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CASE REPORT

Diabetic striatopathy in a patient with hemiballism

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Abstract Diabetic striatopathy is a rare and life-threatening manifestation of diabetes mellitus. The disease commonly affects individuals of Asian descent, females, and the elderly. Patients usually present with hemiballism-hemichorea caused by nonketotic hyperglycemia. Hemiballism-hemichorea is defined as involuntary continuous random appearing movement involving one side of the body. This movement disorder may develop secondary to stroke, diabetic striatopathy, neoplasm, infection, Wilson's disease, and thyrotoxicosis. Despite being rare, prompt recognition of a hyperglycemia-induced hemiballism-hemichorea is essential because the symptoms are reversible with correction of hyperglycemia. Diagnosis is possible based on blood analysis and neuroimaging findings. Laboratory tests reveal raised blood glucose and hemoglobin A1C levels which indicate poorly controlled diabetes. Neuroimaging provides suggestive findings of diabetic striatopathy which are hyperattenuation on computed tomography and hyperintensity on T1-weighted magnetic resonance imaging in the basal ganglia. In this case report, our aim is to present neuroimaging findings in an adult man with sudden onset of hemiballism associated with nonketotic hyperglycemia.

Keywords Diabetic striatopathy · Diabetes mellitus · Hemiballism-hemichorea · Nonketotic hyperglycemia

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Introduction

Diabetic striatopathy is a disease characterized by the sudden onset hemiballism or hemichorea in a poorly controlled diabetic patient [1]. This rare clinical manifestation of diabetes mellitus is also defined as "hemichorea-hemiballism in nonketotic hyperglycemia" or "chorea, hyperglycemia, basal ganglia syndrome" in the medical literature [2, 3]. Imaging findings are relatively helpful in the diagnosis of this life-threatening disorder. Early recognition with appropriate treatment is in favor of good prognosis.

In this case report, we aimed to present the neuroimaging findings of a patient with sudden onset hemiballism associated with nonketotic hyperglycemia.

Case report

A 55-year-old man with known diabetes mellitus presented to the emergency department of our institution with sudden onset of involuntary movements at the left upper extremity in the last 3 days. He was conscious with normal mental functions. Neurological examination revealed no obvious motor or sensory deficit. Laboratory tests, computed tomography (CT), magnetic resonance imaging (MRI), and diffusion-weighted imaging (DWI) were performed. On admission, blood tests revealed elevated levels of glucose and hemoglobin A1C; his blood glucose level was 246 mg/dl (N 70–110 mg/dl) and his hemoglobin A1C was 15.4 % (N 7 %). Urine examination was negative for ketones. These laboratory results were consistent with nonketotic hyperglycemia. CT showed faint hyperdensity in the right putamen (Fig. 1). On MRI, the right putamen was hyperintense on T1-weighed image (Fig. 2). DWI showed no diffusion abnormality in the cerebral parenchyma and the basal ganglia (Fig. 3). Diabetic striatopathy was the final diagnosis based on the imaging

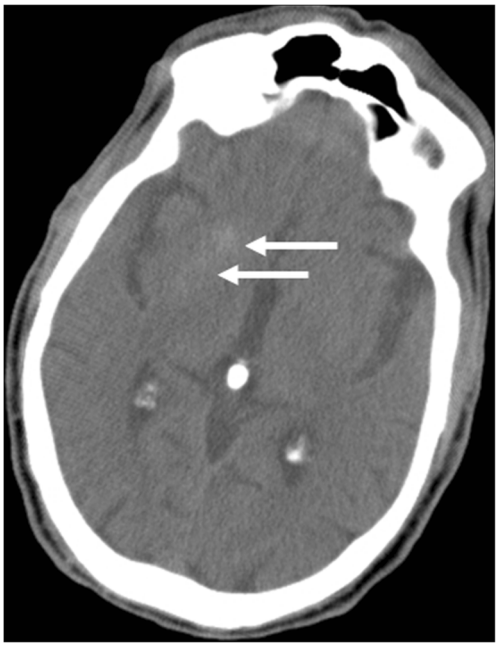

Fig. 1 CT shows subtle hyperattenuation in the right putamen (*arrows*)

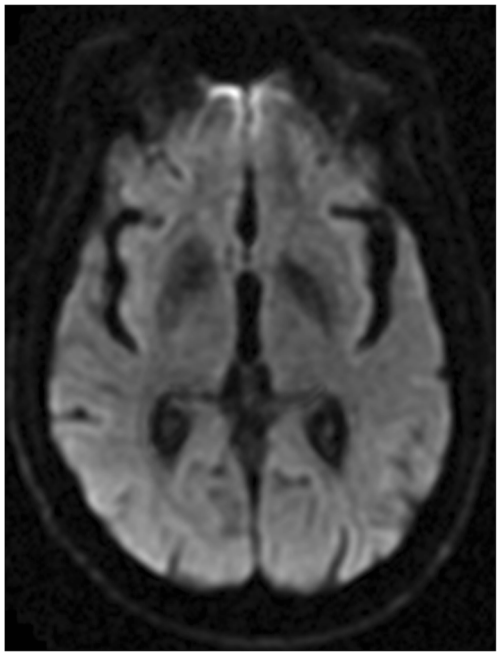

Fig. 3 DWI shows no diffusion restriction in the basal ganglia

findings and the laboratory results. The hemiballism rapidly recovered following insulin treatment.

