Clinical vignette: Hyperglycemic hemichorea hemiballismus: a sugar coated movement disorder

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Hyperglycemic Hemichorea Hemiballismus: A Sugar Coated Movement Disorder

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Case Presentation
An 86 year old man with type II DM, CAD, and anxiety presented to the ED with insomnia and restlessness for 3 days. He appeared dehydrated and disheveled. Physical exam was normal except for abnormal jerking, twitching and writhing of his right upper and lower extremities. There were no focal neurological deficits. HgbA1C was 18% and other than pseudo-hyponatremia (Na of 129 mmol/L) and blood glucose >500 mg/dL, there were no abnormal laboratory findings. Urine toxicology screen, TSH, and vitamin B12 levels were normal.

Patient was treated for hyperosmolar hyperglycemic state. His movements were assumed to be due to insomnia and a possible psychiatric illness but failed to resolve with sedatives. The patient underwent a brain CT scan followed by an MRI that revealed hyper-attenuation of the left putamen. This finding, with the patient’s unilateral involuntary jerking in the setting of prolonged hyperglycemia lead to the diagnosis of hyperglycemic hemichorea hemiballismus.

The patient was treated with glycemic control and short-term clonazepam. His hemichorea resolved after 3 weeks of appropriate glycemic control and clonazepam was discontinued.

Clinical Definition
- Uncontrolled jerky movements
- Unilateral or bilateral
- Prolonged hyperglycemia
- Characteristic MRI findings

The pathophysiology of this condition is poorly understood but biopsies of the basal ganglia lesions reveal swollen astrocytes. Symptoms are usually unilateral and typically resolve completely with glucose control, as do the findings on brain imaging. This striking movement disorder is diagnosed by typical MRI findings in the setting of prolonged hyperglycemia. Because it is reversible, physicians should be aware of this rare and disabling form of chorea so as not to mistake it for psychiatric disease or more serious forms of chorea such as Huntington's.

Conclusions
- Rare complication to a common disease
- Benign condition, disturbing to patient
- MRI findings are characteristic
- Glycemic control essential for cure
- Prompt diagnosis prevents unnecessary interventions

References
- Chorea associated with non-ketotic hyperglycemia and hyperintensity basal ganglia lesion on T1-weighted brain MRI study: a meta-analysis of 53 cases including four present cases: S.H. Oh, K.Y. Lee, J.H. Im, M.S. Lee; Journal of the Neurological Sciences, 200 (2002), pp. 57–62