Case Report

Unilateral hemichorea and hemiballismus: Rare complications of non-ketotic hyperglycemia

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ABSTRACT

Hemichorea-hemiballismus is an extremely rare and dramatic complication of non-ketotic hyperglycemia in patients with uncontrolled diabetes mellitus. Here, we present the case of a 65-year-old male, known hypertensive, diabetic with the left eye retinopathy who presented with the chief complaints of sudden onset hemichorea and hemiballismus on the right upper and lower limbs. Magnetic resonance imaging brain reported T1 hyperintense and T2 isointense/hypointense signal in the left putamen with no restricted diffusion – these signal changes. He was started on oral antihyperglycemics and a combination of tetrabenazine, clonazepam, and haloperidol. Mild resolution of symptoms was seen and the patient is on follow-up with advice to control sugar through diet and regular sugar monitoring.

Key words: Diabetic complications, Hemichorea-hemiballismus, Hyperglycemic hyperosmolar non-Ketotic syndrome, Movement disorders

Hemichorea-hemiballismus is an extremely rare and dramatic complication of non-ketotic hyperglycemia (NKH) in patients with uncontrolled diabetes mellitus. The presence of continuous, jerky, movements of one side of the body, often due to focal lesions in the contralateral side basal ganglia is characteristic of this disease. Ischemic changes in the striatum associated with hyperglycemia and hyperviscosity are assumed to be the pathophysiology behind this dramatic event [1].

NKH is the most common metabolic cause of hemichorea-hemiballismus and it is a reversible condition most often. Chorea and ballism can be reversed through early management of the blood glucose levels in the patient. Various conditions such as cerebrovascular insufficiency, neurodegenerative diseases, neoplastic diseases, immunologic diseases, infectious diseases, and metabolic diseases are known as secondary causes of this rare disorder [2]. Although hyperglycemia is the most common cause, clinical characteristics and mechanisms remain unclear. We present a case of diabetes mellitus 2 who initially presented with NKH later followed by sudden onset of hemichorea and hemiballismus.

CASE REPORT

A 65-year-old male, known hypertensive, diabetic with the left eye retinopathy and on regular medication presented to the Shri Krishna Hospital Trauma Emergency Care with the chief complaints of sudden onset hemichorea and hemiballismus on the right upper and lower limbs. There was a prior history of admission to a civil hospital 10 days back with a blood glucose level (>500 mg/dl). On the 6th day of admission, the patient developed sudden onset choreiform and ballism movements on the right upper and lower limbs and was shifted to the Manavala Lakva Hospital and finally referred to our center.

On presentation, the patient was vitally stable, conscious, well-oriented, and was shifted to the ward. The central nervous system examination revealed no loss of cognition, power of 5/5, and tone in all four limbs. Cerebellar and sensory examinations were normal and only extrapyramidal presentation being hemichorea and hemiballismus on the right side with a gradual onset and progressively worsening into a large amplitude, flailing movements of unilateral upper and lower limbs, and with an inability to walk properly on the right side. The initial clinical impression was sudden onset hemichorea and hemiballismus in a known case of hypertension and diabetes mellitus. Differential diagnosis included NKH or cerebrovascular stroke due to infarct or hemorrhage. The patient displayed no seizure-like activity.

Magnetic resonance imaging (MRI) brain (limited axial study) was performed by obtaining 4 mm thick T1-weighted, T2-weighted, diffusion-weighted, apparent diffusion coefficient (ADC), and susceptibility-weighted imaging (SWI) minimum

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intensity projection axial images. T1 hyperintense (Fig. 1a) and T2 isointense/hypointense signal (Fig. 1b) were seen in the left putamen with no restricted diffusion (Fig. 2). These signal changes were consistent with the differentials of NKH. Few discrete 2/FLAIR hyperintense foci were seen in bilateral corona radiata and periventricular white matter which were suggestive of chronic small vessel ischemic changes. Foci of blooming on SWI (T2*) were seen in bilateral lentiform nuclei, suggestive of physiological calcification/metal deposition. Both the lateral ventricles and third ventricle were mildly dilated. The fourth ventricle appeared normal. Basal cisterns and bilateral cerebral sulci were also prominent, suggestive of generalized cerebral atrophy, most likely age related. MRI brain ruled out differentials of infarct or bleed and clinical impression of NKH was confirmed.

