CASE REPORT

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Diabetic Striatopathy: Case Report and Review

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Abstract

Background: Diabetic striatopathy (DS) also known as hyperglycaemic non-ketotic hemichorea-hemiballismus syndrome presenting with uncontrollable involuntary jerky motions with ballism, typically more proximal, with large amplitude and choreic movements as distal, largely associated with poorly controlled diabetes mellitus. The entity is a rare disorder with an estimated prevalence of 1 in 1,00,000. Screening of patients, young or elderly who present with involuntary movements and hyperglycaemia or presenting without prior history of diabetes mellitus is essential[1]. Unfortunately it's prevalence in the literature is low owing to unfamiliarity of this entity amongst physicians.

Case presentation: 63-year-old male patient having 10 years history of uncontrolled Type 2 diabetes mellitus and essential hypertension presented to our hospital with acute onset of involuntary movements as hemiballismus-hemichorea (HB-HC) of left upper and lower extremities, and involving face and neck. These movements were not noted in sleep. The patient discontinued the diabetes medications a month prior to the onset of the present problem. MRI brain done revealed altered signal intensities in the right lentiform nucleus consistent with a clinical diagnosis of non-ketotic hyperglycaemic HB-HC. This clinical entity is described as diabetic striatopathy. He received insulin injections, tetrabenazine as anti-choreic drug with vitamins and minerals as supportive therapy. The patient had a complete remission of symptoms within just 2 weeks of control of blood sugar.

Conclusion: Diabetic striatopathy (DS) is an uncommon neurological condition of uncontrolled hyperglycaemia induced hemichorea/hemiballismus syndrome. The clinical presentation and brain MRI findings highlight its recognition while stringent glycaemic control with use of anti-choreic drugs revert the manifestations of DS.

Keywords: Hemichorea-Hemiballismus, uncontrolled diabetes mellitus, brain MRI; anti-choreic drug; basal ganglia.

Introduction

Diabetic striatopathy (DS) which is variously known as hyperglycaemic non-ketotic hemichorea-hemiballismus or chorea hyperglycaemic basal ganglion syndrome[1]. Both chorea and ballismus refer to random, uncontrollable, involuntary jerky motions associated with poorly controlled diabetes mellitus, either type 1 or 2. The development of HB-HC is associated with hyperintensity on T1-weighted magnetic resonance imaging of putamen, caudate nucleus and globus pallidus either in isolation or combined. These findings of MRI in presence of non-ketotic hyperglycaemia is considered a *sine qua non* of this entity[2,3].

Case presentation

A 63-year-old male, non-smoker, hypertensive and with uncontrolled diabetes mellitus type 2 presented with continuous involuntary movements of left upper and lower extremities and also involving face and neck. Movement disorders were observed 2 days before presenting to this hospital. He was diabetic for 10 years, on metformin and glimepiride but with poor adherence. He discontinued his diabetes medications 6 weeks prior to hospitalisation without any justification. History of viral febrile illness, COVID-19 and intake of any drugs/alternative medicines was denied. History of movement disorders in the family was also denied. His speech was unaffected. The gait was unsteady and interrupted by violent motions, but suppressed during sleep.

On examination, patient was conscious, oriented and had normal vital features. His height was 172 cms, weight of 60 kgs, and BMI of 20.3 kg/m^2. He had violent high amplitude choreoathetotic movements of the left side of the body including the face, neck and trunk. High amplitude movements were observed at the shoulder, hip, elbow, and ankle (video on *www.jiacm.in*), pupils were reactive to light with normal accomodation reflex. Eye movements were normal in all directions. No nystagmus was seen. The motor and sensory systems were unremarkable. Co-ordination could not be tested on the left side due to irregular movements. Deep tendon and plantar reflexes were normal. Routine investigations were within normal limits with random blood sugar levels of 440 mg/dL and HbA1c of 14% (Normal range - 3.8 - 5.6%). Urine ketones were negative, serum ketones were not tested. MRI T1-weighted images of the brain revealed

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hyperintense signal in the right lentiform nucleus. Diffusion weighted imaging (DWI) revealed subtle restricted diffusion in right lentiform nucleus. No evidence of blooming seen on GRE sequences. Mild age-related atrophic changes were also noted. Based upon the characteristic presence of HB-HC movements, poorly controlled diabetes mellitus, negative family history of chorea and characteristic MRI findings; a strong presumptive possibility of diabetic striatopathy was entertained. Insulin injections for better blood glucose management and tetrabenazine 12.5 mg twice daily for control of abnormal movements were initiated with supportive nutritional supplements. The patient demonstrated substantial improvement in movement disorder in next 2 weeks of time. A follow-up visit after 3 months showed no residual movement abnormalities. The blood glucose levels were well controlled, with HbA1c improvement from 14% to 7.4%.

