Bilateral dystonia in type 1 diabetes: a case report

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Abstract

**Introduction** Diabetic hemichorea-hemiballismus is a rare complication of type 2 diabetes. Here, we first report a case with type 1 diabetes, manifesting hemichorea and bilateral dystonia as hyperglycemia-induced involuntary movement.

**Case presentation** A 62-year-old Japanese women with body weight loss of 30 kilograms during past 1 year developed symptoms of thirst, **polydypsia** and polyuria. She also presented with hemichorea and bilateral dystonia for 5 days. She presented extremely high plasma glucose (774 mg/dl), hemoglobin A1c (21.2%) and glycated albumin (100%) with ketosis. Based on the presence of glutamic acid decarboxylase antibodies (18,000 U/ml; normal<1.3 U/ml), lower daily urinary excretion of C-peptide (7.8 µg), ketosis and HLA typing DR-4, we diagnosed as type 1 diabetes mellitus. We treated with continuous intravenous regular insulin infusion and medication with haloperidol, and finally dystonia completely disappeared within 3 days.

**Conclusion** Dystonia is one of the manifestations of hyperglycemia-induced involuntary movement besides hemichorea-hemiballism.

Introduction

Chorea is defined as irregular, unpredictable, brief and jerky involuntary movements, while ballismus is large-amplitude flailing movements [1]. Hemichorea-hemiballismus is a rare complication of non-ketotic hyperglycemia and only 53 case reports of this particular condition were published between 1985 and 2001 [2]. Most of the cases were more than 60 years old and represented type 2 diabetes and non-ketotic hyperglycemia. The differential diagnosis of diabetic hemichorea-hemiballismus is challenging because this type of hyperkinetic movement disorders is caused by focal lesions, such as ischemic or hemorrhagic stroke, infection, epilepsy, and neoplasm, as well as systemic processes, including systemic lupus erythematosus, Wilson’s disease and thyrotoxicosis [1]. Here, we present a case with type 1 diabetes dystonia is also one of the manifestations of hyperglycemia-induced involuntary movement.

Case presentation

A 62-year-old Japanese women with body weight loss of 30 kilograms during past 1 year developed symptoms of thirst, **polydypsia** and polyuria admitted to our hospital. She also presented with hemichorea and bilateral dystonia for 5 days. She recognized her forefeel for several seconds in prior to the initiation of involuntary movement. At first, she had chorea movement of her right arm at ~3 Hertz, and then involuntary and slowly elevated her right arm accompanied by continuing chorea movement of right hand; she simultaneously stretched her right leg. About 10 seconds later, she slowly flexed her left knee and kept the bilateral and asymmetrical spastic posture. The sequences of slow and continuous muscular contractive movement were defined as bilateral “dystonic movement”. Whole series of her movement terminated in 30 seconds and she finally was relieved from her dystonia and could voluntarily move (See additional file 1: Movie1). Since the exactly same pattern of hemichorea and bilateral dystonic movement intermittently occurred every 10 minutes, she could not stand and had difficulties in taking meals for 2 days. These movements were observed in both waking and sleep states. She presented extremely high plasma glucose (774 mg/dl), hemoglobin A1c (21.2%) and glycated albumin (100%) with ketosis but without
Acidosis. Anti-nuclear antibodies were negative, serum ceruloplasmin and thyroxine levels were within normal range. Magnet resonance imaging demonstrated no brain tumor, hemorrhage and infarction and she had a normal electroencephalogram excluding possibility of epilepsy. In this case, there were no typical MR images seen in diabetic hemichorea-hemiballisms such as high signal basal ganglia lesions, mainly putamen, on T1-weighted images [3]. Based on the presence of glutamic acid decarboxylase antibodies (18,000 U/ml (normal<1.3 U/ml), lower daily urinary excretion of C-peptide (7.8 μg), ketosis and HLA typing DR-4, we diagnosed as type 1 diabetes mellitus. We treated with continuous intravenous regular insulin infusion and medication therapy with haloperidol, and finally dystonia completely disappeared within 3 days. After the discontinuation of haloperidol, the recurrence of dystonia was not observed.

Discussion

Many hypotheses for the development of diabetic hemichorea-hemiballismus were reported, such as local GABA starvation, disinhibition of dopaminergic neurons, local microhemorrhage, microinfarction, demyelination and brain edema [4]. Recent imaging analysis revealed reduced cerebral glucose metabolism on positron emission tomography (PET) scans with concomitant hyperperfusion in affected basal ganglia seen on single photon emission computed tomography (SPECT)[5]. In some cases, the basal ganglia in diabetic hemichorea-hemiballismus revealed hyperdense without mass effect on CT scans and hyperintense on T1-weighted MRI scans but these imaging features completely reverse after therapy [2]. These evidences supported the idea that basal ganglia are generally weak in hyperglycemic stress, and chronic hyperglycemic stress might induce reversible neurotransmitting functional disorders and consequent involuntary movement. Since dystonia is caused by the lesions of basal ganglia, the dystonia is a spectrum of the hyperglycemia-induced involuntary movements in addition to hemichorea-hemiballism.

Diabetic hemichorea-hemiballismus was observed mostly in type 2 diabetes and the cases with type 1 diabetes were extremely rare. In the 53 cases reported in the literature, only one case of type 1 diabetes with acute onset of non-ketotic hyperglycemia was reported and the rest of them were type 2 diabetes in elder patients [2]. The series of the case suggested that long-term exposure to hyperglycemia without ketosis in elderly is related to the development of hemichorea-hemiballismus in diabetes. We speculated that the case was exposed to long-term hyperglycemic stress because she manifested slowly progressive form of type 1 diabetes without acidosis states.

Conclusion

Dystonia is one of the manifestations of hyperglycemia-induced involuntary movement besides hemichorea-hemiballism.

Abbreviations

If abbreviations are used in the text they should either be defined in the text where first used, or a list of abbreviations can be provided.

Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**Competing interests**

The author(s) declare that they have no competing interests.

**Authors' contributions**

AY and HM analyzed and interpreted the patient data regarding type I diabetes and MRI imaging. AY and JW was a major contributor in writing the manuscript. All authors read and approved the final manuscript.

**References**

2. Oh SH, Lee KY, Im JH, Lee MS: **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.

**Additional files**

* File name; Movie 1
* File format; Windows Media audio/video file
* Title of data; Hemichorea and bilateral dystonia in a present case.
* Description of data; Whole series of her movement were terminated in 30 seconds. The exactly same pattern of hemichorea and bilateral dystonic movement intermittently occurred every 10 minutes.
Additional files provided with this submission:

Additional file 1: movie 1.wmv, 1144K
http://www.jmedicalcasereports.com/imedia/5719647920359845/supp1.wmv