Dosing of oral antihyperglycemic was adjusted according to the requirements and daily sugar monitoring was done. He was started on tetrabenazine, clonazepam, and haloperidol with mild clinical improvement, and the patient was discharged when vitally stable. The patient presented on follow-up with mild symptoms and decreased intensity of extrapyramidal signs of hemichorea and hemiballismus and was advised regular sugar monitoring and repeat MRI after complete suspension of initial symptoms.

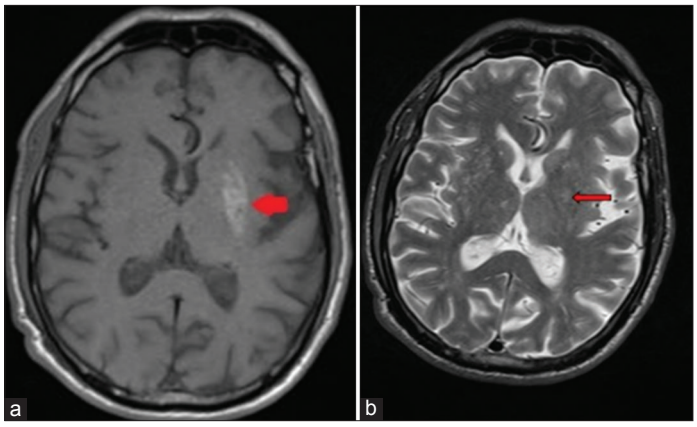

Figure 1: (a) Focal hyperintensity noted in the left putamen on T1-weighted image (marked by red arrow); (b) focal hypointense signal is noted in the left putamen on T2-weighted image (marked by red arrow)

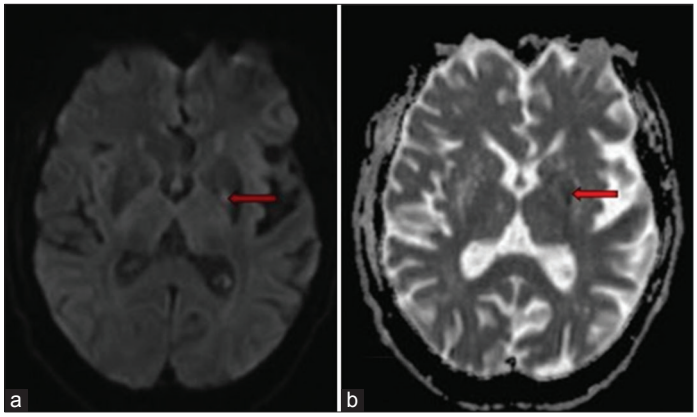

Figure 2: No diffusion restriction is noted in diffusion-weighted imaging-apparent diffusion coefficient image (marked by red arrow)

DISCUSSION

Hyperkinetic movement disorders are a vast category of extrapyramidal signs and chorea and hemiballismus fall under this category. Chorea is rapid, involuntary, non-rhythmic, non-stereotypical, small amplitude movements while ballismus is proximal chorea that produces flinging movements of high amplitude. Hemichorea and hemiballismus is the involvement of one half of the body due to disinhibition at the contralateral striatum or subthalamic nucleus. Cerebrovascular insufficiency, neurodegenerative diseases, neoplastic diseases, immunologic diseases, infectious diseases, and metabolic diseases are known as secondary causes of this rare disorder [2].

Bedwell *et al.* were the pioneers to report hemichorea-hemiballism caused by NKH. They reported the case of a 65-year-old woman presenting with ballistic movements in all four limbs after episodes of hyperglycemia [3]. Various hypotheses have been proposed to explain the pathophysiology, the most commonly invoked theory suggests that depletion of gamma-aminobutyric acid (GABA) and acetylcholine, which are needed as an alternative energy source during NKH, leads to a decreased inhibitory signal to the thalamus resulting in hyperactive movement [2]. However, Satish *et al.* reported that chorea-ballism can also manifest in ketotic patients where acetoacetate is abundant and can be utilized as a source of GABA, suggesting that metabolic alteration might not be the sole mechanism [4]. The pathophysiology remains questioned till date.