Discussion

India is the capital of the diabetic world. Involuntary movements of varied nature are described in association with diabetes mellitus. The myriad aetiopathological concepts of this entity include neurogenetic, autoimmune, infectious, inflammatory. metabolic and neurodegenerative entities as potent aetiologies of this clinical syndrome. The clinical, radiological and laboratory correlation, as observed in the case, narrows down the differential diagnoses of this potential condition. The common presentation of DS is with HB-HC movement disorder, occurring in a subject with poorly controlled diabetes mellitus. Reversibility in clinical, imaging and neurological deficits is invariably observed on achieving adequate blood sugar control.

Choon-bing Chao and coworkers in 2020[4] identified 72 articles comprising of a total of 176 patients of which 96.6% were diabetic subjects, and of these 17% were newly diagnosed diabetics. The average blood glucose level described was 414 mg/dL and HbA1c of 13.1%. The present case manifested as violent unilateral HB-HC movements and had a blood glucose level of 440 mg/dL and HbA1c of 14%. These findings of the case raised the possibility of diabetic striatopathy. These movements disappeared during sleep. However, in literature a few cases are reported with partial or no suppression of DS abnormal movements during sleep[5]. Among patients with poorly controlled diabetes mellitus (HbA1c 10%) hospitalised for any cause, 0.58% had DS, while incidence rose to 1.2% among these who were hospitalised for neurological disorders[6]. The disease prevalence is higher in females than males with male: female ratio of 1:1.77[4]. The hemichoreas are also seen in diabetes type 1[7]. The reported low prevalence of the disease could be because of unfamiliarity of this entity amongst physicians, leading to an erroneous diagnosis of intracerebral haemorrhage. This confusion of diagnosis is noticeable chiefly with findings of hyperdensities on CT[8]. High prevalence of this entity in females is attributed to estrogenic changes that are observed in post-menopausal women (representing elderly age group) affecting GABA and dopamine receptors.

There are several considerations for MRI sequence findings of hyperintensities. These imaging abnormalities could be induced by a) presence of calcifications (should show HU value of 80) b) microhaemorrhages which are often transient, acute in onset and match with tissue density for hyperdensity of 40-50 HU[9]. c) T1 shortening and hyperintensities are found with a reduction in velocity of blood flow, presence of fat, high protein contents in the cell and presence of paramagnetic metals, viz., iron, zinc, copper and manganese[10]. In absence of mass effect or oedema, the presence of MRI findings is congruous with the concept of microbleed in tissue. Mestre and colleagues in 2009 demonstrated deposits of hemosiderin in putamen and suggested that hyperglycaemic status may transiantly induce blood-brain barrier dysfunction, resulting in RBCs and hemosiderin extravasation in putamen[11]. Protein hydration may cause swollen gemistocytes inside the cytoplasm as seen in sclerosing panencephalitis or epilepsy with resultant hyperdensities. Prolonged period of hypoperfusion and ischaemia often result in dysfunction of neurons. The hypoperfusion does not explain the presence of bilateral chorea with observed unilateral pathology on MRI. A few cases are reported with presence of focal microhaemorrhages based upon this vascular theory. Applications with gradient echo-weighted (T2-GRE) and susceptibility weighted imaging (SWI) MRI sequences help further to narrow the wide differential diagnoses. Presence of paramagnetic materials in basal ganglion like copper, iron or manganese simply can be excluded as the cause of hyperdensities with judicious clinical assessment of patient. Copper and iron disorders may have a chronic course as seen with Wilson's and other degenerative disorders. Manganese deposition is usually related to parkinsonism while fat separation images can rule-out the possibility of presence of fat as the cause of MRI hyperintensities. The resolution of involuntary movement disorder is highly variable from few days to about 10 months after correction of hyperglycaemic status. Few cases are described with partial improvement between 3 months and 5.6 years[12]. Most patients also need anti-choreic drugs to achieve symptom control. Tetrabenazine (TBZ) exerts anti-choreic effects by decreasing the amount of dopamine in the brain. Further, this drug has the highest binding in caudate nucleus and putamen. These are the pathological locations in most diabetic striatopathies. Resistant cases not

responding to conventional therapies may require deep brain stimulation of putamen, caudate nucleus, and subthalamic nucleus for control of movement disorder.

Conclusion

The recognition of the entity of DS is important in cases of uncontrolled diabetes mellitus. The clinical scenario and brain MRI findings that are consistent with entity, support this diagnosis. Stringent glycaemic control and use of anti-choreic medications help the to achieve faster recovery.

Patient video available at *www.jiacm.in*

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