Pearls And Oy-Sters: Hemiballism and Orbitofrontal-Like Syndrome in a Patient With Unilateral Tuberothalamic Stroke

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Pearls

Unilateral tuberothalamic stroke frequently causes behavioral changes and impairment of recent memory especially in the left hemisphere, while motor signs are mostly minimal or even absent.

The tuberothalamic artery supplies the paramedian thalamic area including the subthalamic nucleus, with ischemic stroke of this structure being the most common non-genetic cause of acquired chorea.

Major behavioral findings are negative symptoms such as abulia or apathy often accompanied by a reduced speech suggestive of transcortical aphasia.

Oy-sters

Positive behavioral symptoms such as logorrhea are a rare finding in tuberothalamic stroke likely caused by disruption of orbitofrontal pathways.

The diagnosis of tuberothalamic stroke is challenging due to a great deal of variability in the anatomy of the supplying arteries and a broad range of clinical symptoms.
Case report

A 72-year-old right-handed woman presented with a two-day history of acute onset logorrhea and right-sided movement disorder with hemiballism and choreic movements. In addition, the patient reported an urge to talk that she could not suppress (video 1). Further, she referred several arguments with close friends since the onset of her symptoms due to insulting language. The patient was well aware of a behavioral change that contrasted with her personality but did not seem to mind it. In addition, she reported involuntary movements of the right hand and leg that were initially mostly ballistic. Three days after symptom onset the movements were more distal and had a lower amplitude consistent with chorea (video 1). Her past medical history was unremarkable except for primary arterial hypertension.

Neurological examination at the time of admission revealed persistent involuntary proximal limb movements consistent with hemiballism that were confined to her right side. The movement disorder worsened during the serial sevens test, but the patient was still able to walk unaided. On neuropsychological testing she was cooperative, had an increased speech delivery rate, occasional semantic paraphasias and paragrammatical errors as well as impaired monitoring. She displayed severely impaired word list learning with deficits in encoding and recall and many intrusions hinting to temporal context confusion.

Brain CT showed a faint hypodense lesion in the left subthalamic nucleus. MRI with diffusion-weighted imaging (DWI) on day 3 after symptom onset confirmed a DWI hyperintense lesion in the left subthalamic nucleus extending to the anterior thalamus with hypointensity on apparent diffusion coefficient (ADC) maps consistent with acute ischemic stroke (figure 1, A-B). A source of embolism could not be detected. Echocardiography revealed mild aortic insufficiency and hypertensive heart disease. Electrocardiographic monitoring and extra-/transcranial ultrasound were unremarkable. Antiplatelet therapy with acetylsalicylic acid 100mg/d and lipid lowering treatment with atorvastatin 40mg/d was initiated permanently. For symptomatic treatment of the movement disorder, a medication with clonazepam 1.5 mg/d was started.
The movement disorder and logorrhea improved over the course of her hospitalization on our stroke unit but were still present at discharge to outpatient rehabilitation on day 7 after symptom onset.

Discussion

This patient with acute onset right-sided hemiballism, logorrhea, monitoring deficits and memory impairment had a left-sided stroke of the subthalamic nucleus and the anterior thalamus. The case is unusual because a small unilateral lesion in an uncommon location caused logorrhea and personality changes.¹

The subthalamic nucleus and anterior thalamus are supplied by the tuberothalamic artery, the most prominent of up to twelve perforators of the posterior communicating artery (Pcom).² It originates most often from the middle third of the Pcom and supplies the reticular nucleus, the ventral anterior as well as the rostral ventrolateral nucleus, the ventral pole of the medial dorsal nucleus, the anterior nuclei, the ventral internal medullary lamina, the ventral amygdalofugal pathway and the mamillothalamic tract (figure 1 D).³ In rare cases, the tuberothalamic artery may arise from the proximal segment of the posterior cerebral artery.³

Hemiballism occurs in around 1% of stroke patients hence making stroke the most common non-genetic cause of acquired chorea.⁴, ⁵ The location of the lesion is variable. Most often it is localized in the subthalamic nucleus, less frequently in the caudate nucleus, thalamus, putamen or globus pallidus.⁶ However, strokes located in the cortex or subcortical white matter may also cause hemiballism.⁴ It has been suggested that most stroke lesions causing hemiballism have a network overlap in the posterolateral putamen.⁷

Most patients with tuberothalamic stroke present with personality changes and memory disturbance.⁸ Impairment of recent memory is particularly prominent in tuberothalamic artery infarctions of the left side like in our patient.⁸ Lesions in the anterior nuclei or the mamillothalamic tract lead to a disruption of the Papez circuit mainly causing deficits in memory encoding, apathy and abulia.⁹⁻¹¹
In contrast, an orbitofrontal-like syndrome with disinhibition of speech and behavior has only rarely been reported in tuberothalamic stroke.\textsuperscript{12, 13} While apathy and abulia suggest a disruption of pathways to the medial frontal lobe, the disinhibition of speech and behavior observed in our patient as well as temporal context confusion indicate a disruption of orbitofrontal pathways.

Recognizing tuberothalamic stroke is challenging for several reasons. First, more than 60\% of patients have coexisting lesions in other vascular territories.\textsuperscript{14} Second, vascular variants give rise to a heterogeneity of symptoms of tuberothalamic strokes. Especially the adjacent paramedian artery from the P1 segment of the posterior cerebral artery provides collaterals to varying extend.\textsuperscript{8} Third, the neuro-anatomical organization of the thalamus and the resulting functional connectivity is complex, comprising extensive thalamocortical and – subcortical networks.\textsuperscript{15} As such, the thalamus can be considered as an “integrative hub”\textsuperscript{15} for most divergent functions such as sensory and motor function, visual and auditory function, language, memory and behavior.
## Appendix 1. Authors

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Figure 1: **Subthalamic Stroke:** Acute ischemic stroke of the left subthalamic nucleus extending to the anterior thalamus disrupting the Papez circuit. (A-B) Hyperintensity on diffusion weighted imaging in the vascular territory of the tuberothalamic artery with corresponding decreased apparent diffusion coefficient (C). (D) Scheme of vascular supply of the thalamus in lateral view. Abbreviation: 1, internal carotid artery; 2 posterior cerebral artery P1 segment; 3, posterior communicating artery; 4, posterior cerebral artery P2 segment; 5, tuberothalamic artery; 6, paramedian artery; 7, inferolateral artery; 8, posterior choroidal artery; DM, dorsomedial nucleus; VA, ventral anterior nucleus; VL, ventrolateral nucleus; VP, ventral posterior complex; P, Pulvinar.
Video 1: 72-year-old patient with acute onset right-sided movement disorder with logorrhea three days after symptom onset.

References:

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