Diabetic striatopathy

Takanobu Iri,¹ Hiroyuki Yano,² Mitsuyo Kinjo ²

¹Medicine, Okinawa Hokubu Hospital, Nago, Japan

²Medicine, Okinawa Chubu Hospital, Uruma, Japan

Correspondence to
Dr Mitsuyo Kinjo;
kinjomitsuyo@gmail.com

Accepted 27 July 2021

DESCRIPTION

A 70-year-old man presented with involuntary movements of the left lower leg. The patient had recent admission 4 weeks ago with increased drowsiness and raised capillary blood glucose reading of 935 mg/dL with elevated serum osmolality level at 321 mosmol/kg and no ketoacidosis (pH 7.404), resulting in the diagnosis of hyperosmolar hyperglycaemic syndrome (HHS). Intravenous fluid and insulin improved his consciousness. With lower serum C-peptide level and high haemoglobin A1c level (17.6%), the patient was diagnosed with latent autoimmune diabetes in adults with a positive anti-glutamic acid decarboxylase antibody level at high tit ≥ 2000 U/mL. Regular insulin was initiated.

Three days beforehand, involuntary movements of the left lower leg occurred at rest. Other history was unremarkable. He did not take any medications except for insulin. He used tobacco daily for 20 years but quit 30 years before. He did not consume alcohol or use illicit drugs.

On examination, vital signs were stable. His body mass index was 20.8. He was alert and oriented. Neurological examination revealed involuntary movement of the left upper arm and lower limb ([video 1](#)). Strength was 5/5 throughout. Sensation to light touch and pinprick was intact. Reflexes were symmetrical. Cranial nerves were intact. Cerebellar signs were absent. Romberg sign was not performed. Other system examination results were normal.

Laboratory studies revealed normal complete blood count, liver and renal function tests, and electrolytes. There was no acidemia (pH 7.36, HCO_3^-

Video 1 Involuntary movement of the left upper arm and lower limb was notable.

Figure 1 A brain MRI scan showed right putamen T1 hyperintensity.

Figure 2 The abnormal signals of the right putamen on MRI images had almost disappeared.

© BMJ Publishing Group Limited 2021. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Iri T, Yano H, Kinjo M. *BMJ Case Rep* 2021;**14**:e244248. doi:10.1136/bcr-2021-244248

Images in...

27.3 mmol/L). A chest radiograph, ECG and CT of the head were unremarkable. A brain MRI scan showed right putamen T1 hyperintensity (figure 1) with normal T2 intensity and diffusion weighting. Ballism secondary to diabetic striatopathy (DS) was diagnosed. Insulin and sulpiride were commenced and glucose normalised, along with improvement of involuntary movements. Six months later, the abnormal signals on MRI images had almost disappeared (figure 2).

Chorea consists of involuntary, rhythmic, purposeless, jerky movements involving distal limbs. Proximal muscle involvement with larger amplitude is commonly seen in ballism.¹ Basal ganglia and subthalamus dysfunction causes chorea or ballism. Aetiologies of chorea or ballism include cerebrovascular, metabolic, structural, inflammatory, infectious, autoimmune and iatrogenic diseases.¹ DS is characterised by the sudden onset of hemichorea or hemiballism associated with hyperglycaemic and striatal abnormality either by hyperdensity on CT or hyperintensity on T1-weighted MRI.² DS usually develops in the elderly population with a female predominance and long-standing, poorly controlled type 2 diabetes mellitus (DM) with an average haemoglobin A1c level of 13.1%. DS infrequently occurs in type 1 DM³ and even after correction of hyperglycaemic.⁴ However, it can be the first manifestation of DM in 17% of patients.¹ The precise pathogenic mechanism of DS is unclear in non-ketotic hyperglycaemic status.⁵ However, it is hypothesised that the depletion of gamma-aminobutyric acid (GABA) due to the upregulation of an alternative anaerobic pathway in the Krebs cycle leads to the disinhibition of the subthalamus and basal ganglia associated with hyperkinetic movement.¹

CT or MRI can detect the striatal abnormality of DS: T1-weighted MRI demonstrates hyperintensity in the putamen,

caudate nucleus and globus pallidus,^{6–8} which presumably result from petechial haemorrhage, mineral deposition, myelin destruction or infarction with astrocytosis.¹

Management of DS includes correction of hyperglycaemic and symptom control. Chorea may improve by glucose control in 25% of patients, while the majority may need treatment including GABA-receptor agonists, selective serotonin reuptake inhibitors and dopamine-depleting agents.⁹

New onset of ballism or chorea in a diabetic patient following HHS should alert the clinician about DS, which can be confirmed by MRI.

Acknowledgements We thank Rita McGill, MD, MS (Department of Nephrology, University of Chicago), for English correction of the manuscript.

Contributors TI wrote manuscript and collected data. HY contributed to the writing of the manuscript and edited data. MK contributed to the overall writing of the manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iD

Mitsuyo Kinjo <http://orcid.org/0000-0002-8342-230X>

REFERENCES

- 1 Chua C-B, Sun C-K, Hsu C-W, *et al.* "Diabetic striatopathy": clinical presentations, controversy, pathogenesis, treatments, and outcomes. *Sci Rep* 2020;10:1594.
- 2 Abe Y, Yamamoto T, Soeda T, *et al.* Diabetic striatal disease: clinical presentation, neuroimaging, and pathology. *Intern Med* 2009;48:1135–41.
- 3 Alves C, Sampaio S, Barbosa V, *et al.* Acute chorea and type 1 diabetes mellitus: clinical and neuroimaging findings. *Pediatr Diabetes* 2012;13:e30–4.
- 4 Cho H-S, Hong C-T, Chan L. Hemichorea after hyperglycemia correction: a case report and a short review of hyperglycemia-related hemichorea at the euglycemic state. *Medicine* 2018;97:e0076.
- 5 Lin JJ, Chang MK. Hemiballism-hemichorea and non-ketotic hyperglycaemia. *J Neurol Neurosurg Psychiatry* 1994;57:748–50.
- 6 Shan DE, Ho DM, Chang C, *et al.* Hemichorea-hemiballism: an explanation for MR signal changes. *AJNR Am J Neuroradiol* 1998;19:863–70.
- 7 Oh S-H, Lee K-Y, Im J-H, *et al.* Chorea associated with non-ketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a meta-analysis of 53 cases including four present cases. *J Neurol Sci* 2002;200:57–62.
- 8 Cherian A, Thomas B, Baheti NN, *et al.* Concepts and controversies in nonketotic hyperglycemia-induced hemichorea: further evidence from susceptibility-weighted MR imaging. *J Magn Reson Imaging* 2009;29:699–703.
- 9 Das L, Pal R, Dutta P, *et al.* "Diabetic striatopathy" and ketoacidosis: Report of two cases and review of literature. *Diabetes Res Clin Pract* 2017;128:1–5.

Learning points

- Diabetic striatopathy is characterised by the sudden onset of hemichorea or hemiballism associated with hyperglycaemic and striatal abnormality either by hyperdensity on CT or hyperintensity on T1-weighted MRI.
- Chorea may improve by glucose control in 25% of patients, while the majority may need treatment including GABA-receptor agonists, selective serotonin reuptake inhibitors and dopamine-depleting agents.

Copyright 2021 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

Customer Service

If you have any further queries about your subscription, please contact our customer services team on +44 (0) 207111 1105 or via email at support@bmj.com.

Visit casereports.bmj.com for more articles like this and to become a Fellow