ORIGINAL RESEARCH PAPER

General Medicine

CHOREA: A RARE PRESENTATION OF NON KETOTIC HYPERGLYCEMIA

KEY WORDS:

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ABSTRACT

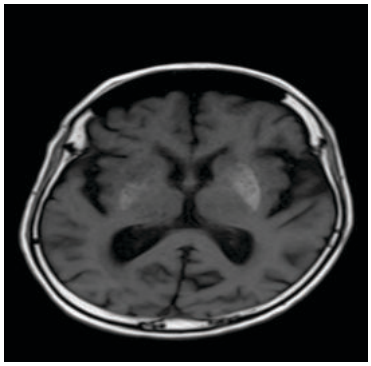
This is a case of rare and uncommom presentation of diabetes mellitus, 65-year-old female with no known comorbidities presented to emergency department with choreiform movement for 9 days. She had history of polyuria and polydipsia one month before the presentation. Her random blood sugar on presentation was 425mg/dl with HbA1c of 15.9. The patient was started on insulin and clonazepam following which her symptoms improved within 2 days with almost complete resolution on attainment of adequate glycemic control. This is the first such case to our knowledge of NKH-Chorea with hyperintense signals in basal ganglia.

INTRODUCTION

Chorea is characterized by involuntary, irregular, purposeless, random and non-rhythmic hyperkinesias[1]. The movements are abrupt , brief, jerky and unsustained. Some of the conditions associated with chorea include chorea gravidarum, systemic lupus erythematosus, antiphospholipid syndrome, hyperthyroidism, polycythemia vera and non ketotic hyperglycemia and behcet disease[2]. Hyperglycemia is the most common metabolic cause of chorea-ballism[3]. Cases of NKH-chorea have more commonly reported in elderly females of East-Asian origin[5]. Majority of patients require only glycemic control for management thus making it the key treatment in such presentations.

Case

65 year old elderly female presented to our emergency department with 9 days history of flailing movements of left upper and lower extremity associated with rapid protrusion and retraction of the tongue. She had history of polydipsia and polyuria since one month. She had no history of loss of consciousness, weakness, headache, vomiting, convulsion, blurring of vision, slurring of speech or any febrile illness. There was no history of similar illness in the family. She had no past history of diabetes mellitus, Hyperthyroidism of any other chronic illnesses. No history of neuroleptic drug intake. On examination patient was conscious, oriented with GCS of E4V5M6 , vitally stable. On auscultation she had normal vesicular breath sounds. On cardiovascular examination heart sounds were normally heard with no added sound. On examination of central nervous system, she was observed to have choreiform movements involving left upper and lower extremity with power of 5/5 in all four limbs. Tone and reflexes were normal with bilateral planter flexor response. Cranial nerve examination was normal. Cerebellar signs were absent.



T1 weighted Mri image above shows hyperintensities in bilateral lentiform nucleus

Blood investigations

Parameter	Results	Reference Range
Hemoglobin	12.1 gm/dl	12-15gm/dl
Wbc count	6500/cmm	4000-10000/cmm
Platelets	319000 /cmm	150000-450000
ALT	35 u/l	0-35 u/l
Total bilirubin	0.4mg/dl	0.3-1.2 mg/dl
Direct bilirubin	0.0	
Sodium	133.4 meq/l	135-145 meq/l
Potassium	3.44 meq/l	3.5-4.5 meq/l
Creatnine	0.79 mg/dl	0.5-1.02 mg/dl

Her random blood glucose was 425mg/dl with negative ketones and normal abga (pH-7.39, pc02-40, hco3 -24.T1-weighted (MRI) showed ill-defined hyperintense signal involving bilateral lentiform nucleus, which showed iso to hypointense signal on T2-weighted and FLAIR images with blooming on GRE.

The patient was managed with iv fluids and insulin in emergency department. In view of her symptoms and high blood sugar levels at the time of presentation, diagnosis of NKH- chorea was made and patient was managed with insulin and clonazepam during her stay in neurology ward.

DISCUSSION

Choreiform movement disorders include chorea, athetosis and ballism. Non-ketotic hyperglycemic chorea is a rare metabolic cause of chorea with a prevalence of 1/100,000[4]. It is characterized by triad of chorea, hyperglycemia and hyperintensities in basal ganglia. The mean of age of presentation for patients is 71 years and it is most commonly seen in elderly females[5].

The first ever case NKH-Chorea was reported by Bedwel in 1960[4]. Since then, a number of cases have been described around the world with similar presentation.

The pathology behind NKH-chorea remains uncertain despite a number of hypotheses that have been proposed to explain the choreiform movements in presence of hyperglycemia. One of such mechanism is hyperviscosity[6] secondary to hyperglycemia which may lead to regional blood brain barrier disruption and metabolic damage. Another mechanism is decreased availability of GABA and

acetylcholine leading to regional metabolic failure and basal ganglia dysfunction[7].

The prognosis of NKH-chorea has been reported to be excellent with complete reversibility seen in almost all cases with prompt recognition and timely treatment.[5]

Given above is the link for patient's presentation

CONCLUSION

In conclusion although uncommon chorea/ballism may be associated with Non ketotic hyperglycemia and patient with long standing diabetes may come with this presentation for the first time. Hence this uncommon and rare presentation should always be kept in mind while evaluating patients. As timely diagnosis with neuroimaging and management of hyperglycemia can completely resolve the symptoms.

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