

Nonketotic Hyperglycemic Chorea-Ballism (NKHCB): An Atypical Case and a Review of Literature

Syed H. Shabbir, MD,¹ and Neville M. Jadeja, MD¹

¹ Department of Neurology, Montefiore Medical Center, Bronx, NY.

Nonketotic hyperglycemic chorea-ballism (NKHCB) is a rare movement disorder characterized by hemichorea-hemiballism, contralateral striatal abnormality, and rapid resolution following glycemic control. We describe an atypical case of NKHCB in a 71-year-old female with uncontrolled type 2 diabetes exhibiting hemichorea and hemiballism limited to the right upper extremity. While NKHCB patients typically show abnormalities on com-

puted tomography (CT) imaging of the head, a CT scan of our patient during the acute phase was unremarkable. The movements subsided following glycemic control and fluid administration. The current literature on NKHCB is sparse and largely limited to case reports and series. We discuss several typical and atypical presentations and findings on imaging.

INTRODUCTION

Nonketotic hyperglycemic chorea-ballism (NKHCB) is a rare movement disorder typically associated with uncontrolled type 2 diabetes (Lai et al., 1996), though a few cases have also been reported in the setting of type 1 diabetes (Hashimoto et al., 2012). NKHCB is characterized by hemichorea-hemiballism that resolves rapidly following glycemic control. Isolated cases of more generalized, atypical, and unremitting disorders have also been reported (El Otmani et al., 2009; Massaro et al., 2012).

We describe a case of NKHCB limited to the right upper extremity that did not show any detectable abnormalities on computed tomography (CT) imaging.

CASE REPORT

A 71-year-old female with past medical history significant for poorly controlled type 2 diabetes, hypertension, hyperlipidemia, coronary artery disease, and a cerebral vascular accident without residual deficits, presented to the Jacobi Medical Center emergency department for acute onset of right upper extremity involuntary movements. The patient was in her usual state of health and was watching television at home when she noticed a fine tremor of her right upper extremity. The tremor worsened with intentional movement of the arm. Although the patient initially denied ballismic movements, she later recalled her arm having a "mind of its own." The movements kept her awake all night and prompted her to visit the hospital. She denied any numbness or weakness, but had difficulty holding objects in her right hand due to the movements. Except for a mild headache, she had no other complaints and the neurological exam was otherwise normal.

On examination, the patient was noted to have a kinetic and postural tremor punctuated by involuntary hemiballismic movements when the right arm was at rest. The patient reported being noncompliant with her insulin regimen and was found to have a serum glucose of 711 mg/dL (normal <180mg/dL) and HbA1C of 11.5% (normal <6.5%) on

testing. Her motor symptoms resolved within a couple of hours with correction of the hyperglycemia and with fluid hydration.

A head CT (Figure 1) showed mild cerebral atrophy and chronic small vessel ischemic changes. However, no evidence of a mass, mass effect, hydrocephalus, intracranial hemorrhage, acute segmental infarct, or extra-axial fluid collection was noted. Magnetic resonance imaging (MRI) was not performed due to the rapid resolution of the patient's symptoms.

The patient was discharged home on a regimen of pre-meal insulin lispro and nightly insulin glargine for long-term glycemic control.

DISCUSSION

Chorea-ballism is a poorly understood disorder with a diverse etiology and variable pathoanatomic findings (Dewey & Jankovic, 1989). The underlying pathophysiology also remains uncertain. A recent study of the acute phase and remission of 18 patients with NKHCB found strong evidence of reversible basal ganglion involvement. However, findings on T2W1 MRI were more variable and elevated Choline/Creatinine (Cho/Cr) ratios during remission suggested a more permanent neuronal change (K. Chang et al., 2010). Autopsy studies performed on patients with NKHCB have found ischemic changes of the basal ganglia, with one study also reporting calcification and mineralization (Nath, Jambhekar, Rao, & Armitano, 2006; Ohara, Nakagawa, Tabata, & Hashimoto, 2001). Positron emission tomography (PET) and single-photon emission computerized tomography (SPECT) studies similarly showed hypofunction of the striatum, suggesting a role for regional metabolic dysfunction secondary to hyperglycemia and vascular insufficiency (M. Chang, Li, Lee, & Men, 1996; Hsu, Wang, & Hsu, 2004).

