

Non-ketotic hyperosmolar hyperglycemic chorea

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INTRODUCTION

Chorea is a type of hyperkinesia characterized by the presence of involuntary, brief and unsustained movements caused by irregular sequential muscle activation that flows continuously in a disorderly and unpredictable way.¹ It can be a manifestation of a primary neurologic genetic disorder, such as Huntington disease, or may occur as a neurologic complication of a systemic, toxic or metabolic cause, e.g., hypo/hypercalcemia or hyperglycemia.² Bedwell³, in 1960, was the first author to describe the rare clinical syndrome of nonketotic hyperosmolar hyperglycemic (NKHH) chorea. Although rare, it is a treatable condition and, therefore, should be recognized.

OBJECTIVES

To describe the clinical presentation and neuro-radiologic findings of a typical case of NKHH chorea.

CASE

An 80-year-old man was admitted after the abrupt onset of involuntary movements that affected his whole body three months before admission. Medical history revealed diabetes mellitus and systemic arterial hypertension with irregular follow-up and poor glyceemic control.

Neurological examination showed orofacial dyskinesias and gait associated choreoathetoid move-

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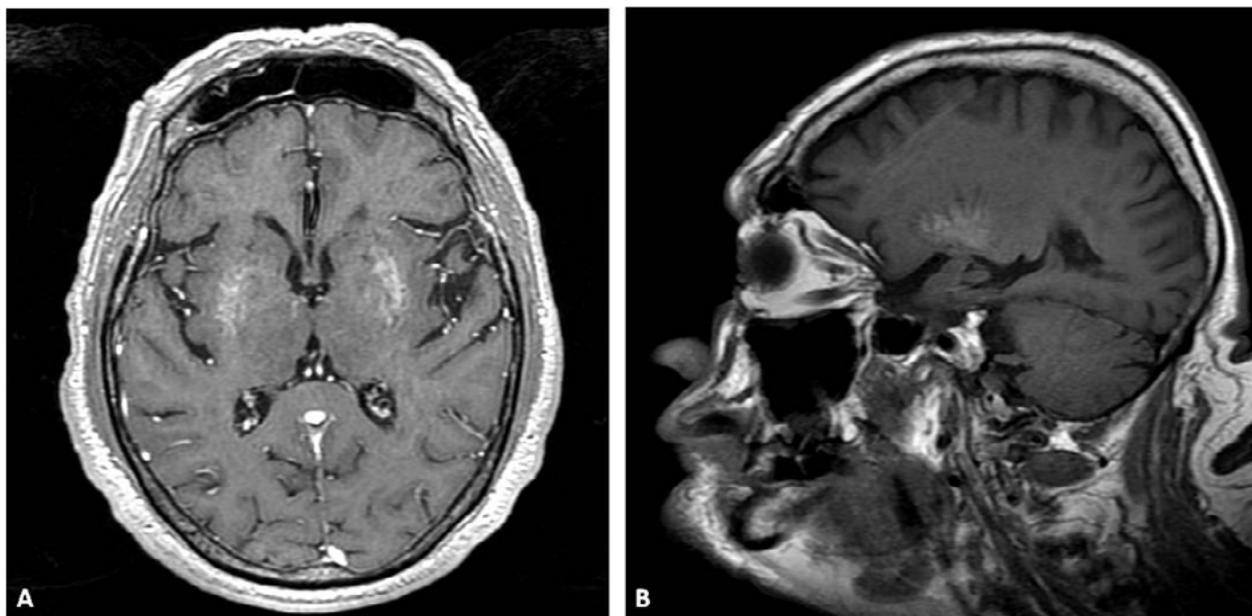


FIGURE. AXIAL (A) AND SAGITTAL (B) T1-WEIGHTED IMAGES SHOWING BILATERAL SPONTANEOUS HYPERINTENSE AREAS IN THE BASAL GANGLIA

ments affecting mainly the left lower limb. Hemoglucose was 462mg/dL on admission and brain MRI showed non-specific hyperintense areas in the basal ganglia on T1 (FIGURE).

The diagnosis of chorea secondary to nonketotic hyperosmolar hyperglycemic state was established through clinical and imaging findings, and the patient was managed with intensive diabetes control and haloperidol 5mg orally twice daily. There was a remarkable improvement in the next few days, and the patient was subsequently discharged with almost no symptoms.

DISCUSSION

Chorea pathophysiology is still not widely understood. However, unlike in parkinsonism and dystonia, intracortical inhibition of the motor cortex is normal.⁴ Semiquantitative analysis of single photon emission computed tomography in patients with hemichorea due to various causes suggests that there is an increase in activity in the contralateral thalamus, possibly due to disinhibition as a result of loss of normal pallidal inhibitory input.⁵

Non-ketotic hyperglycemia-induced chorea occurs more often in women and is usually associated with very high blood glucose⁶. The exact pathophysiology of NKHH chorea remains unclear. However,

many hypotheses as blood hyperviscosity, petechial hemorrhage, depletion of gamma-aminobutyric acid (GABA) and cerebral vascular insufficiency have been suggested.⁷ The correction of the metabolic abnormality usually is curative, but it can rarely continue for months after resolution of hyperglycemia.⁸ Striatal permanent vascular changes may mean persistence of chorea for long periods.⁹

Many of the metabolic choreas are associated with abnormalities on MRI scans. Nevertheless, the etiology of the MRI changes is not fully understood.¹⁰ Hepatocerebral degeneration and hyperglycemic chorea are often associated with high signal intensity on T1-weighted MRI involving the striatum and pallidum.⁸

Chu et al.¹¹, in a report of two patients with hyperglycemic hemichorea-hemiballism, found high signal intensities on T1- and T2-weighted images as well as on diffusion-weighted MRI accompanied by a reduction in diffusion coefficient, suggestive of hyperviscosity, rather than petechial hemorrhages, as the mechanism of edema in the striatum. This is also corroborated by another study of seven patients with hyperglycemic choreoathetosis using MRI and MR spectroscopy.¹² Interestingly, the presence of high counts of acanthocytes may predispose patients with diabetes to develop hyperglycemic chorea.¹³

CONCLUSION

A thorough physical examination and compatible clinical history and imaging are essential tools for diagnosing and treating metabolic chorea. In this case of symptoms secondary to diabetes decompensation,

glycemic control added to a central dopaminergic inhibitor were effective.

Authors Disclosures

No conflict of interest to disclose.

PALAVRAS CHAVE: *Coreia. Discinesias. Hiperglicemia.*

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