Chorea Associated with Nonketotic Hyperglycemia (Diabetic Striatopathy) in an Elderly Male

Sir,

In a patient of diabetes with nonketotic hyperglycemia (NKH), high signal intensity lesions on T1-weighted magnetic resonance imaging (MRI) confined to basal ganglia and contra lateral chorea is a unique syndrome often termed as diabetic striatopathy.[1] We present the details of an elderly person who was unsuccessfully treated for his movement disorder for many weeks before a proper diagnosis was made.

A 64-year-old male, not a known case of diabetes, with a past history of poorly controlled hypertension, noticed sudden onset of choreo-ballistic movements of the right upper and lower extremities for the past 3 months. These involuntary movements were present even during sleep. He was seen by a local physician and was put on trihexiphenidyl tablets with minimal improvement in symptoms. At present patient was admitted with history of osmotic symptoms for few weeks, investigations revealed random blood glucose of 490 mg/dL. On examination, he had an unstable gait, hypotonia in right upper and lower limbs. He had uncontrollable involuntary chorea like movements more in his right lower than upper limb. His muscle power was normal and systemic examination was unremarkable.

Investigations revealed initial plasma glucose of 490 mg/dL, with HbA1c of 14.4%, serum sodium was 130 meq/L, and calculated serum osmolality was 290 mosm/kg. Serum ketones were negative and the venous blood gas analysis was normal. Serum creatinine was 1.2 mg/dL and the liver enzymes were normal. A review of non-contrast computed tomography (CT) of brain done 3 months back which was reported as normal, revealed subtle hyperdensity in the basal ganglia region. MRI brain showed T1 hyperintensity in the head of left caudate nucleus and putamen with T2 hypointensity in the same areas [Figure 1]. With this clinical presentation, NKH and MRI findings, a diagnosis of diabetic striatopathy was made. His diabetes was managed with insulin and he was put on low dose of haloperidol (0.25 mg twice daily) to control his involuntary movements. Within 5 days of admission, he showed marked improvement in symptoms and choroid form movements disappeared during his sleep. The patient is doing well and under follow up with a plan to repeat MRI brain after 3 months.

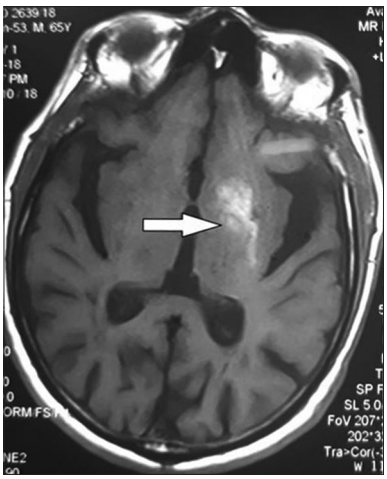

Figure 1: Magnetic resonance imaging brain showing T1 hyper-intensity in the basal ganglion region (arrows)

Hemichorea-hemiballismus, as a manifestation of NKH, is a rarely described entity, typically seen in elderly Asian women with type 2 diabetes, though very rarely it has also been reported in type 1 diabetes, those with diabetic ketoacidosis and in children.[2,3] It is characterized by high signal on T1-weighted MRI, which is likely due to accumulation of lipid-laden macrophages, confined to the striatum. Most accepted pathophysiological mechanism in diabetic striatopathy involves hyperviscosity leading to local tissue hypoperfusion, depletion of gamma-Aminobutyric acid, and accumulation of manganese-containing gemistocytes in the basal ganglia which typically appear as T1 hyperintense lesions.[1] In a large meta-analysis of 53 cases, two-third of

the patients had complete resolution of the symptoms during follow up after 3 months, one-third of them only with control of hyperglycemia.[4] The high signal intensity T1-weighted lesion in basal ganglia is not unique to this syndrome, it may also be seen in hepatic encephalopathy, post-cardiac arrest encephalopathy, and hypoglycemic coma.[4] Though generally having a benign outcome, this condition can lead to caudate atrophy and persistent movement disorder if not recognized and treated properly.[5] In the present case, a significant resolution of symptoms was seen despite late diagnosis and treatment.

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Conflicts of interest

There are no conflicts of interest.

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