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CASE REPORT

A Case of Vascular Hemichorea Responding to Topiramate

Jee-Ae Kim
San Jung
Min-Ju Kim
Seok-Beom Kwon
Sung-Hee Hwang
Ki-Han Kwon

Department of Neurology,
Hallym University College of Medicine,
Seoul, Korea

Although vascular chorea often comes into remission spontaneously, a few patients may remain with persistent movement disorder. Most movements respond well to neuroleptics as well as other antidopaminergic drugs, but some patients show poor responses to those neuroleptics. Topiramate is a widely used of broad-spectrum anticonvulsant possessing a complex mechanism of action. It has been proven to enhance gamma-aminobutyrate acid activity and to be effective in the control of other movement disorders. We describe a 63-year-old woman with intractable vascular hemichorea which was controlled with anti-convulsant, topiramate.

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Key Words: Hemichorea, Anticonvulsant, Topiramate.

Hemichorea refers to irregular, flowing, non-stereotyped, random involuntary movement on one side of the body. Recently, it has been reported that chorea represents the most common post-stroke movement disorder, usually located in the contralateral basal ganglia and subthalamic nucleus.[1] The epidemiology of movement disorders following a stroke is unknown. Although spontaneous recovery usually occurs, in some cases hemichorea or hemiballism can be disabling and irreversible.[2] Most movements respond well to the conventional (haloperidol, perphenazine) or atypical neuroleptics (clozapine, risperidone, olanzapine) as well as other antidopaminergic drugs, but some patients show poor responses to those neuroleptics.[3] Moreover use of these agents is limited due to the side effects of mainly extrapyramidal symptoms (EPS), including tardive dyskinesia and Parkinsonism,[4] besides sedation, hyperarousal, and metabolic dysfunction.[5] Herein we report a patient with intractable vascular hemichorea who was successfully treated with topiramate.

Case Report

A 63-year-old woman presented sudden involuntary, dance-like movements of her right arm and leg. The patient had a 10-year history of diabetes mellitus and hypertension managed with medication. There was no history of prior use of dopamine agonist, antipsychotics and anticonvulsants. Her blood pressure was 130/80 mmHg and blood sugar was 236 mg/dL. Physical and neurological examinations were unremarkable, with the exception of the involuntary movements of the right limbs. Routine biochemical and hematologic tests such as antinuclear antibodies, serum complement level, rheumatoid factor, serum copper, ceruloplasmin, and thyroid function test were normal apart from hyperlipidemia. Psychiatric evaluation and Mini-Mental State Examination (MMSE) score did not reveal remarkable cognitive impairment (score 24/30) or other psychiatric manifestations. Brain MRI revealed acute infarction of the left subthalamic nucleus and left lateral midbrain (Figure 1). The patient was placed on aspirin 100 mg daily, atorvastatin 40 mg daily for acute stroke medication. As a diagnosis of vascular chorea had been established, we tried on risperidone 1 mg bid to control the choreic movements for 3 days, but failed to improve and complained of somnolence. Then we tried use clonazepam 1.5 mg/day for 4 days but the involuntary movements continued. No optimal doses had been achieved because the patient complained of worsening symptoms and somnolence. At 8 days after the onset, administration of topiramate was started at 25 mg twice daily for 3 days and was increased in quantity up to 50 mg bid.

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Corresponding author
San Jung, MD
Department of Neurology,
Hallym University College of Medicine,
948-1 Daerim 1-dong, Yeongdeungpo-gu,
Seoul 150-950, Korea
Tel +82-2-829-5125
Fax +82-2-847-1617
E-mail neurojs@hallym.ac.kr

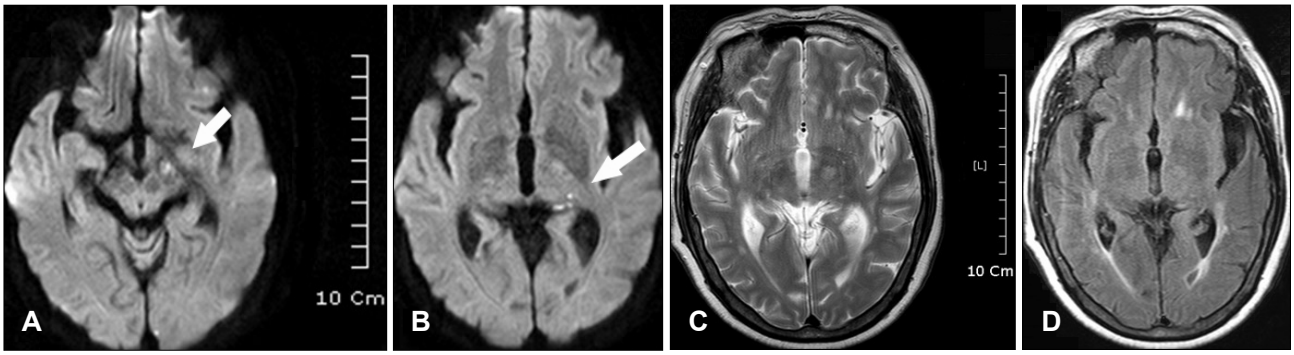

Figure 1. Diffusion weighted MR images shows acute small infarctions on left subthalamic area and upper midbrain (A and B, arrows). These tiny lesions are scarcely visible on T2 weighted (C) or FLAIR image (D). FLAIR: fluid-attenuated inversion-recovery.

The involuntary movements improved to mild degree within 1 week of starting topiramate, and the optimum hemichorea control occurred with the dose of 50 mg of topiramate twice daily. She experienced no side effects with topiramate. However, when she discontinued topiramate due to poor compliance after 1 month of treatment, her movements were aggravated. On the contrary, her hemichorea disappeared almost completely on final examination performed 1.5 months after restart of the medication which maintained at 100 mg per day.

Discussion

Although not completely understood, it suggests that chorea results from the imbalance in the direct and indirect pathways in the basal ganglia circuitry.[4] The disruption gamma-aminobutyric acid (GABA)-ergic transmission of the indirect pathway causes a loss of inhibition on the pallidum, allowing hyperkinetic movements to occur.[4] Anticonvulsants may be useful for the treatment of chorea. In particular, GABA-ergic transmission such as valproate, may be effective in some cases of chorea.[6] Topiramate is a broad-spectrum anticonvulsant and is now widely used for adult and pediatric epilepsy possessing a complex mechanism of action. It has been proven that topiramate can enhance GABA activity and may be effective in control of other movement disorders.[7] Its advantage over other neuroleptics is the lack of EPS in spite of longterm use.[8] Only few case reports have described partial effect of topiramate on control of hemichorea.[9,10] We report a case of intractable hemichorea which was controlled with anti-convulsant, topiramate. In this patient, we have tried conventional drugs which are known to be effective to control choreoathetosis. Whereas several atypical and conventional neuroleptics have failed to control the movement, this patient responded to topiramate which acts with GABA-ergic inhibition. We suggest that topiramate could be a useful therapeutic option in the management of symptomatic generalized chorea. However, prospective clinical trials should be undertaken to confirm these results.

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