

Letter to the Editor

Stereotypy after cerebellar infarction



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1. Introduction

Stereotypy is operationally defined as a non-goal-directed continuous or continual repetition of the same movements [1]. Differential diagnosis from similar disorders can be made based on the presence of concomitant clinical features including internal urge or paroxysmal presentation.

Frontal or cerebellar dysfunction has been suggested to be mechanisms of stereotypy in various disorders such as frontotemporal lobar degeneration, Alzheimer's disease, and developmental disorders. However, stereotypy was only an accompanying feature to other predominant manifestations and usually lacking temporal correlation with the evolution of the disorders [2–4].

Herein we report a case of stereotypy developed after a cerebellar infarction.

2. Case report

A 71-year-old woman presented with a 1-week history of abnormal movements of her right hand. She had an acute infarction in the right cerebellum with cognitive impairment 1 month prior to presentation and had been treated for hypothyroidism for 2 years.

Neurologic examination revealed slowness in rapid alternating movement of the right limbs without ataxia. She exhibited repetitive supination and pronation of the right hand or rubbed her thigh when seated, and flexion and extension of the right elbow while walking, which were patterned and seemingly purposeless (Video). She denied any sensory urge to move her arm. She could suppress the abnormal movements, which reappeared when distracted.

Neuropsychological examination revealed disorientation to time and place, impaired verbal memory, buccofacial and ideomotor apraxia, and frontal dysfunction [Mini-Mental Status Examination (MMSE) = 7, Clinical Dementia Rating Scale = 3].

Laboratory studies were unremarkable except mild hypothyroidism with autoantibodies [anti-thyroglobulin antibody = 329.9, anti-thyroperoxidase antibody = 1517 (U/ml)], Cerebrospinal fluid examination was normal. Electroencephalography showed mild diffuse cerebral dysfunction.

Initial brain magnetic resonance imaging (MRI) at the onset of stroke showed acute infarction in the right cerebellum and left lingual gyrus, and steno-occlusion of intracranial arteries (Fig. 1A). A follow-up MRI

after onset of stereotypy showed no new lesions. Brain single photon emission computed tomography (SPECT) showed a perfusion defect in the right cerebellum (Fig. 1B). ¹⁸F-Fluorodeoxyglucose positron emission tomography revealed diffuse hypometabolism in the right hemisphere. Metabolism was preserved in the left sensorimotor cortex (Fig. 1C).

Stereotypy gradually improved with antiplatelet (cilostazol) and acetylcholinesterase inhibitor (donepezil). Her cognitive function was ameliorated (MMSE = 15) and stereotypy never returned during 3-year follow-up.

3. Discussion

Stereotypy has been reported occasionally in neurodegenerative dementias [5]. The involvement of the cortico-striato-thalamo-cortical pathway has been suggested in stereotypy of frontotemporal dementia [2]. However, because her cognition abruptly deteriorated after cerebral infarction, she was diagnosed with vascular dementia rather than neurodegenerative dementia. Her stereotypy was accompanied by cognitive impairment and partly reversible with medical treatment, which suggested that cognitive dysfunction might play an important role in her stereotypy. As her cognition globally deteriorated, an independent role of frontal lobe cannot be inferred from the cognitive evaluation. Hashimoto's encephalopathy was excluded by her 2-year history of hypothyroidism and resolution without steroid treatment.

Although an acute infarction was found in the left lingual gyrus, the lingual gyrus is related to vision and language and has never been associated with stereotypy. In case of the cerebellum, there were a few studies underscoring the involvement of the cerebellum in the stereotypies of developing children and Down syndrome [3,4]. In this patient, the follow-up MRI showed no new lesion, and the SPECT showed perfusion defect only in the right cerebellum; the cerebellar infarction was the most likely culprit responsible for her stereotypy. It is not easy to explain the 3-week delay between the infarction and the development of stereotypy. As her cerebral vasculature was very poor and her cognition and stereotypy improved with vasodilating agent (cilostazol), vascular compromise could have occurred in the interim period between the infarction and the stereotypy. The vascular compromise might not be severe enough to result in a new stroke but critical enough to trigger an abnormal network related to stereotypy.

