Radiology Case Reports 16 (2021) 895–898

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Bilateral striatum with high-signal intensity on T1-weighted MRI: A case of hemichorea induced by nonketotic hyperglycemia ☆

Yuanyi Pan[a], *Fattyang Chew, PhD*[b], *Rongping Wang, PhD*[a], *Xuntao Yin, PhD*[a], *Yaying Li, PhD*[a,*]

[a] *Department of Radiology, Guizhou Provincial People's Hospital, Zhongshan East Rd.83, Nanming Dist., Guiyang, 550002, Guizhou, China*

[b] *Department of Medical Imaging, China Medical University Hospital, Taichung City, Taiwan (R.O.C.)*

ARTICLE INFO

Article history:
Received 23 December 2020
Revised 9 January 2021
Accepted 11 January 2021

Keywords:
Diabetes mellitus
Nonketotic hyperglycemia
T1-weighted MRI

ABSTRACT

Hemichorea induced by nonketotic hyperglycemia is a rare complication of diabetes mellitus. Here we present a case of 80-year-old female patient. She had a history of involuntary movement of the left body. Imaging examination showed abnormal signs of bilateral basal ganglia. After relevant treatment was conducted, the patient's hemichorea symptoms were significantly improved. Imaging features of nonketotic hyperglycemia Hemichorea plays a very important role in diagnosing and treatment of this disease.

Introduction: Hemichorea refers to the involuntary movements of one or more limbs of the same side. These movements are continuous and irregular, variable amplitude, and usually involve the arms and legs together. Stroke is the most common reason, hyperglycemia is relatively rare. Characteristic imaging findings can be very suggestive of diagnosis, and also closely related to the development of disease. Here we present a case of hemichorea induced by nonketotic hyperglycemia which was confirmed by bilateral abnormal signals of basal ganglia in MRI.

Case report

An 80-year-old female patient presented to our emergency department with involuntary movement of her left limbs for more than 10 days. She has a history of type 2 diabetes mellitus without any treatment. Neurological examination showed that the patient had a slow response, poor muscle strength, and positive tendon reflexes of her left limbs. Involuntary movements were observed in the left upper and lower limbs and the movements were continuous and irregular with variable amplitude. Laboratory examinations on admission

☆ Declaration of competing interest: No conflict of interest.

* Corresponding author.
E-mail addresses: 872524179@qq.com (Y. Pan), c_fyang@hotmail.com (F. Chew), liyaying1120@163.com (Y. Li).

https://doi.org/10.1016/j.radcr.2021.01.024

revealed glucose: 8.59 mmol/L (3.9 mmol/L-6.1 mmol/L) and HBA1c 7.43%(4%-6%), and other laboratory data were unremarkable. Brain computed tomography showed slightly high density in the bilateral striatal region, and no obvious signs of stroke were seen. Subsequent brain MRI demonstrated high-signal intensity on T1-weighted of bilateral basal ganglia. Hence, the diagnosis of nonketotic hyperglycemia hemichorea was made. The involuntary movement of the patient has ameliorated after treatment with metformin and dietary modification. She was discharged from the hospital. However, the involuntary movement of her left limbs aggravated on 1-month follow-up due to her poor control of her blood glucose level.

Discussion

Hemichorea induced by nonketotic hyperglycemia is a rare complication of poor blood glucose control. Additional complications of it include seizures, myasthenia, dystonia and pyramidal tract signs, which may also occur in combination [1,2]. It is possible to develop when the patient is being nervous. It may be alleviation and disappear when the patient is being in relaxation and sleeping.

The incidence rate of it is about 1/100,000 [3]. It occurs mostly in Asian women over 70-year-old, which may be related to genetic factors and high dopaminergic response due to racial and gender differences. After menopause, decrease of estrogen leads to hypersensitivity phenomenon of dopamine receptor in the substantia nigra striatum system, so that the dopamine function is relatively enhanced. Nonketotic hyperglycemia related hemichorea usually occurs a few weeks later after high blood glucose levels [4].This response indicates a delayed response to severe hyperglycemia [5]. In our case, there was no symptom when the blood glucose was 30 mmol/L. Laboratory test results include high blood glucose and glycosylated hemoglobin. Although classical description is ketosis hypertonic hyperglycemia, but patients may appear normal serum osmotic quantity and serum and urine ketone, the patients were moderately elevated blood ketone.