In a retrospective report of 25 patients diagnosed with NKHCB at five Korean university hospitals between 1995 and 2010, six patients underwent a CT of the head, of which



Figure 1 | A head CT during the acute phase of the patient's NKHCB showed mild cerebral atrophy and chronic small vessel ischemic changes. No characteristic hyperattenuation of the basal ganglia was detected.

five showed contralateral hyperattenuation of the basal ganglia. The sixth patient had bilateral upper extremity ballismus with both CT and MRI of the head showing non-specific changes (Lee et al., 2011). Similarly, in a case series of three NKHCB patients who underwent CT imaging, two exhibited contralateral lesions of the basal ganglia, while the third patient, who had generalized chorea with no ballismic components, had no obvious abnormalities (El Otmani et al., 2009). Other patients with bilateral involuntary movements exhibit symmetric hyperattenuation of the lentiform nuclei on both CT and MRI imaging (Massaro et al., 2012).

CONCLUSION

NKHCB is a rare movement disorder that is not well understood. This case provides further evidence that clinical and radiological findings in NKHCB are variable, and adds to our understanding of NKHCB disease development and pathophysiology.

Corresponding Author: Syed H. Shabbir, MD
(syedhshabbir@gmail.com).

Author Contributions: SHS and NMJ researched data and prepared the manuscript.

Disclosure: The authors have completed and submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. No conflicts were noted.

References

- Chang, K., Tsou, J., Chen, S., Ro, L., Lyu, R., Chang, H., ... Chen, C. (2010). Temporal features of magnetic resonance imaging and spectroscopy in Nonketotic hyperglycemic chorea-ballism patients. *European Journal of Neurology: The Official Journal of the European Federation of Neurological Societies*, 17(4), 589–593.
- Chang, M., Li, J., Lee, S., & Men, C. (1996). Nonketotic hyperglycaemic chorea: A SPECT study. *Journal of Neurology, Neurosurgery, and Psychiatry*, 60(4), 428–430.
- Dewey, R. B., Jr, & Jankovic, J. (1989). Hemiballism-hemichorea. Clinical and pharmacologic findings in 21 patients. *Archives of Neurology*, 46(8), 862–867.
- El Otmani, H., Moutaouakil, F., Fadel, H., & El Ouafi, N. (2009). Chorea-ballismus in acute Nonketotic hyperglycaemia. *Functional Neurology*, 24(3), 129–132.
- Hashimoto, K., Ito, Y., Tanahashi, H., Hayashi, M., Yamakita, N., & Yasuda, K. (2012). Hyperglycemic chorea-ballism or acute exacerbation of Huntington's chorea? Huntington's disease unmasked by diabetic ketoacidosis in type 1 diabetes mellitus. *The Journal of Clinical Endocrinology and Metabolism*, 97(9), 3016–3020.
- Hsu, J. L., Wang, H.-C., & Hsu, W.-C. (2004). Hyperglycemia-induced unilateral basal ganglion lesions with and without hemichorea. A PET study. *Journal of Neurology*, 251(12), 1486–1490.
- Lai, P. H., Tien, R. D., Chang, M. H., Teng, M. M., Yang, C. F., Pan, H. B., ... Kong, K. W. (1996). Chorea-ballismus with nonketotic hyperglycemia in primary diabetes mellitus. *AJNR. American Journal of Neuroradiology*, 17(6), 1057–1064.
- Lee, S.-H., Shin, J.-A., Kim, J.-H., Son, J.-W., Lee, K.-W., Ko, S.-H., ... Ahn, Y.-B. (2011). Chorea-ballism associated with nonketotic hyperglycaemia or diabetic ketoacidosis: characteristics of 25 patients in Korea. *Diabetes Research and Clinical Practice*, 93(2), e80–83.
- Massaro, F., Palumbo, P., Falcini, M., Zanfranceschi, G., & Pratesi, A. (2012). Generalized chorea-ballism in acute non ketotic hyperglycemia: Findings from diffusion-weighted magnetic resonance imaging. *Parkinsonism & Related Disorders*, 18(8), 998–999.
- Nath, J., Jambhekar, K., Rao, C., & Armitano, E. (2006). Radiological and pathological changes in hemiballism-hemichorea with striatal hyperintensity. *Journal of Magnetic Resonance Imaging: JMIR*, 23(4), 564–568.
- Ohara, S., Nakagawa, S., Tabata, K., & Hashimoto, T. (2001). Hemiballism with hyperglycemia and striatal T1-MRI hyperintensity: An autopsy report. *Movement Disorders: Official Journal of the Movement Disorder Society*, 16(3), 521–525.