The cerebellum normally receives efferent copies the motor cortices and sends out corollary discharges to be compared with the sensory information from movements performed [6]. In this patient, individual automatic movements of the hands seemed not to be dyspraxic, which were repeated without deterrence. The intactness of the left hemisphere, especially the left sensorimotor cortex, suggested by the preservation of metabolic activities, was essential for learning individually normal-looking movements. However, the efferent copies from intact motor cortices could have resulted in a mismatch with the corollary discharges from the damaged contralateral cerebellum. Thus, she might have less difficulty in acquisition of new automatic movements but greater problems in checking the repetition due to impaired feedback secondary to the destruction of cerebellar hemisphere.

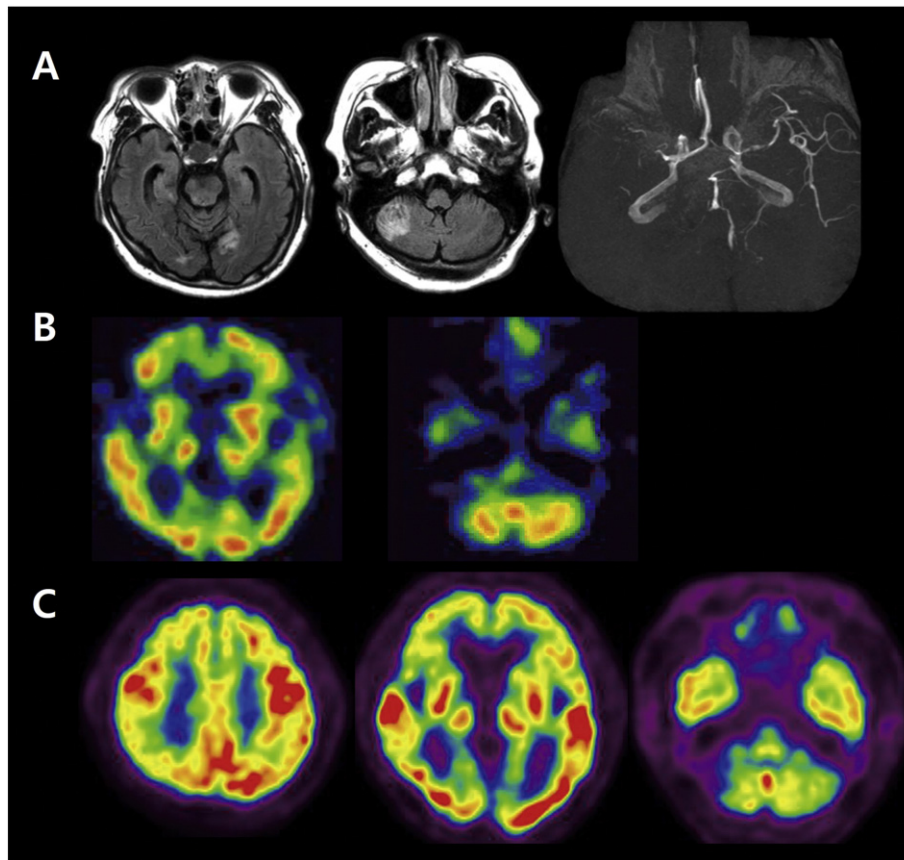


Fig. 1. T2 Fluid attenuated inversion recovery images at the onset show high signal intensities in the right cerebellum and left lingual gyrus and magnetic resonance angiography shows steno-occlusion of intracranial vessels (A). Brain single photon emission computed tomography shows a perfusion defect in the right cerebellum (B). ^{18}F -Fluorodeoxyglucose positron emission tomography reveals hypometabolism in the right cerebral cortices. Metabolism is also decreased in the right sensorimotor cortex, whereas it is preserved in the left sensorimotor cortex (C).

Ritualistic or stereotypic behavior could be a feature of cerebellar cognitive affective syndrome (CCAS) [7]. However, in this patient, if her stereotypy fell within the spectrum of CCAS, cortical function contralateral to the cerebellar infarction would likely be more severely impaired; instead, the reverse was true of the present patient.

In conclusion, we suggest functional mismatch between cerebellum and cortex combined with underlying diffuse cognitive dysfunction as a hypothetical mechanism of stereotypy related to cerebellar infarction.

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Competing interest

None.

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