Non-Ketotic Hyperglycemia Induced Chorea (NKHIC): A Case Report and review of literature

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Abstract

Case report: we report a 68-year-old woman known diabetic patient who presented with uncontrolled involuntary movement of left half of body associated with non-ketotic hyperglycemia and respond to treatment of hyperglycemia.

Methods: A retrospective review of clinical, radiological and laboratory findings in a patient with chorea associated with non-ketotic hyperglycemia.

Results: we report a patients presented with acute hemichorea, with a history of long standing diabetes mellitus and uncontrolled diabetes mellitus. The initial cranial CT scans revealed hyper density bilateral putamen and caudate nuclei more marked on right side contra-lateral to the side of chorea. Her laboratory investigation showed a non-Ketotic hyperglycemia and, hence, hyperglycemia-induced hemi-chorea diagnosis was made. Chorea partially responded to treatment for hyperglycemia and patient also treated with tetrabenzine.

Conclusion: Hemi-chorea associated with diabetic non-Ketotic hyperglycemia may present with acute neurologic manifestation resemble stroke. Diagnosis of this clinical and radiological condition is important because treatment of the underlying hyperglycemia will lead to correction of chorea.

Keywords: diabetic non-ketotic hyperglycemia, CT scan,chorea, putamen, hypedensity

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Introduction:

Chorea, an involuntary jerky movement, consists of random and fast jerking motions in distal parts of the limbs. Chorea can be caused by a wide variety of vascular, infection, drugs degenerative, and toxic metabolic diseases.[1] which usually involve basal ganglia particularly the striatum. Systemic lupus erythematosus is the most

common systemic disorder that causes chorea; the chorea can last for days to years. Choreas can also be seen with hyperthyroidism, autoimmune disorders including Sjögren's syndrome, infectious disorders including HIV disease, metabolic alterations, polycythemia rubra vera (following open-heart surgery in the pediatric population), and in association with many medications (especially anticonvulsants, cocaine, CNS stimulants, estrogens, lithium). Chorea can also be seen in paraneoplastic syndromes associated with anti-CRMP-5 or anti-Hu antibodie.Non-ketotic hyperglycemia has been occasionally associated with hemiballism-hemichorea (HB-HC).[2] A characteristic clinical and radiological feature could provide an early clue for diagnosis and treatment.

Case report:

We report a 68-year-old woman known diabetic patient from 24yrs was admitted due to an acute-onset of chorea involving her left half of body. There was no history of exposure to neuroleptic drugs, hyperthyroidism, autoimmune disorders including SLE, Sjögren's syndrome, infectious disorders including HIV disease and trauma. She had no history or family history of movement disorders.Her blood glucose level was 585 mg/dl and glycosylated hemoglobin A1C was 11.9%.Urine for ketones negative. Other routine blood tests including CBC, RFT and serum electrolytes were normal. Her brain non-contrast CT scan showed hyper density at the bilateral putamen and caudate nuclei more marked on right side (figure-1, 2). The chorea completely disappeared following correction of hyperglycemia and tetrabenzine (250mg/day) orally.

Discussion:

Non-ketotic hyperglycemia is an unusual cause of hemiballism-hemichorea.[2] this condition was first described by Sanfield et al in an uncontrolled diabetic patient presented with chorea, who's CT showed unilateral faint hyperdensity over the putamen.[3] Clinical manifestation can be either unilateral or bilateral. If unilateral, the imaging abnormality is typically at the contra-lateral basal ganglia. Non-ketotic hyperglycemia associated HB-HC is more commonly occurs in females than males and most reported cases are elderly Asians, suggesting a genetic influence or an inadequate diabetes control in this less developed countries.[4]

The mechanism of brain damage in HB-HC associated with hyperglycemia is not completely known. A meta-analysis of 53 case of non-ketotic hyperglycemia-induced

chorea by Oh et al., suggested synergistic effects of uncontrolled non-ketotic hyperglycemia and vascular insufficiencies cause an incomplete transient dysfunction of the striatum and chorea.[4] A reduction in both GABA and acetylcholine in the basal ganglia in combination with metabolic acidosis and the lack of energy production may then cause a basal ganglia dysfunction and, subsequently, chorea.[4] An ischemic process is another possible mechanism of non-ketotic hyperglycemia associated HB-HC.[4] An explanation of the imaging appearance on CT and MRI is still largely under debate. MRI examination of the previously reported case revealed an increased density on CT which corresponded with an abnormal hyper signal intensity in T1-Weighted and normal or hypo signal intensity on T2-Weighted or gradient-echo T2-Weighted images.[7] These findings yield only a few differential diagnoses of intracranial lesion including methemoglobin.[6] All findings accompany with continuous resolution of high density on CT scans suggest the evolution of a petechial hemorrhage with hemosiderin deposition rather than previously a postulation of calcification, which is unlikely to resolve over time.[7] One study supported a hypothesis of petechial hemorrhages as the cause of this syndrome, possible secondary to erythrocyte diapedesis due to hyperglycemia induced blood-brain barrier dysfunction.[8] Hyper viscosity caused by hyperglycemia can result in partial neuronal death and dysfunction on the vulnerable striatum, similar to the partial ischemic injury models, and may be associated with Wallerian degeneration of the internal white matter of the putamen.[5,9] The protein desiccation that occurs during the course of Wallerian degeneration could explain the hyper attenuation in the noncontrast CT brain, and the hyper signal intensity on T1-Weighted and restricted diffusion in the DW images in the early phase.[9] one case revealed calcium depositions and a focal micro hemorrhage within a confluent area of an infarct.[10] one case showed astrocytic gliosis and extra vascular hemosiderin depositions consistent with small previous hemorrhage in the striatum.[11]

Our patient presented with the classic triad of acute chorea, non-ketotic hyperglycemia and hyper density on CT scan. The diagnoses should be made promptly with no further complicated investigation. Involuntary movements in patient completely recovered after correction

of hyperglycemia and treatment with tetrabenzine.

Conclusion:

Hemi chorea associated with diabetic non-ketotic hyperglycemia may present with acute neurologic manifestation resembling stroke. Chorea associated with nonketotic hyperglycemia should be early diagnosed because early treatment of hyperglycemia could lead to partial or complete recovery of involuntary jerking movement. Further study is required to understanding the pathophysiology of this rare complication of hyperglycemia in uncontrolled elderly diabetic patient.

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Figure 1 Non-contrast CT scan showed hyper density at the bilateral putamen and caudate nuclei more marked on right side

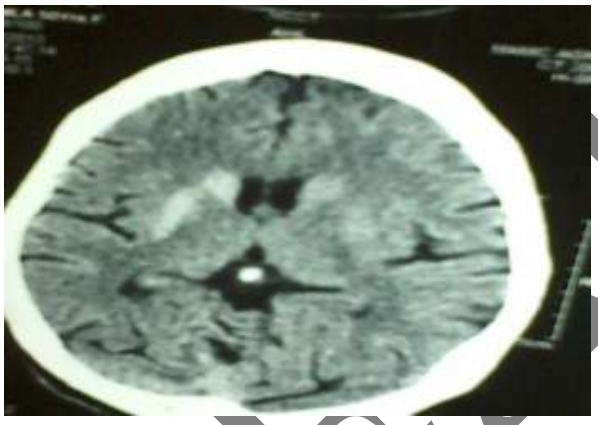

Figure:2 Non-contrast CT scan showed hyperdensity at the bilateral putamen and caudate nuclei more marked on right side