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Atypical parkinsonism with marked asymmetry due to a superimposed developmental venous anomaly

Young Eun Huh *MD PhD*, Jonguk Kim *MD*, Won-Chan Kim *MD*

Department of Neurology, CHA Bundang Medical Center, CHA University, Korea

Abstract

Intracranial developmental venous anomalies (DVAs) are the most common cerebral vascular malformation and are usually asymptomatic. Movement disorders are rarely associated with DVAs within basal ganglia regions. We report a case of markedly asymmetric parkinsonism due to unilateral DVA in the basal ganglia, which occurred together with symmetrical nigrostriatal dopaminergic deficits. A 57-year-old woman presented with resting tremor in the right hand lasting for 6 months. She also experienced problems with gait and started falling while walking one month ago. The neurological examination found a resting tremor in the right hand and moderate rigidity and bradykinesia in the right extremities. She reported light headedness on standing up. The patient displayed minimal response to treatment with 300 mg levodopa. The FP-CIT PET scan revealed symmetrical decrease of radiotracer uptake in bilateral basal ganglia. Brain MRI and cerebral angiography identified a large DVA draining the basal ganglia, thalamus, and surrounding deep white matter in the left side.
Conclusion: A DVA may contribute to the prominent asymmetrical manifestation in our patient, in combination with symmetrical dopaminergic loss from neurodegenerative Parkinsonian syndrome. A marked asymmetry in patients with signs of atypical Parkinsonism can be a clue for further imaging investigation to exclude superimposed structural lesions such as DVAs.

Keywords: Developmental venous anomaly, parkinsonism, basal ganglia, dopamine transporter positron emission tomography

INTRODUCTION

Intracranial developmental venous anomalies (DVAs) are the most common cerebral vascular malformation with a prevalence of approximately 3% in autopsy examinations and neuroimaging series.[1,2] The lesion are characterized by the coalescence of radially displayed medullary veins into a dilated collecting vein, which provides normal cerebral venous drainage.[2] DVAs are usually asymptomatic. However, they may be symptomatic due to concomitant vascular malformations, direct compression of adjacent structures, or venous hypertension caused by restricted venous outflow.[2] Manifestations of DVAs include neurological deficits from stroke, seizure, hydrocephalus, and trigeminal neuralgia.[3]

Here, we report a patient with a large DVA in the basal ganglia, who manifested as markedly asymmetrical parkinsonism, along with symmetrical striatal dopaminergic depletion.

CASE REPORT

A 57-year-old woman presented with a 6-month history of progressive tremor in the right hand. She gradually developed gait difficulty and started to fall while walking one month before examination. She had no other medical history except for hypertension for 3 years. She reported light headedness on standing up and denied any urinary problems including incontinence. She had no family history of neurological diseases. Neurological examination identified a resting tremor in the right hand and moderate rigidity and bradykinesia in the right extremities. While walking, she displayed more shuffling steps in the right side than the left, and her arm swings were reduced in the right side. A dopamine transporter PET image with F-18 FP-CIT revealed symmetrical reduction of radiotracer uptake in bilateral striatum (Figure 1A). Brain MRI and cerebral angiography identified a large DVA draining the basal ganglia, thalamus, and surrounding deep white matter in the left side (Figure 1B and C). The patient was treated with 300 mg of levodopa, which slightly alleviated her gait difficulty. She showed no further improvement after increase in levodopa dosage up to 600mg, which only aggravated her postural dizziness.

Written informed consent was obtained from

Address correspondence to: Won-Chan Kim, MD, Department of Neurology, CHA Bundang Medical Center, CHA University, 59 Yatap-ro Bundang-gu, Seongnam-si, Gyeonggi-do, 13496, Korea. Tel: 82-31-780-5482, E-mail: wckim@cha.ac.kr

