An Unusual Case of Hemiballism-Hemichorea Associated with Nonketotic Hyperglycemia in Association with a Centrum Semiovale Stroke

Giulia Lancellotti, Catherine Sagot, Anne Forest, Sandrine Greffard, Anne Bertrand, Marc Verny

AN UNUSUAL CASE OF HEMIBALLISM- HEMICHOREA ASSOCIATED WITH NONKETOTIC HYPERGLYCEMIA IN ASSOCIATION WITH A CENTRUM SEMIOVALE STROKE

To the Editor: An 86-year-old man with a medical history of diabetes mellitus, hypertension, dyslipidemia, severe tobacco use (10 pack-years), and brain trauma was admit-

ted to the hospital for a left-sided hemichoreoathetosis that had started suddenly 48 hours before. Family history was unremarkable for movement disorders.

On neurological examination, he was disoriented to time and space and presented with left-sided abnormal involuntary movements suggestive of hemichoreoathetosis associated with left facial spasms.

Brain computed tomography (CT) revealed spontane- oushyperdensity within the right striatum. Blood sample examination showed high fasting glucose (306 mg/dL, nor- mal <126 mg/dL), high glycosylated hemoglobin (HbA1c; 11.9%, normal <6.5%), and high osmolarity (303.3 mO- smol/kg, normal 285–295 mOsmol/kg); urinary ketones were absent; and cerebrospinal fluid analysis revealed high glucose and lactate levels. On brain magnetic resonance imaging (MRI), the right striatum was hyperintense on T1-weighted images without contrast administration, sug- gestive of diabetic striatopathy. Focal hyperintensity on diffusion-weighted imaging within the right centrum semiovale suggested the presence of an associated recent ischemic stroke (Figure 1).

Diabetes mellitus decompensation, with fasting glu- cose levels reaching 540 mg/dL during the hospital stay, was treated with insulin, and platelet antiaggregationther- apy for stroke treatment and recurrence prevention was started. As soon as blood glucose decreased toward normal values, hemichorea gradually subsided, making symptom- atic treatment unnecessary; dramatic cognitive and motor improvement were obtained at discharge.

Clinical and MRI follow-up were performed. Hypersig- nal of right lenticular and caudate nuclei and recurrence of left hemichorea were evident until 9 months later in a context of not-yet-satisfactory glycemic control (HbA1c 9.1%). Imaging and clinical findings had disappeared at 24- month follow-up, when better control of diabetes mellitus was evident (HbA1c 8.3%), reinforcing the diagnosis of hyperglycemic hemichorea. He had three recurrent strokes during 24 months of follow-up in the deep territory of the left middle cerebral artery, the right cerebellar hemisphere, and the left medulla oblongata and right corona radiata.

This individual was characterized by the overlap of diabetic decompensation with hemichoreicsymptomatol- ogy and ischemic stroke manifesting with confusion and postural instability. Hemichorea was recurrent during the 9-month period of persistence of basal ganglia abnormali- ties in the context of poor glycemic control. Diabetic striatopathy is a well-recognized but rare cause of hemibal- lism-hemichorea, associated with contralateral basal ganglia hyperdensity on CT and hyperintensity on T1- weighted MRI.[1–4] The cause of clinical and imaging abnormalities is unclear. Several mechanisms attributed to hyperglycemia have been proposed: transient ischemic injury with microhemorrhage resulting in hyperviscosity caused by hyperosmotic state, calcium deposition, gemisto- cytosis, altered GABAergic and dopaminergic

neurotrans- mission,[5–8] paramagnetic mineral deposition including zinc-containing metallothionein expressed in swollen astro- cytes,[9] and autoimmunity-mediated inflammation.[10]