Choreiform and ballism develops usually after initial attacks of hyperglycemia, however, several reports depict otherwise, Ifergane *et al.* in their study reported involuntary symptoms being the presenting symptoms of diabetes without any previous history of hyperglycemic attacks and also proposed that all cases of sudden hemichoreiform and ballism should be suspected for undiagnosed diabetes mellitus [5]. Cosentino *et al.* in their report of 20 cases presented that all patients on admission had hyperglycemia except one which was evaluated 4 weeks after involuntary movement onset. Our patient on admission had a blood glucose level of 500 mg/dl which coincided with this finding. They also reported a higher incidence of NKH-induced hemichorea-hemiballismus in Asians but cases were also reported in some other ethnic groups worldwide. Our report is of a patient in an Asian setting [6].

Since then, several cases have turned up and varied pathophysiology has been derived. One of the largest study samples of hemichorea-hemiballismus with 53 patients was documented by Sh *et al.* [7]. In their study, out of the 53 patients, about 47 presented with hemichorea and only around six had bilateral chorea and the body parts affected by chorea were upper and lower extremities in almost 30 patients. Our study correlated with the findings with the patient having hemichorea on the right side upper and lower extremities which was the most common presentation.

High signal intensity basal ganglia lesion on the T1-weighted brain MRI was reported in all patients. The putamen was involved in 31 patients. There were no patients with an isolated lesion confined to the caudate nucleus or the globus pallidus. In

contrast to T1, T2-weighted studies showed low signal intensity basal ganglia lesions in 24 patients, isosignal intensity lesions in 17 patients. Our findings revealed similar findings with T1 hyperintense, T2 isointense/hypointense signal seen in the left putamen and no diffusion restriction is noted in diffusion-weighted imaging-ADC (DWI-ADC) image.

Nath *et al.* in their study found iterated T1 hyperintensity involving the right basal ganglia, including the globus pallidus and T2 weighted, and DWI sequences showed low signals. Restricted diffusion was not seen on DWI or an ADC map. Multiple old infarcts were also seen in the contralateral cerebral hemisphere and cerebellum [8]. Our findings were in favor with additional findings of lateral ventricles and third ventricle which were mildly dilated. Normal fourth ventricle and basal cisterns and bilateral cerebral sulci were prominent suggestive of generalized cerebral atrophy which was most likely age related.

Ohara *et al.* in an autopsy report of a 92-year-old male with hemichorea-hemiballism associated with hyperglycemia and striatal hyperintensity on T1-weighted MRI, described histological findings of multiple foci of recent infarcts on the putamen contralateral to the hemiballismus [9]. This was contrasted by Fujioka *et al.* in their study which reported that transient internal carotid artery-middle cerebral artery occlusion which leads to spectacular shrinking deficit produces a specific ischemic change with delayed onset in the basal ganglia and cerebral cortex in humans on MRI but not on computed tomography scans. They speculated that the lesions represented incomplete ischemic injury, including selective neuronal death, a proliferation of glial cells, paramagnetic substance deposition, and/or lipid accumulation [10].

Shan *et al.* reported the presence of hyperintense putamen precedence in two patients out of 10 in the study cohort which coincided with our findings and they concluded that neurons in the ventral striatum and striatonigral pathway played a critical time in the generation of ballism in the patient [11]. Hemichorea and hemiballismus is reversible complication with a good prognosis and can be treated by controlling the hyperglycemia as reported by Lin and Chang in their study [12].

Our patient showed significant improvement in the reduction of hyperglycemia by oral antihyperglycemic agents and a combination of tetrabenazine, clonazepam, and haloperidol. Sitburana and Ondo reported the first use of tetrabenazine in hyperglycemia-induced hemichorea-hemiballismus and it showed dramatic results. Tetrabenazine has been a known neuroleptic and it inhibits vesicular monoamine transporter 2, it is also a mild dopamine inhibitor. Rapid action of tetrabenazine on symptoms suggests a dopaminergic or less likely adrenergic contribution to pathophysiology. This contradicts the usual hypothesis described before [2,4,13]. Our patient showed improvement to some stage but complete resolution of symptoms was not found when the patient presented to follow-up with milder symptoms than before. Furthermore, the efficacy of neuroleptics was not determined as the improvement could also be attributed to the control over hyperglycemia due to oral antihyperglycemic agents. The patient is still on follow-up with advice to continue regular medication.

CONCLUSION

Unilateral hemichorea and hemiballismus are rare complications but a more common presenting sign of NKH and this differential should always be considered to exclude this from a cerebrovascular stroke. Hemichorea and hemiballismus presents with specific striatal lesions on brain MRI and this forms the basis of their diagnosis.

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