Neurology Asia 2022; 27(2) : 489 – 491

Hemichorea without MRI findings accompanied by significant internal carotid artery stenosis: A case report

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Abstract

Hemichorea is a clinical syndrome characterized by rapid, purposeless, irregular, and uncontrollable choreographic movements of the limbs and/or face, and it is frequently associated with cerebral infarction. We report here a case of acute hemichorea, as the first sign of severe stenosis of the contralateral terminal internal carotid artery without cerebral infarction, with complete remission after the endovascular intervention.

Keywords: Hemichorea, cerebrovascular disease, movement disorder

INTRODUCTION

Hemichorea, a rare motor disorder, is characterized by involuntary choreographic movements of the limbs and head, with neck twisting, shoulder shrugging, intermittent finger flexion, and extension, hand swinging, and arm extension.[1] Contralateral stroke lesions in the subthalamic nucleus and basal ganglia are the most common cause of this movement disorder.[2] Vascular cause of hemichorea without an imaging lesion are rare. We present here a case of acute onset hemichorea that was the first presentation of severe stenosis of the contralateral terminal internal carotid artery without cerebral infarction and was in full remission after the endovascular procedure.

CASE REPORT

A 59-year-old Chinese woman with hypertension for 5 years and no history of psychotropic medication use was admitted for intermittent 'seizures' with involuntary movement of her left arm and leg for 5 days. She also denied taking any illicit drugs or prolonged use of over the counter medications, there was no history of neurological illness in her family.

Neurological evaluation revealed hemiballismus and chorea-like movements in the left upper and lower limbs. There was no pronator muscle drift, and the muscle power and sensation were normal in the upper and lower extremities on both sides. Cerebellar and cranial nerve examinations were also normal. She was normoglycemic. There was no evidence of hemorrhage, infarct, or mass effects in her CT brain scan. The patient was admitted to the neurology ward for further investigation. Laboratory tests included ceruloplasmin, screening for human immunodeficiency virus and syphilis infection were negative. The electrolytes and glucose level were normal. Magnetic resonance imaging (MRI) and diffusion-weighted imaging (DWI) of the brain did not show any evidence of ischemic infarct. Computerized tomography angiography of the neck and head revealed severe stenosis of the terminal internal carotid artery. There were no significant abnormalities in the rest of the cerebral arteries. Cranial CT perfusion imaging showed delayed perfusion of the right basal ganglia. (Figure 1)

Following a 5-day course of aspirin 100 mg and clopidogrel 75 mg, a digital subtraction cerebral angiography (DSA) was performed under general anesthesia. The severe terminal right internal carotid artery stenosis was confirmed. (Figure 2A) A 2.5*15mm Neuroform EZ stent (USA, Stryker) was successfully implanted after balloon dilation and stent angioplasty in the carotid artery. (Figure 2B) Postoperative residual stenosis was less than 20%. On the second day after the operation, the hemichorea was no longer seen and there was no recurrence during the 30-day follow-up.

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Date of Submission: 27 December 2021; Date of Acceptance: 18 January 2022

https://doi.org/10.54029/2022dew

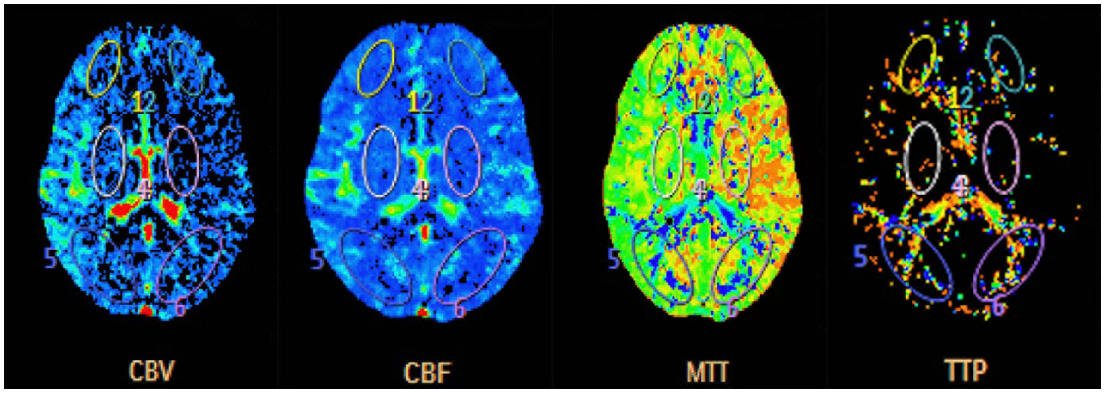

Figure 1. CT perfusion (CTP) showed prolonged mean transit time (MTT) and time to drain (TTP) in the right basal ganglia, while the cerebral blood flow (CBF) and cerebral blood volume (CBV) had no significant difference on both sides.

DISCUSSION

Hemichorea is a term that refers to a collection of clinical symptoms and signs produced by extrapyramidal lesions that are marked by rapid, purposeless, coarse, irregular, involuntary choreiform movements of the lateral limbs and/or face.[3] It is frequently caused by basal ganglia vasculopathy, or as a component of St. Vitus' dance or Huntington's chorea, and rarely by tumors or degenerative disease of the basal ganglia.

