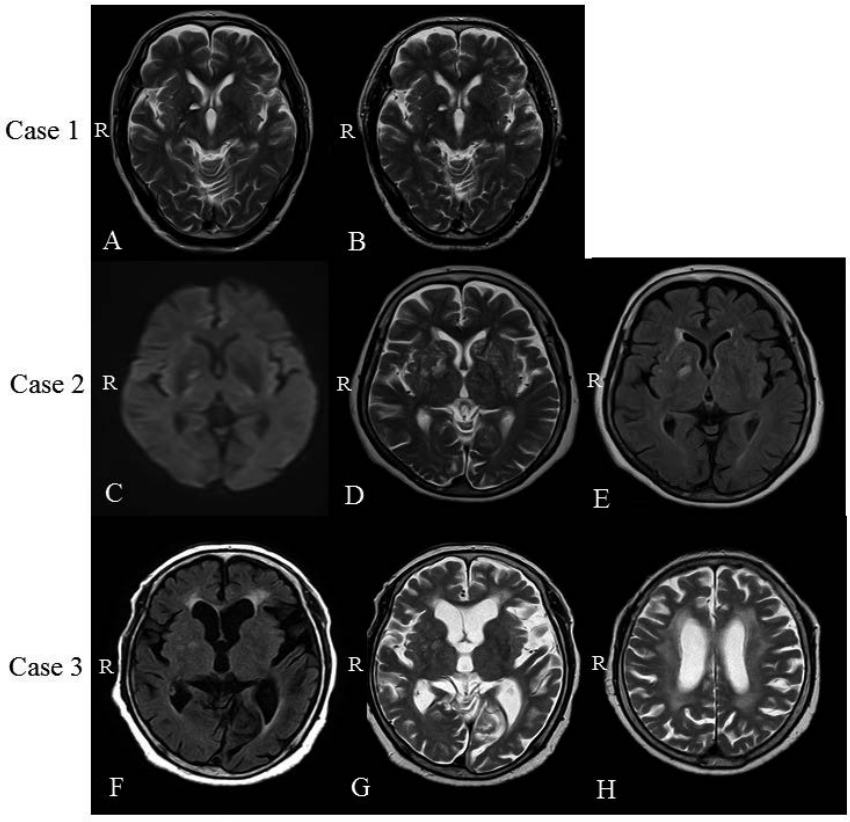

Figure 1. Magnetic resonance image findings for the three hemichorea-hemiballism cases. In case 1, brain magnetic resonance image (MRI) shows the old cerebral infarct in the right globus pallidus present for the past four years (A: T2 weighted MRI image (T2WI)at the onset, B: T2WI at the four years before the onset). In case 2, MRI shows the subacute cerebral infarcts in the right globus pallidus (C; Diffusion weighted image(DWI), D; T2WI, and E; Fluid attenuated inversion recovery(FLAIR)). In case 3, MRI shows periventricular hyperdensity (PVH), old infarcts in right globus pallidus and putamen (F; FLAIR, G; T2WI, and H; T2WI).

