Case 17472

Hyperglycemia induced hemichorea-hemiballismus in a 71-year old female presenting with involuntary unilateral movements

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Section: Neuroradiology
Area of Interest: CNS Neuroradiology brain
Imaging Technique: MR
Special Focus: Endocrine disorders Metabolic disorders
Case Type: Clinical Cases
Authors: Michelle Regina L. Castillo, Catherine Jessica M. Lazaro
Patient: 71 years, female

Clinical History:

A 71-year-old female with poorly controlled diabetes presented with a one-month history of involuntary, irregular, writhing movements starting with intermittent elevation of the left shoulder, followed by the ipsilateral forearm and lower extremity. Cranial nerves were intact, and no sensory and motor deficits were noted. Initial serum glucose levels were elevated.

Imaging Findings:

Computerized tomography (CT) of the brain revealed a hyperdense right basal ganglia, which likely represents mineral deposition.

Magnetic resonance imaging (MRI) of brain without Gadolinium was also done showing abnormal T1W1-hyperintense signals in the right putamen, globus pallidus, and caudate nucleus. T2W1-/FLAIR- imaging revealed slightly hyperintense signals in the right putamen, globus pallidus, and caudate nucleus. Diffusion-weighted imaging revealed areas of restricted diffusion. Susceptibility-weighted imaging showed no evidence of haemorrhage nor abnormal calcifications.

Discussion:

Background

Hyperglycemia induced hemichorea-hemiballismus is characterized by non-patterned, involuntary, unilateral movements. This presents in the elderly female population and may resolving over days or persist for years. [1]

These continuous, irregular, and involuntary jerky movements of one side of the body, is often the result of a focal lesion of the contralateral basal ganglia. Non-ketotic hyperglycemic hemichorea is a rare disorder with an incidence ranging from less than 1 in 100,000. [2]

The disease is characterized by the abundance of gemistocytes in the basal ganglia, which explains the shortened T1 relaxation time, and the cause of excessive neuronal activity, generating features of hemichorea-hemiballismus. [3]
Clinical Perspective

Patients with uncontrolled diabetes presenting with non-patterned, involuntary, irregular, jerky, unilateral movements or hemiballismus-hemichorea is rare. Other possible etiologies of hemichorea-hemiballismus include infection, medications, perinatal hypoxia, immunological disorders, and metabolic disorders such as diabetes mellitus. The presentation can mimic stroke, thus a vascular aetiology is always primarily considered.

Imaging Perspective

Computed tomography (CT) findings are usually normal, but sometimes show subtle high densities in the basal ganglia contralateral to the side of the symptoms with no associated mass effect or volume loss, and no involvement of the internal capsule. [4,5]

Magnetic resonance (MR) findings are pathognomonic for nonketotic hyperglycemic hemichorea. [1] The most commonly described MRI findings include high-intensity signals in the basal ganglia on T1-weighted brain MRI on the side contralateral to patient symptoms. [3] While the thalamus is usually spared, the putamen is almost always involved. [1,5] A shortened T1 relaxation time is also noted. The signal abnormality probably represents zinc deposition, not calcium deposition or haemorrhage. [1,3]

The hyperdense lesions within the basal ganglia found in patients with hemichorea-hemiballism was first thought to be due to calcifications or haemorrhage. However, several follow-up studies concluded that the foci in the T1-weighted images may persist for years, and did not follow the typical evolution of extravasated blood. [6]

The characteristic unilateral T1 basal ganglia hyperintensity with sparing of the thalamus, and a decreased T1 relaxation time is pathognomonic for nonketotic hyperglycemic hemichorea. The constellation of all the presenting clinical and imaging findings strongly points towards a metabolic condition such as hyperglycemia-induced hemichorea-hemiballismus.

Outcome

Secreatagogues can be given to decrease serum glucose levels. [7] Imaging findings and clinical presentation can mimic stroke. CT/MRI abnormal signal in unilateral basal ganglia, mostly in the putaminal area, should still rule out non-ketotic hyperglycemia, however rare because prompt diagnosis is key in this reversible benign condition.

Take-Home Message / Teaching Points

Hyperglycemia induced hemichorea-hemiballismus is a rather benign condition with good prognosis given that it is diagnosed early to promptly correct the hyperglycemia.

Differential Diagnosis List: Hyperglycemia induced hemichorea-hemiballismus, Vascular etiology, Infection, Medications, Immunological disorders, Metabolic disorders, including diabetes mellitus

Final Diagnosis: Hyperglycemia induced hemichorea-hemiballismus

References:


**Description:** Axial CT images at the level of the basal ganglia showed white matter hyperdensities on the right. **Origin:** Philippine General Hospital, Manila, Philippines.
Description: Axial T1-weighted images showed hyperintensities involving the right caudate nucleus, putamen, and globus pallidus. Origin: Philippine General Hospital, Manila, Philippines.
Description: Sagittal T1-weighted images showed hyperintensities at the basal ganglia.

Origin: Philippine General Hospital, Manila, Philippines
Description: Coronal T1-weighted images showed hyperintensities at the right putamen, globus pallidus, and caudate nucleus. Origin: Philippine General Hospital, Manila, Philippines.
Description: Axial FLAIR images showed hyperintense signals in the right putamen, globus pallidus, and caudate nucleus. Origin: Philippine General Hospital, Manila, Philippines.