Diabetic striatopathy- rare case of Chorea associated with Non-ketotic hyperglycemia

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ABSTRACT

Non-ketotic hyperglycemia (NKHG) has been reported to increase the probability of seizures and movement disorders especially in elderly population. Here we report a case of long standing diabetes presenting with choreoathetoid movements and poor glycemic control. They may get frequently misdiagnosed to have neurological disease, however recognition of this association is important as the condition may quickly resolve when NKHG is controlled.

Keywords: *diabetic striatopathy, hyperglycemia*

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INTRODUCTION

Hyperkinetic movement disorders are characterized by involuntary movements not associated with weakness. Several types of hyperkinetic disorders have been described like tremors, chorea, hemiballismus, athetosis, tics and dystonia. Chorea is a rare movement disorder that can be familial or seen to be associated with a number of other disorders¹. Association of chorea with vascular diseases, metabolic derangements like hypoglycemia² and hyperglycemia^{3,4} has been described in literature, though rare. Chorea associated with non-ketotic hyperglycemia (NKHG) and high signal intensity lesions on T1-weighted brain magnetic resonance images (MRI) is recognized as a unique syndrome, known as diabetic striatopathy. This has been mainly described to affect elderly women³. However, its overall clinical features and pathogenesis are still unclear. Here we describe a case of this unique syndrome in an elderly male who presented with choreoathetoid movements and NKHG.

CASE REPORT

55y/M, plumber by occupation, known case of T2 DM for 5 years, complicated by tripathy presented with history of sudden onset choreoathetoid movements right hand and

right angle of mouth for 10 days. These involuntary movements were present even during sleep. There was no history suggestive of connective tissue disorders, fever, use of anticonvulsants, cocaine, CNS stimulants or lithium. There was no family history of movement disorder. On examination there were uncontrollable slow, distal, writhing, involuntary, semi-purposeful choreoathetoid movements involving right arm and hand with some involuntary chewing movements involving right angle of mouth, however gait was normal. His muscle tone, power, reflexes and sensory examination was normal. Systemic examination was unremarkable. Investigations revealed random blood glucose of 468 mg/dl, HbA1c- 9.9 %, serum sodium 134mmol/l, calculated serum osmolality 294mosm/kg, serum ketones were negative normal venous blood gas analysis and thyroid function test. Serum creatinine-1.8mg/dl, eGFR-41 ml/min/1.73m², normal hemogram, peripheral blood film and liver function tests. MRI brain was done which revealed T1 hyperintensity, T2

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hypointensity in left basal ganglia region suggestive of diabetic striatopathy. Keeping in mind his clinical features, NKHG and characteristic MRI features, diagnosis of diabetic striatopathy was made. Hyperglycemia was managed with insulin therapy- basal bolus regimen. Besides insulin therapy, patient was started on tetrabenazine with marked improvement of clinical features within 3 days of start of treatment. Repeat MRI brain was planned after 3 months to look for resolution of brain lesion.

DISCUSSION

Choreoathetosis, is a rarely described entity, typically seen in elderly women with T2DM and NKHG. However it has been also reported in patients with T1DM children⁵ and those with diabetic ketoacidosis⁶. It is characterized by high signal intensity T1 weighted MRI confined to basal ganglia⁷⁻¹¹. Initially these lesions were attributed to be due to post-anoxic ischemic calcifications; frequent disappearance of lesion on follow-up suggested it to represent petechial hemorrhage rather than calcification. A biopsy and postmortem pathological studies have shown selective neuronal loss, gliosis, and reactive astrocytosis at the striatal areas, although, no apparent hemorrhage was reported^{7,10}. Precise mechanism of diabetic striatopathy is unknown, various causative hypothesis have been postulated. In hyperglycemia, the cerebral metabolism shifts to the anaerobic pathway leading to increase metabolism of GABA, and decreased regeneration of acetate leading to decreased acetylcholine synthesis. A reduction in both GABA and acetylcholine in the basal ganglia and the lack of energy production may then cause a basal ganglia dysfunction and subsequent chorea.¹¹. Other postulated mechanism is that poorly controlled diabetes mellitus increases the risk of cerebrovascular ischemia and reduces cerebral blood flow¹². Combined effects of neurotransmitter deregulation, hyperglycemia and vascular insufficiencies cause an incomplete and transient dysfunction of the basal ganglia clinically manifesting as chorea.

Radiologically, lesion is characterized by characteristic hyper-intense basal ganglia lesions in T1-weighted brain MRI, but the T2-weighted brain MRI findings vary. High signal intensity in basal ganglia region on the T1-weighted MRI studies is not specific to diabetic striatopathy and can be observed in chronic hepatic encephalopathy¹³, post-cardiac arrest encephalopathy¹⁴, hypoglycemic coma¹⁵, and mild focal ischemia¹⁶.

Patient usually respond well after management of hyperglycemia with insulin therapy, with variable time to response from one day to months^{17,18}. Some patients need

other medications temporarily to control involuntary movements. But overall prognosis has been reported to be excellent¹⁷⁻²⁰, and only few atypical cases with delayed onset after resolution of the hyperglycemia, unremitting severe disorder movements, and late recurrence have been reported^{8,9}.

To conclude diabetic striatopathy as a rare cause of chorea with characteristic T1 hyper-intense lesion in basal ganglia region occurs in elderly women with longstanding and poorly controlled diabetes, but not exclusively. Its clinical presentation is variable course is benign, and most patients have transient movement disorders that respond well to medical treatment as evident with resolution of clinical and radiological features. Thus, diabetic striatopathy should be kept in mind especially while managing an elderly patient of movement disorder and underlying longstanding uncontrolled diabetes and typical lesion on MRI, due to good prognosis of disease if managed properly.

Fig 1a: T1W MRI showing hyperintensity at caudate nucleus (Arrow) and lentiform nucleus (Star)

Fig 1b: T1W MRI showing normal hypointense caudate nucleus (Arrow) and lentiform nucleus (Star) Comparative image.

Fig 2a: FLAIR MRI showing isointensity at caudate nucleus (Arrow) and iso/hypointense lentiform nucleus (Star)

Fig 2b: FLAIR MRI showing normal hyperintense caudate nucleus (Arrow) and lentiform nucleus (Star) Comparative image.

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