



An Unusual Cause of Chorea

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Introduction

Hemichorea-hemiballism is an uncommon but recognized side effect of hyperglycemia which is frequently reversible with improved glycemic control and anti-chorea medications. The prevalence of diabetic striatopathy is likely underestimated due to lack of awareness of this condition by physicians.

Case Presentation

Month 1

Month 2

Month 3

Month 4

Months 5-18

55-year-old man with disseminated sarcoidosis and type 2 diabetes mellitus has indeterminate imaging findings concerning for neurosarcoidosis and is started on high dose steroid taper

Hemoglobin A1c 6.9%

Patient has acute onset of unilateral, involuntary choreiform movements of left upper and lower extremities

Hemoglobin A1c increased to 14.3% on metformin monotherapy, insulin regimen initiated

Hemoglobin A1c 14.3%

Admitted to hospital after MRI brain shows new T1 hyperintensities within the right putamen and minimally in the right caudate head. Multidisciplinary team agrees on diagnosis of non-ketotic hyperglycemic hemichorea

Hemoglobin A1c 10.0%

Chorea only modestly improved despite strict glycemic control. Symptoms failed to improve with trials of risperidone, haloperidol, clonazepam, and gabapentin

Hemoglobin A1c \leq 6.0%

PMH

Disseminated sarcoidosis (pulmonary hilar adenopathy with splenic involvement s/p splenectomy), stroke, hypertension, hyperlipidemia, type 2 diabetes mellitus

Objective Findings

Vitals on hospital admission:

Afebrile, BP 161/89 mmHg, HR 85bpm, RR 16/min, SpO₂ 99% on RA

Physical Exam:

Within normal limits except for choreiform movements of LUE and LLE

Labs on hospital admission:

CBC and bmp within normal limits
Vbg with normal pH of 7.45
HgbA1c 10.0%
ACE level within normal limits

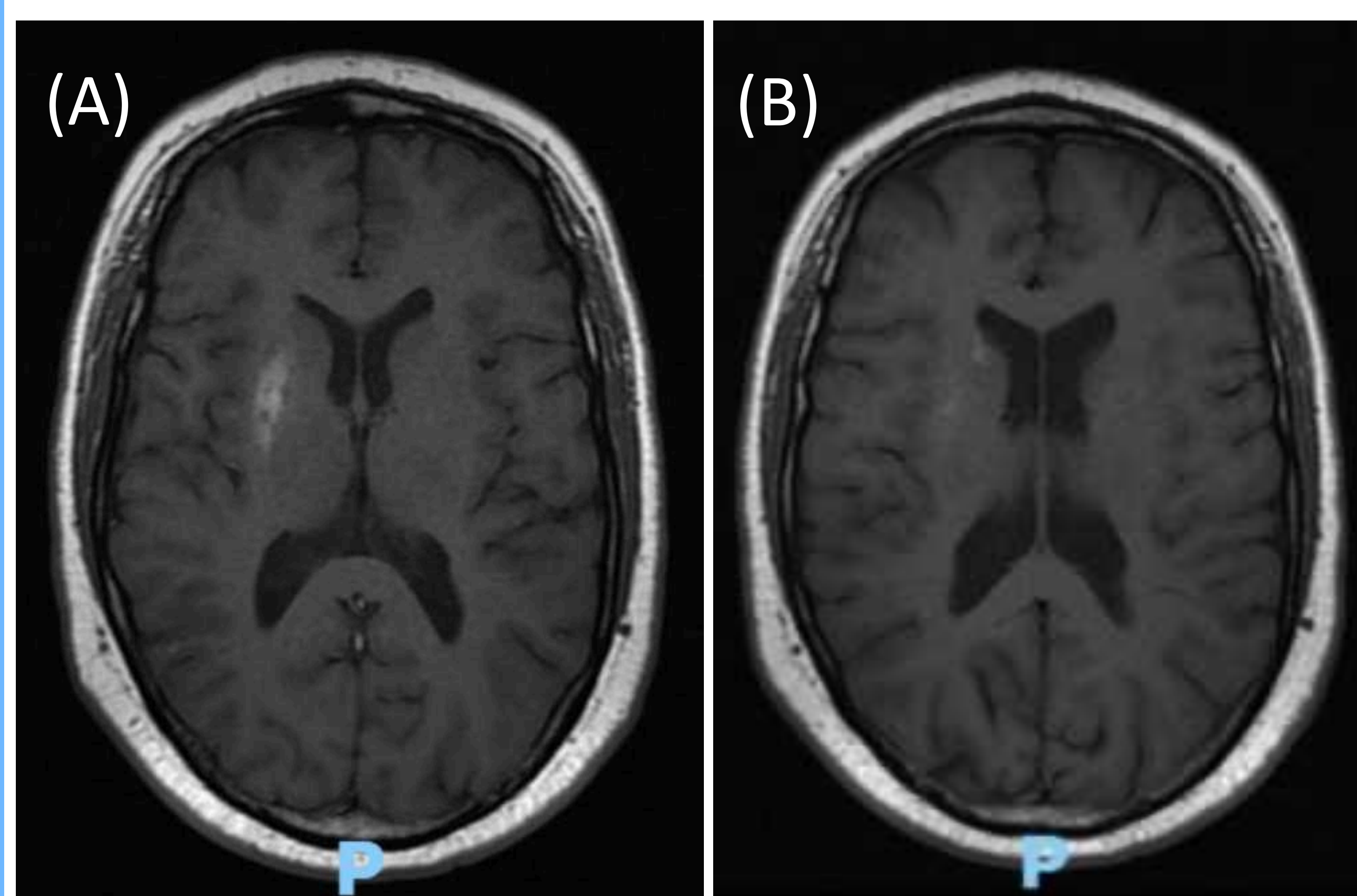


Figure 1: new T1 hyperintensities within the right putamen (A) and minimally in the right caudate head (B). T1 axial images of patient from 7/14/2020.

References

- 1) "Chua, Choon-Bing et al. "Diabetic striatopathy": clinical presentations, controversy, pathogenesis, treatments, and outcomes." Scientific reports vol. 10,1 1594. 31 Jan. 2020, doi:10.1038/s41598-020-58555-w
- 2) Nagai J, Yamada T, Cao X, Fukui A, Tajitsu M, Yamakawa F, Yambe Y, Murase T, Watanabe M, Shimada F. Cranial magnetic resonance imaging and angiography findings in a patient with hyperglycemic hemichorea-hemiballism. J Clin Endocrinol Metab. 2015 Jan;100(1):11-2. doi: 10.1210/jc.2014-2576. PMID: 25313912.

Discussion

- This case illustrates a rare and possibly underrecognized complication of hyperglycemia.
- The cause of this phenomenon is theorized to be either hyperglycemia related petechial hemorrhage or cerebral ischemia resulting in depletion of GABA and acetylcholine within the basal ganglia (2)
- Awareness of this condition is important as it is estimated 25% and 75% of patients achieve improvement in symptoms with strict glycemic control and anti-chorea medications, respectively (1)

Teaching Points

- Hyperglycemia can cause abnormal imaging findings of the basal ganglia resulting in hemichorea-hemiballism
- First line therapy includes correction of hyperglycemia and sustaining strict glycemic control
- Additional anti-chorea medications have been reported to have therapeutic benefit in many patients