Sugars and Shakes

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BACKGROUND

Nonketotic Hyperglycemic Hemichorea (NHH) is a rare clinical syndrome characterized by severe hyperglycemia without ketoacidosis, hemichorea/hemiballismus, and basal ganglia enhancement on MRI. The pathogenesis is related to cerebral hypoperfusion secondary to hyperglycemia, resulting in depletion of GABA neurons in the basal ganglia. NHH has high potential for misdiagnosis, leading to treatment that may be detrimental to patient safety. We present an unusual case of NHH.

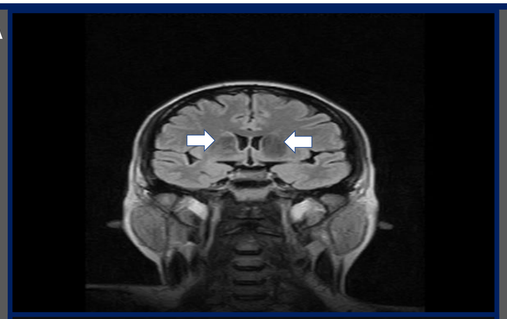
CASE PRESENTATION

A 72-year-old woman with a history of Type 2 DM treated with insulin presented after one day of upper extremity weakness. She was severely hyperglycemic (609 mg/dL) without evidence of DKA. She reported nonadherence with home insulin and her HbA1c was 14%. She underwent extensive cerebrovascular imaging with no evidence of stroke. Subsequent MRI brain demonstrated enhancement of bilateral basal ganglia. The patient reported resolution of her weakness after normalization of blood glucose. She clarified that her initial symptoms involved “wobbling” of her arm, and suspicion was raised for an atypical seizure presentation. Though EEG was negative for epileptiform activity, she was diagnosed with complex hyperglycemia-related partial seizures and discharged.

IMAGE REFERENCES

- Case courtesy of Dr Chee Kok Yoon, Radiopaedia.org, rID: 21798
- Case courtesy of Dr Franco A. Scola, Radiopaedia.org, rID: 57454
- Case courtesy of Dr Naqibullah Foladi, Radiopaedia.org, rID: 88319

A



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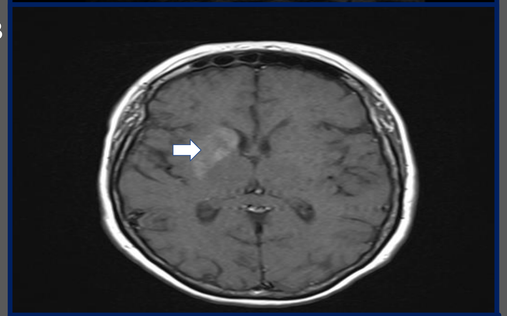

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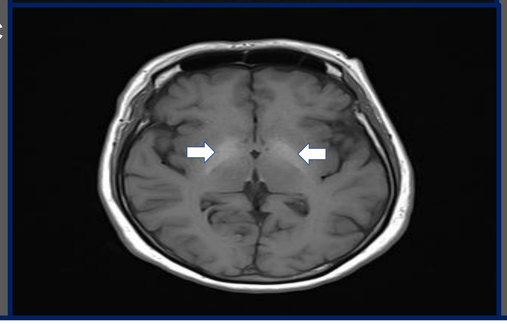

Legend

Exhibits A: T2/ Flair showing hypodensity of the caudate/putamen
Exhibits B: T1 demonstrating classical unilateral hyperdensity of the putamen
Exhibits C: T1 demonstrating bilateral hypodensity of caudate nuclei

HOSPITAL COURSE

One week later, the patient presented with similar complaints and visible choreiform movements of the left face as well as severe hyperglycemia. Given her recent diagnosis of seizures, the patient was started on levetiracetam without effect. Repeat MRI brain was negative for subthalamic stroke. Repeat EEG was negative. Chorea improved noticeably after blood sugar control was achieved. She was started on haloperidol twice daily for movement control. Further chorea workup was negative for evidence of Wilson’s disease, SLE, APLS, or Huntington’s. Diagnosis of NHH was made and the patient was discharged on haloperidol and insulin.

CONCLUSION

NHH is a rare neurological complication of uncontrolled diabetes that is important to recognize, as reversal of symptoms is achievable with strict glycemic control. Patients with NHH can be misdiagnosed with more common causes of chorea such as subthalamic stroke, Wilson’s disease, and Huntington’s disease. For our patient, failure to recognize NHH resulted in expensive workup for other neurologic syndromes and inappropriate initiation of antiepileptics. Ultimately, prompt recognition of NHH and timely initiation of insulin therapy is vital for patients with this clinical syndrome.

REFERENCES

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- Tan Y, Xin X, Xiao Q, Chen S, Cao L, Tang H. Hemiballism-hemichorea induced by ketotic hyperglycemia: case report with PET study and review of the literature. Translational Neurodegeneration. 2014;3(1):14.