An imbalance in the basal ganglia circuit may lead to the onset and development of the chorea. There are numerous nuclei in and around the cerebral cortex, which form a network of projection pathways with the caudate nucleus, putamen, pallidum, substantia nigra, and subthalamus to deliver excitatory or inhibitory stimuli to the brain and regulate the dopaminergic projective bundles from the substantia nigra to the striatum.[4] When these nuclei are damaged, the striatum

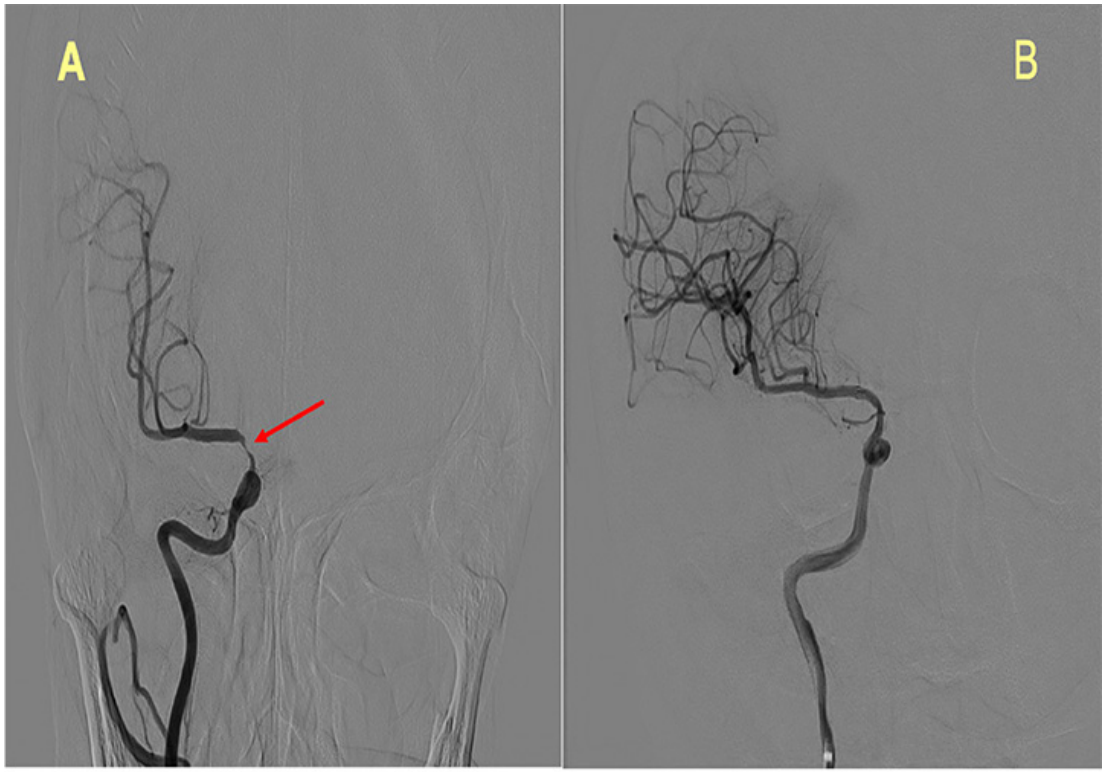

Figure 2: A. DSA revealed severe stenosis of the right internal carotid artery (red arrow). B. Post-stent insertion, the stenosis was reversed.

loses its ability to inhibit the globus pallidus, the lateral part of the released pallidum increases its ability to inhibit the nucleus accumbent, and the downstream hypothalamic inhibition is relieved, resulting in chorea and energy expenditure, acid-base metabolism disruption, and a reduction in GABA synthesis.[5]

In previous studies, CTP has been shown to be able to quantify hemodynamic changes in slow-acting cerebral hypoperfusion.[6] MTT and TTP are sensitive indicators of perfusion anomalies, while CBF and CBV are critical indicators of ischemia damage severity. During the hemodynamic compensatory period, MTT and TTP were frequently moderately prolonged, while CBV and CBF remained normal or slightly decreased, eventually CBV increased.[7] As a result, we were able to conclude that the patient had less perfusion in the right basal ganglia than on the left.

Limb-shaking TIA is an important differential diagnosis of hemichorea to eliminate. Limb shaking TIA is frequently caused by significant stenosis or blockage of the artery supplying the anterior circulation of the contralateral limb. The common symptoms are episodic and brief involuntary jerking of the limbs that typically last less than 5 minutes but can persist up to 1 hour.[8] The shaking is triggered by actions that result in hypoperfusion. Standing upright, moving, neck hyperextension, coughing, and hyperventilation are common provocations.[9] As our patient's symptoms persisted for more than an hour, with no regular jerking movement therefore, limb-shaking TIA was unlikely.

Although our patient did not show MRI changes of ischemia infarct, there was however, imaging evidence of internal carotid artery stenosis, hypoperfusion in the basal ganglia area, and significant improvement after the endovascular procedure. The striatum is known to be particularly vulnerable to ischemic and hypoxic damage in the basal ganglia. After ruling out other possible causes of chorea, it was hypothesized that right internal carotid artery stenosis caused hypoperfusion of the basal ganglia-thalamic-cortical circuit, resulting in cell dysfunction, neurotransmitter imbalance, and the hyperkinetic left sided involuntary movement.

In conclusion, we present here a case of hemichorea due to cerebrovascular disease is not uncommon, clinical cases of vascular-derived hemichorea without an associated MRI infarct lesion which is extremely rare in the literature.[10] However, our patient demonstrate that further cerebrovascular imaging with cerebral angiography and CTP may help to identify the underlying structural abnormalities and demonstrate the ischemia, thus guide further treatment.

DISCLOSURE

Ethics approval: This study was approved by our institutional ethical review board and was conducted by the ethical standards and with the 1964 Declaration of Helsinki and its later amendments. Written informed patient consent was obtained.

Conflict of interests: None

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