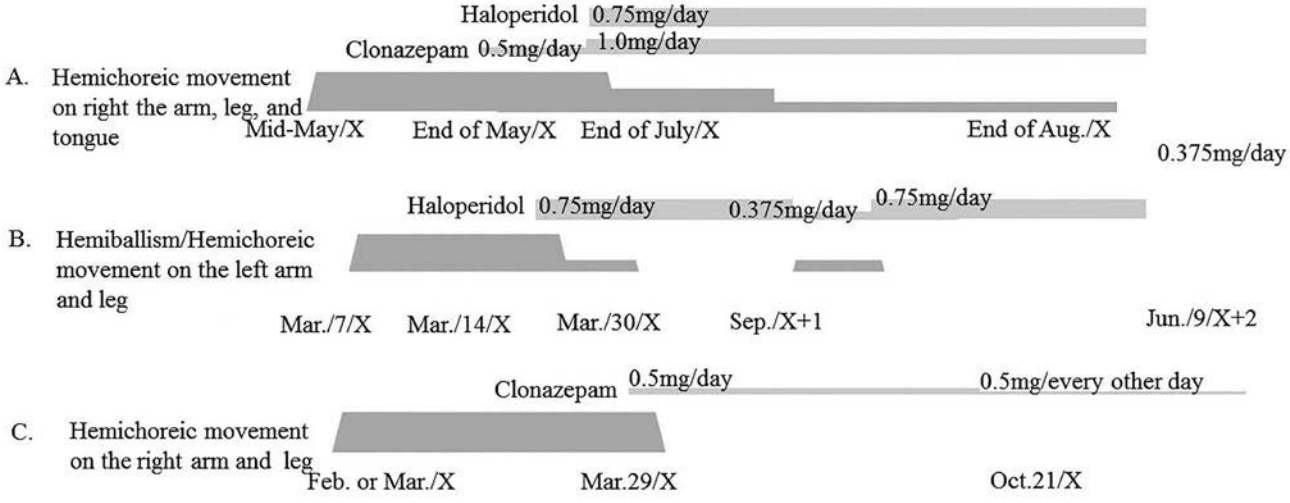

Figure 2. Clinical courses of the three hemichorea-hemiballism (HCHB) cases (A; case 1, B; case 2, C; case 3) X means the year when HCHB occurred.

vascular diseases, HCHB occurs unilaterally due to nonketotic hyperglycemia.[4-6] Among the three patients, one had diabetes; however, his blood sugar levels were not high enough to be considered HCHB-associated. Therefore, our cases were considered purely stroke-related.

Here, lesions in the globus pallidum or putamen contralateral to the dyskinesia were observed in all cases. The pathogenesis for stroke-related HCHB is unclear; previous studies have suggested mechanisms involving the basal ganglia motor circuitry, despite the absence of a specific and reliably predictive anatomical location.[1-3] Grandas[7] stated that in clinical radiological series of hemiballism, which are classically related to lesions in the subthalamic nucleus, most patients had lesions outside this nucleus, mainly in other basal ganglia structures. It has also been suggested that abnormal neuronal firing patterns in the internal segment of the globus pallidus may be related to the pathogenesis of hemiballism.[7] Furthermore, Zijlmans[8] reviewed patients with vascular-related chorea affecting one side of the body in which the lesions are most frequently found in the thalamus and lentiform nucleus.

HCHB is also known to be a hyperkinetic movement disorder in diabetic patients. In diabetic hemichorea-hemiballism, striatal hyperintensity is typically seen on T1-weighted magnetic resonance imaging.[4-6] These reports support that HCHB is associated with basal ganglia, as demonstrated in our three cases.

In some cases, HCHB can occur immediately after an acute stroke, whereas others can experience a progressive or delayed onset.[1] Among our HCHB cases, the MRI for case 2, which was taken seven days after the onset of HCHB, showed the subacute cerebral infarcts in the globus pallidus contralateral to the symptoms, and in cases 1 and 3, old infarction was seen in the basal ganglia contralateral to the symptoms. These observations suggest that the onset of HCHB after stroke varies from case to case.

We successfully controlled the symptoms using pharmacotherapy with GABA-agonist (clonazepam) or anti-dopaminergic (haloperidol) activity. However, due to side effects, such as oral dyskinesia and drowsiness, we attempted withdrawal in situations where the symptoms were controlled but could not do so due to recurrence.

Conclusions

Among the three HCHB patients, all showed subacute or old infarction in the basal ganglia contralateral to the symptoms. Symptoms were controlled using pharmacotherapy with GABA-agonist (clonazepam) or anti-dopaminergic (haloperidol) activity.

References

1. Mehanna R, Jankovic J. Movement disorders in cerebrovascular diseases. Lancet Neurol 2013;12:597-608.
2. Nakawah MO, Lai EC. Post-stroke dyskinesias Neuropsychiatr Dis Treat 2016;12:2885-93.
3. Laganiere S, Boes AD, Fox MD. Network localization of hemichorea-hemiballismus Neurology 2016;86: 2187-95.
4. Hashimoto T, Hanyu N, Yahikozawa H, Yanagisawa N. Persistent hemiballism with striatal hyperintensity on T1-weighted MRI in a diabetic patient: a 6-year follow-up study. J Neurol Sci 1999;165:178-81.
5. Kitagawa M, Yamanaka Y, Adachi T, et al. Diabetic hemichorea-hemiballism after prompt improvement in hyperglycemia. Intern Med 2017;56:3 073-6.
6. Son BC, Choi JG, Ko HC. Globus pallidus internus deep brain stimulation for disabling diabetic hemiballism/ hemichorea. Case Rep Neurol Med 2017;2017:2165905.
7. Grandas F. Hemiballismus. Handb Clin Neurol 2011;100:249-60.
8. Zijlmans JC. Vascular chorea in adults and children. Handb Clin Neurol 2011;100:261-70.