Discussion

Hemiballism is defined as unilateral large-amplitude involuntary movements of the proximal extremities. Hemichorea is a hyperkinetic disorder characterized by nonrhythmic and nonrepetitive involuntary movements in one side of the body. Hemiballism-hemichorea is a unilateral involuntary movement disorder secondary to abnormalities affecting the contralateral basal ganglia [3]. It most commonly results from ischemic or hemorrhagic stroke [4]. Nonketotic hyperglycemia is the second most common etiology of hemiballism-hemichorea [4]. Neoplasm, infection, Wilson's disease, and thyrotoxicosis may also cause this rare clinical presentation [3].

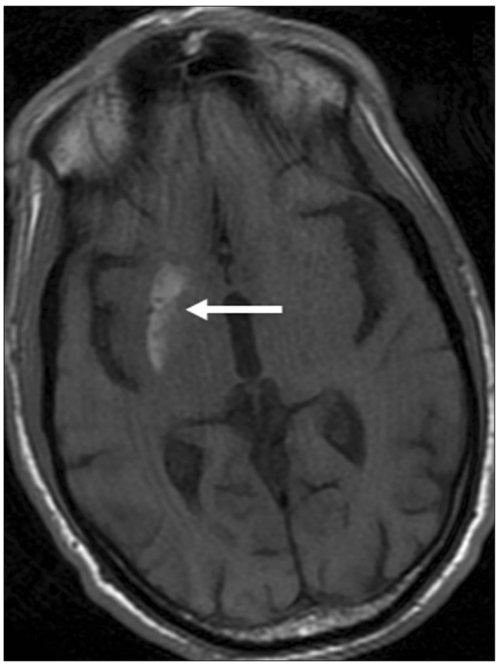

Fig. 2 T1-weighted MRI demonstrates increased signal intensity in the right putamen (*arrow*)

Diabetic striatopathy is typically seen in elderly patients with type 2 diabetes mellitus [3]. Women are affected more commonly than men [5]. It is more common in Asians, a feature which may suggest an underlying genetic predisposition [4]. Patients usually present with acute onset of hemiballism-hemichorea; however, in rare instances, a generalized movement disorder secondary to the involvement of bilateral basal ganglia or stroke-like complaints may be seen [2, 5, 6]. Laboratory findings reveal long-standing uncontrolled diabetes with increased blood glucose and hemoglobin A1C levels and no ketones in the blood and urine.

The pathogenesis of the abnormal movements is not fully understood. According to the suggested theory, hyperviscosity induced by hyperglycemia causes a decrease in cerebral perfusion, an increase in anaerobic metabolism, and a reduced GABA level. The depletion of this inhibitory neurotransmitter results in hemiballism-hemichorea [4]. However, this theory is inefficient to explain unilateral basal ganglia involvement and the clinical presentation.

Diabetic striatopathy has characteristic neuroimaging findings predominantly seen in the putamen. The involvement of

the caudate nucleus may be in association with the putaminal findings [4]. CT may show hyperattenuation in the basal ganglia; however, this finding is not found in all patients. T1-weighted MRI typically demonstrates increased signal intensity in putamen/caudate nucleus [4]. Signal changes are variable on T2-weighted MRI. There is no edema, mass effect, or contrast enhancement on CT or MRI [4]. DWI usually shows no diffusion restriction. Susceptibility-weighted imaging (SWI), a MRI sequence used in detection of venous blood, hemorrhage, iron deposition, or calcification, may show no abnormality or signal loss [7]. Fluorodeoxyglucose (FDG) positron emission tomography (PET) reveals metabolic failure in the affected basal ganglia [8].

The pathogenesis of the imaging findings is controversial. Histological findings include neuronal loss, gliosis, and reactive astrocytosis. No infarction, calcification, or macrohemorrhage is evident on histological or imaging examinations. Mineral deposition, protein denaturation, or petechial hemorrhages are the proposed reasons of the imaging findings [4, 7].

Abnormal movements rapidly recover within hours to few days after the treatment of hyperglycemia in most cases. Imaging findings are also reversible although complete resolution of abnormalities may take up to 6 years after the initial presentation [9].

In conclusion, diabetic striatopathy is a treatable disorder characterized by hyperglycemia-induced hemiballism-hemichorea. Neuroimaging findings associated with an increased blood glucose level indicate the appropriate diagnosis. Diabetic striatopathy should be considered in patients with unilateral movement disorders and contralateral neuroimaging abnormalities (hyperattenuation on CT, hyperintensity on T1-weighted MRI) in the basal ganglia.

Conflict of interest The authors declare that they have no conflict of interest.

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