This was the first case, to the authors' knowledge, of contemporary occurrence of hyperglycemia-related hemi- chorea and stroke. Two possible diagnostic hypothesis were taken into account: the first considered stroke to have played a central role in diabetes mellitus decompensation, which, in a context of cerebral frailty due to stroke, led to the development of hyperglycemia-related hemiballism- hemichorea; the second considered hyperglycemia as the trigger of stroke through an ischemic injury due to hyper- viscosity.[5] The available anamnestic data did not help identify which of the two disorders first appeared. The proximity of the stroke lesion to the altered basal ganglia, both in the right middle cerebral artery area, is interesting; although this strengthens the hypothesis of the ischemic mechanism for imaging abnormalities associated to hyper- glycemia, it seems in accord with the second hypothesis. It could also be related to the first hypothesis, if the abnor- mal basal ganglia is considered to enter the ischemic penumbra of the documented stroke. Moreover, a partici- pating direct role of the documented ischemic stroke to the abnormal movements cannot be excluded given the exis- tence also of vascular hemichorea-hemiballism typically associated with lesions of the basal ganglia and, as in this case, of the adjacent white matter.[1]

Difficulties in optimal glycemic control probably contributed to the persistence for several months of basal ganglia abnormalities accompanied by recurrence of hemi- chorea, both finally disappearing 24 months later when diabetes mellitus compensation was also evident.

Hyperglycemia-related hemiballism-hemichorea and the occurrence of four ischemic strokes during a 24-month per- iod are rare in clinical practice. This man's case raises doubts also about the possible predictive role of hyperglycemia- related hemiballism-hemichorea for recurrence of stroke.

Giulia Lancellotti, MD Division of Gerontology and Geriatrics, Department of Biomedical, Metabolic and Neural Sciences, University of Modena and Reggio Emilia, Modena, Italy

Catherine Sagot, MD Department of Geriatrics, Assistance Publique-Hopitaux de Paris, University Hospital Department "Fight Ageing and Stress," Piti e-Salpetri ere Hospital, Paris, France

Anne Forest, MD Department of Geriatrics, Hospital Center Marmande, Marmande, France

Sandrine Greffard, MD Department of Geriatrics, Assistance Publique-Hopitaux de Paris, University Hospital Department "Fight Ageing and Stress," Piti e-Salpetri ere Hospital, Paris, France

Anne Bertrand, MD, PhD Department of Functional and Diagnostic Neuroradiology, Assistance Publique-Hopitaux de Paris, Piti e-Salpetri ere Hospital, Paris, France

Marc Verny, MD, PhD Department of Geriatrics, Assistance Publique-Hopitaux de Paris, UniversityHospitalDepartment "FightAgeing and Stress," Piti e-SalpetriereHospital, Paris, France Universit e Pierre et Marie Curie, Sorbonne Universit es, Paris, France

REFERENCES

1. Cardoso F, Seppi K, Mair KJ et al. Seminar on choreas. Lancet Neurol2006;5:589–602.

2. Piccolo I, Defanti CA, Soliveri P et al. Cause and course in a series of patients with sporadic chorea. J Neurol2003;250:429–435.

3. Chu K, Kang DW, Kim DE et al. Diffusion-weighted and gradient echo magnetic resonance findings of hemichorea-hemiballismus associated with diabetic hyperglycemia: A hyperviscosity syndrome? Arch Neurol2002;59:448–452.

4. Koh YHM, Lim WS, Seng TT et al. An unusual case of non-ketotic hyperglycemia presenting as hemichorea. J Am Geriatr Soc 2007;55: 1691–1693.

5. Chang KH, Tsou JC, Chen ST et al. Temporal features of magnetic resonance imaging and spectroscopy in nonketotic hyperglycemic chorea- ballism patients. Eur J Neurol2010;17:589–593.

6. Battisti C, Forte F, Rubenni E et al. Two cases of hemichorea-hemiballism with nonketotic hyperglycemia: A new point of view. NeurolSci2009;30:179–183.

7. Nath J, Jambhekar K, Rao C et al. Radiological and pathological changes in hemiballism-hemichorea with striatal hyperintensity. J MagnReson Imaging 2006;23:564–568.

8. Shan DE, Ho DM, Chang C et al. Hemichorea-hemiballism: An explana- tion for MR signal changes. Am J Neuroradiol1998;19:863–870.

9. Cherian A, Thomas B, Baheti NN et al. Concepts and controversies in nonketotic hyperglycemia induced hemichorea: Further evidence from susceptibility-weighted MR imaging. J MagnReson Imaging 2009;29: 699–703.

10. Wang JH, Wu T, Deng BQ et al. Hemichorea-hemiballismus associated with nonketotic hyperglycemia: A possible role of inflammation. J NeurolSci2009;284:198–202.