The pathophysiology of the disease is not fully elucidated yet. Hypothesis of GABA metabolism disorders is now accepted most widely. Hemichorea can be resulted from decreasing of GABA function or increasing of dopamine function. Because of microvascular lesion, high blood viscosity and cerebral ischemic anoxia damage of blood-brain barrier, basal ganglia region-related neurons use GABA for alternative energy sources for anaerobic metabolism. While patients of ketosis take advantage of ketone to synthesize acetoacetate and resynthesis of GABA accordingly, so they may not develop to hemichorea. However, the lack of materials in nonketosis patients results in a significant decrease of GABA, and shows dance-like extrapyramidal symptoms. 18F-FDG PET/CT shows obvious low metabolism in the lesion of basal ganglia [6]. Morbidity of nonketotic hyperglycemia-related hemichorea is higher in patients of women after 70 may furtherly testify this hypothesis. During in postmenopausal period, dopamine function get to increase due to low levels of estrogen, which can sensitize the dopamine receptors in the substantia nigra striatum system.

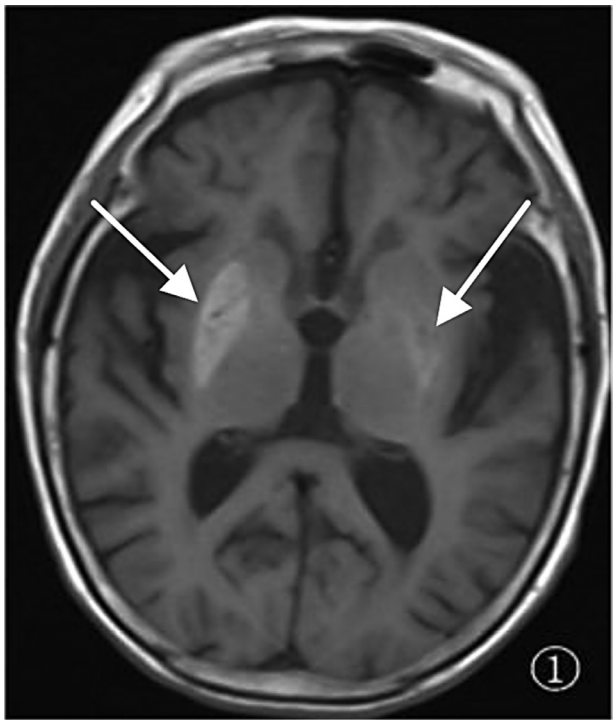

Fig. 1 – T1WI of Transverse section: High-signal intensity on T1-weighted of bilateral basal ganglia, more obvious on the right side.

The characteristic imaging manifestations are these: Most of previous cases showed high-signal intensity on T1-weighted in the basal ganglia region, especially striatum, and the putamen was always susceptible. The lesions had no surrounding edema and mass effect. There were also a few reports of bilateral negative finding and ipsilateral basal ganglia involvement. In our case, T1-weighted high signals were found in both sides of basal ganglia, but relatively heavy in the contralateral area of limb, indicating that the range of T1-weighted high signals in the striatum was closely related to the severity of the disease. Studies have showed semi-quantitative volume analysis of MRI T1 images quantifies lesions and evaluate the improvement of clinical symptoms [7]. T2-weighted striatum signals are variable and can be equisignal or low signal. Recent studies have reported that low signal of T2 sequence has suggestive significance for the diagnosis [8]. Negative changes were found on DWI and ADC images. CT images show slightly high-density changes in bilateral striatum, which can be adjusted to narrow window and high window level. Some cases have no positive found. Interestingly, the affected areas of CT and MRI images are often mismatched, it may indicate there are two different pathophysiological mechanisms [9].Various mechanisms have been proposed to explain the imaging manifestations, such as micro-hemorrhage caused by damage to the blood-brain barrier, calcification caused by microinfarction, astrocyte hyperplasia, ischemia-reperfusion injury, and cell edema [9,10].

