Tetrabenazine in Hyperglycemic Induced Hemichorea Hemiballismus

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ABSTRACT

OBJECTIVE: To report a case of hyperglycemic induced hemichorea hemiballismus (HHHB) successfully treated with tetrabenazine (TBZ). BACKGROUND: HHHB is a rare condition manifest by acute onset hemichorea, associated with characteristic unilateral MRI hyper-intensities in the striatum. The exact pathology is not known. TBZ is a monoamine depletor that treats a variety of hyperkinetic movement disorders but has never been evaluated for HHHB. METHODS: A 74-year old woman presented with an acute onset of left hemiparesis and left hemichorea-hemiballismus shortly after the diagnosis of new-onset diabetes mellitus. She was treated with clonazepam 1 mg two times a day without benefit. The intensity was stable for one month prior to evaluation and resulted in marked functional disability. Neurological examination at presentation was normal except for the involuntary movements. Her Abnormal Involuntary Movement Scale (AIMS) was 21. CT scan and MRI imaging of the brain showed well defined hyperintensities in the right lentiform nucleus and caudate, consistent with previous reports of HHHB. RESULTS: The patient was started with TBZ one month after the onset of HHHB at 12.5 mg two days for 5 days and titrated to 25 mg three times a day. Within 2 hours of the first dose she reported marked improvement, and by three days had almost complete resolution of the movements. CONCLUSION: TBZ dramatically improved this case of HHHB. This was much more robust than when TBZ is used to treat hemichorea-hemiballismus associated with lesions of the subthalamic nucleus, suggesting a different pathophysiological mechanism.

INTRODUCTION

Hemichorea-hemiballismus (HCHB) is a continuous, involuntary, random movement involving proximal and/or distal muscles on one side of the body, including the face in some cases. [Dewey and Jankovic, 1989] It is usually associated with structural brain lesions but can occur with metabolic abnormalities. [Dewey and Jankovic, 1989] There are many reports of nonketotic hyperglycemia provoking hemichorea-hemiballismus with characteristic brain imaging including hyperintensity of the contralateral basal ganglia on brain CT scan and increased signal intensity on T1W MRI. [Hashimoto et al., 1999; Oh et al., 2002; Wintermark et al., 2004; Nakano et al., 2005] Hyperglycemic induced hemichorea-hemiballismus (HHHB) may resolve in days, or persist. [Oh et al., 2002; Wintermark et al., 2004; Li and Chang, 1994; Rergane et al., 2001] Chronic cases were reported to have slight or incomplete response to medical treatment. [Ahlskog et al., 2001] We report a persistent HHHB case who dramatically responded to TBZ.

METHODS

• A 74-year old Nigerian woman with medical histories of hypertension, hypercholesterolemia, and bradycardia developed polyuria. She has been diagnosed with new-onset diabetes mellitus.
  - One month later she developed left hand and foot “jerking” after she woke up in the morning. This quickly progressed to involve her entire left side, including face. A brain MRI done 5 days after the onset showed T1W hyperintensity, T2W hyperintensity and non-restricted DWI of the right caudate and putamen. [Figure 1A,1B,1C]
  - Repeated brain CT scan 10 days after the onset showed mild hyperintensity at the right caudate and putamen corresponding to the abnormal MR signal. [Figure 1D] She was treated with aspirin and cilostazol.
  - The initial examination at Baylor College of Medicine Movements Disorders Clinic revealed mild dysarthria and a slightly decreased left nasolabial fold.
  - The involuntary movements were not suppressible (Video Segment 1A) Her Abnormal Involuntary Movement Scale (AIMS) was 21. [Munetz and Benjamim, 1968] The volitional motor examination was otherwise mitigated by the marked hemichorea.
  - The patient was diagnosed with HHHB based on her history, examination, and classic radiographic features.
  - She was prescribed TBZ 12.5 milligrams two times a day with a slow titration to 25 milligrams three times a day. Within 2 hours of the first dose she reported marked improvement.
  - Examination revealed only very mild intermittent choreiform movement of her left foot (AIMS = 1) without any other abnormality. [Video Segment 1B]
  - Over the next three months, the left HCHB recurred shortly after stopping TBZ on two occasions.
  - She currently remains on the relatively low dose of 12.5 milligrams per day with continued excellent control.

CONCLUSIONS

TBZ dramatically improved this case of HHHB. This was much more robust than when TBZ is used to treat hemichorea-hemiballismus associated with lesions of the subthalamic nucleus, suggesting a different pathophysiological mechanism.

We report a case of an elderly woman who developed left HCHB one month after the diagnosis of a new-onset diabetes mellitus. The distance between hemichorea and hemiballism is phenomenological and likely represents a matter of severity. [Dewey and Jankovic, 1989]

TBZ dramatically improved this case of HHHB. This was much more robust than when TBZ is used to treat hemichorea-hemiballismus associated with metabolic failure from cerebral vascular insufficiency and metabolic derangement. [Hu et al., 2004], and protein desination in the course of Wallerian degeneration. [Wintermark et al., 2004]. Some have also combined this with the dystonic and choreiform movements seen in non-ketotic hyperglycemia, which resolve immediately upon glucose correction. [Jankovic, 1999; Dewey, 1999]

TBZ inhibits vesicular monoamine transporter 2 (VMAT2) which interns prevents the release of monoamine neurotransmitters and is a dopamine receptor blocker. [Jankovic and Beach, 1997] There are many reports using TBZ in hyperkinetic movement disorders including tardive dystrophy, myoclonus, Huntington’s disease, tardive dystonia, idiopathic dystonia, hemichorea-hemiballismus and hemiballism from structural lesions around the STN. [Jankovic and Beach, 1997]

HCHB is complicated by early reports that include a heterogeneous collection of causes including petechial hemorrhage [Ahlskog et al., 2001; Shan, 2008] calcification [Shan, 2005], demyelination [Lai et al., 1996], regional metabolic failure from cerebral vascular insufficiency and metabolic derangement. [Hu et al., 2004], and protein desination in the course of Wallerian degeneration. [Wintermark et al., 2004]. Some have also combined this with the dystonic and choreiform movements seen in non-ketotic hyperglycemia, which resolve immediately upon glucose correction. [Jankovic, 1999; Dewey, 1999]

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"The exact mechanism of HIHH remains speculative. This literature is complicated by early reports that include a heterogeneous collection of causes including petechial hemorrhage [Ahlskog et al., 2001; Shan, 2008] calcification [Shan, 2005], demyelination [Lai et al., 1996], regional metabolic failure from cerebral vascular insufficiency and metabolic derangement. [Hu et al., 2004], and protein desination in the course of Wallerian degeneration. [Wintermark et al., 2004]. Some have also combined this with the dystonic and choreiform movements seen in non-ketotic hyperglycemia, which resolve immediately upon glucose correction. [Jankovic, 1999; Dewey, 1999]"