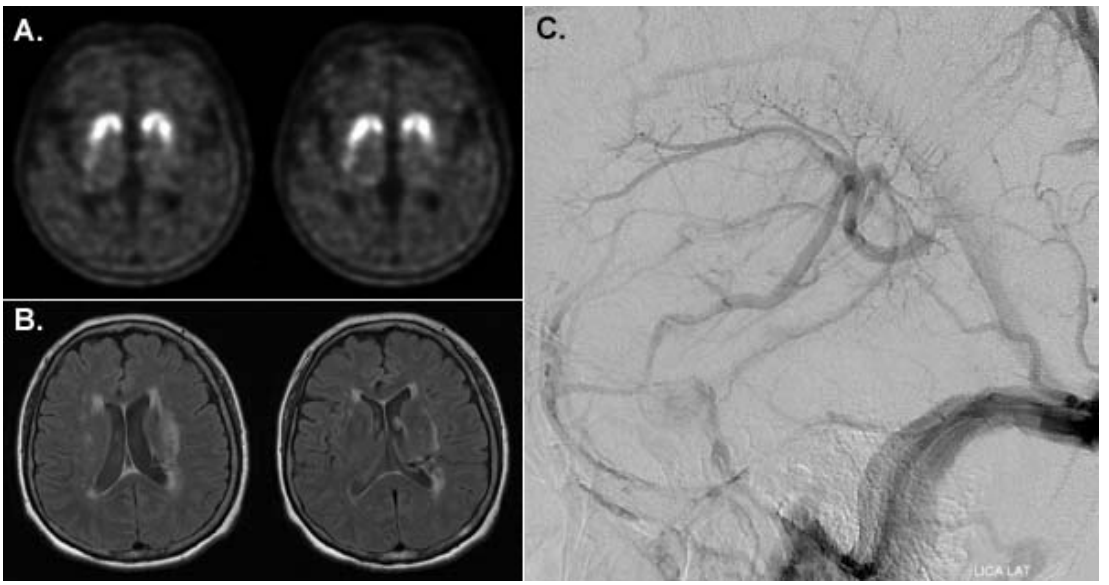

Figure 1. (A) F-18 FP-CIT PET shows symmetrical reduction of striatal tracer uptake in both posterior putamen. (B) Axial FLAIR MR image shows increased signal intensity in the white matter region drained by a developmental venous anomaly in the left basal ganglia. (C) Cerebral angiography during the venous phase confirms radiating medullary veins in the middle of the left basal ganglia region, suggesting a developmental venous anomaly.

the patient, and the study protocol was approved by the Institutional Review Board of CHA Bundang Medical Center.

DISCUSSION

DVAs can occur in basal ganglia areas with a frequency of 6%.[4,5] However, movement disorders are rarely associated with DVAs within basal ganglia regions, including hemichorea, hemichorea-hemiballism, cervical dystonia, tremor, and parkinsonism.[6-11] We report the first case of markedly asymmetrical parkinsonism associated with DVA in the basal ganglia, which presented in combination with symmetrical nigrostriatal dopaminergic degeneration. Our case is distinguishable from the previous case on a young woman presented with rapidly developing parkinsonian features associated with a DVA[11], in terms of a unilateral DVA manifested as asymmetrical neurodegenerative parkinsonism in the presence of symmetrical dopaminergic deficits. Most symptomatic DVAs in basal ganglia areas have been reported in association with concurrent or complicated conditions, such as hyperglycemia, microbleeding, and cavernous malformation.[6-9] In line with previous cases, a unilateral DVA in our patient might impose lateralized motor signs on neurodegenerative parkinsonism due to concurrent symmetrical dopaminergic deficiency, which in itself is not sufficient to account for the substantial asymmetry of motor signs in our patient.[12,13] In addition to the DVA, we observed increased signal intensity of white matter adjacent to the DVA in a FLAIR sequence of brain MRI. Abnormal signal changes in white matter regions drained by DVAs are considered to be caused by chronic local venous congestion, and are markers that predict the risk of symptomatic DVAs.[2,14] Accordingly, white matter lesion, along with the DVA, may contribute to the development of unilateral motor manifestation of our patient by asymmetrically disrupting the BG circuits. Relatively symmetrical motor involvement along with symmetrical striatal dopamine denervation is served as diagnostic clues to distinguish atypical parkinsonism from idiopathic Parkinson's disease.[12] Except for prominent asymmetry in motor signs, features observed in our patient indicate a diagnosis of atypical parkinsonism, including symmetrical striatal dopamine depletion, early postural imbalance, and poor response to levodopa treatment.[15,16] Accordingly, prominent asymmetric presentation of our patient may stem from a unilateral DVA involving the basal ganglia.

Our case demonstrates that a unilateral DVA in the basal ganglia together with symmetrical nigrostriatal degeneration may result in movement disorders by imposing marked asymmetry on

neurodegenerative parkinsonism. Prominent lateralization of motor signs in patients with signs of atypical parkinsonism can be an indication for further imaging evaluation to exclude superimposed structural lesions such as DVAs.

DISCLOSURE

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Conflict of interest: None

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