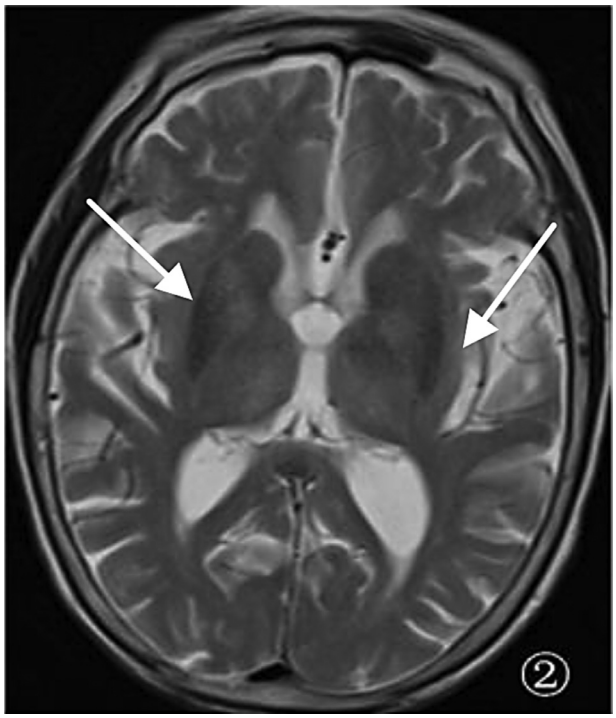

Fig. 2 – T2WI of transverse section: Symmetry low-signal intensity on T1-weighted of bilateral basal ganglia.

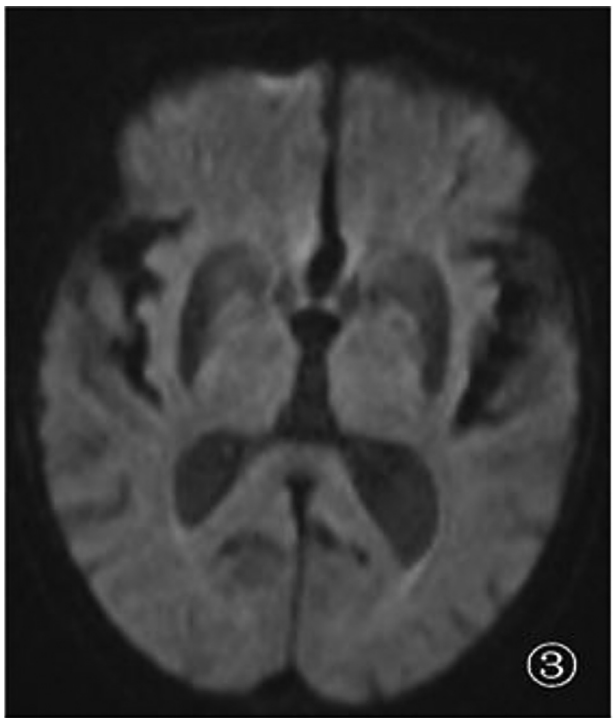

Fig. 3 – DWI: Negative changes were found on DWI and ADC images.

Fig. 4 – CT: Slightly high-density changes in bilateral striatum.

The differential diagnosis of this disease mainly includes clinical symptoms and imaging examination, and also medical history, laboratory examination, and other clinical data. T1WI high-signal intensity of basal ganglia is mostly seen in chronic liver diseases, neurofibromatosis, manganese deposition, calcification, etc. The signal changes caused by metabolic diseases are usually bilateral and symmetrical, while ketosis hyperglycemia is often partial side. Our case indicates Hemichorea of asymmetrical bilateral signal, which makes it more difficult to make a differential diagnosis. In terms of the symptoms of unilateral disease, cerebrovascular disease is the primary inducing factor, nonketotic hyperglycemia is the second most common cause, and other causes include brain trauma, amyotrophic lateral sclerosis, tumors, tuberculoma, demyelinating plaques, human immunodeficiency virus, metabolic disorders, and drugs [11]. Therefore, differential diagnosis can be effectively made by the combination of hyperglycemia symptoms and imaging images.

Control of blood glucose is the main treatment clinically, hemichorea symptoms usually can be improved significantly or disappeared as blood glucose is under controlled, dopamine receptor blockers can be used in the treatment of sustained and severe symptoms, sometimes clonazepam, diazepam can be jointly used. Dopamine receptor antagonist may cause tardive dyskinesia, so treatment plan should be taken considerably and individually. The high signal of T1WI generally lasts for months to several years, signal changes in the treatment process, and can be reduced or even disappear with the improvement of the disease. Most patients have a benign prognosis, and a few symptoms persist. In our case, after blood glu-

cose control, the condition of hemichorea was improved, but after discharge, the patient's blood glucose control was poor again and the situation worsened.

Although hemichorea induced by nonketotic hyperglycemia usually takes a benign prognosis, this persistent abnormal movements of limbs cause inconvenience and pain for patients and their families. Therefore, clinicians should also pay more attention to the psychological and social effects in the treatment of diseases.

Hemichorea induced by nonketotic hyperglycemia typically shows tri-signs, which are unilateral involuntary dance-like movement, high-signal intensity of contralateral striatal region on T1-weighted images, and symptoms affected by the blood glucose levels (Figs. 1–4).

Authors contribution

Yuanyi Pan: Drafting the manuscript and revising the work.

Fattyang Chew, Rongping Wang, Xuntao Yin, Yaying Li: Critical revision of the manuscript.

Patient consent

Patient consent has been obtained.

Acknowledgements

This study was supported by Science and Technology Project of Guizhou Provincial Health Commission (gzwjkj2016-1-067) and Doctor Foundation of Guizhou Provincial People's Hospital (GZSYBS[2019]02).

REFERENCES

[1] Wang X. Nonketotic hyperglycemia-related epileptic seizures. Chin Neurosurg J 2017;3(1).
[2] Kim R, Cho HJ, Lee HW, Jun JS. Combined hemichorea and seizures in a patient with nonketotic hyperglycemia. J Mov Disord 2020;13(1):72–3.
[3] Jaafar J, Rahman RA, Draman N, Yunus NA. Hemiballismus in uncontrolled diabetes mellitus. Korean J Fam Med 2018;39(3):200–3.
[4] H-Sa Cho, C-Ta Hong, L Chan. Hemichorea after hyperglycemia correction: a case report and a short review of hyperglycemia-related hemichorea at the euglycemic state. Medicine (United States) 2018;97(10) e0076.
[5] Hsiao PJ, Kuo CC, Kuo TY, Kao YH, Chan JS, Lin YY, et al. Investigation of the relationship between non-ketotic hyperglycemia and hemichorea-hemiballism: a case report. Medicine (Baltimore) 2019;98(28):e16255.
[6] Kumar Vadi S, Mehta S, Kumar R, Tandyala N, Singh H, Mittal BR. Severe contralateral striatal hypometabolism in a case of diabetic nonketotic hyperglycemic hemichorea on 18F-FDG PET/CT brain. Clin Nucl Med 2020;45(2):e117–19.
[7] Lin YT, Chen SC, Yip PK, Wang V. Magnetic resonance imaging volumetric analysis for diabetic striatopathy with two episodes of hemichorea-hemiballism syndrome: a case report. Medicine (Baltimore) 2019;98(38):e17249.
[8] Yu F, Steven A, Birnbaum L, Altmeyer W. T2*-based MR imaging of hyperglycemia-induced hemichorea-hemiballism. J Neuroradiol 2017;44(1):24–30.
[9] Shan D, Ho D, Chang C, Pan H, Teng M. Hemichorea-hemiballism: an explanation for MR signal changes. Am J Neuroradiol 1998(No.5):863–70.
[10] Johari B, Hanafiah M, Shahizon AM, Koshy M. Unilateral striatal CT and MRI changes secondary to non-ketotic hyperglycaemia. BMJ Case Rep 2014;2014.
[11] Priola AM, Gned D, Veltri A, Priola SM. Case 204: Nonketotic hyperglycemia-induced hemiballism-hemichorea. Radiology 2014;271(1):304–8. doi:10.1148/radiol.14120840.