

Case Report

Persistent Hemichorea and Caudate Atrophy in Untreated Diabetic Striatopathy: A Case Report

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Keywords

Hemichorea · Caudate atrophy · Diabetic striatopathy · Magnetic resonance imaging

Abstract

Background: Neurological complications of diabetes and hyperglycemia are relatively common but the specific manifestations can vary widely. Diabetic striatal disease or “diabetic striatopathy” is an uncommon condition usually thought to result from hyperglycemic injury to the basal ganglia, producing a hyperkinetic movement disorder, usually choreiform in nature. Symptoms are generally reversible with treatment of the hyperglycemia. **Case Description:** We report the case of a 57-year-old woman presenting with a unilateral choreo-athetosis of the left upper extremity, persistent for 4 years. Contemporaneous imaging demonstrated severe atrophy of the right caudate nucleus, while imaging obtained at the onset of symptoms was consistent with a right diabetic striatopathy. Symptoms improved with the use of dopamine antagonists and benzodiazepines. **Conclusion:** Although generally considered to be fully reversible, this case demonstrates that diabetic striatopathy can result in permanent structural lesions with persistent symptoms if left untreated.

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Introduction

Diabetic striatopathy is a rare disorder that classically presents with a limited period of choreiform or ballistic movements associated with an episode of nonketotic hyperglycemia or diabetic ketoacidosis along with characteristic MRI findings. The movement disorder and imaging abnormalities typically resolve with treatment of the hyperglycemia or shortly thereafter [1, 2], although in some cases the movements may become permanent for unclear reasons [3]. To our knowledge, there has not been a reported case in which untreated diabetic striatopathy eventually resulted in structural brain changes along with a permanent movement disorder, which we describe here.

Case Presentation

A 57-year-old right-handed woman with poorly controlled type 2 diabetes mellitus and mild peripheral neuropathy presented to our clinic for involuntary movements of her left hand. Five years previously she experienced the acute onset of these movements with gradually increasing intensity over about 1 month. The movements were essentially stable for the 4 years prior to her presentation and interfered significantly with her work as a hairdresser.

She noted no abnormal movements elsewhere in the body and had no neuropsychiatric issues. The movements would cease during sleep and worsen with stress. She could inhibit the movements with concentrated effort but there was no urge, rebound, or sense of relief when the movement was allowed to occur. Trials of trihexyphenidyl, ropinirole, and carbidopa-levodopa had no effect. Clonazepam reduced the movements somewhat.

She had no history of neuroleptic or antiemetic exposure. Her only medications were insulin and lisinopril. She was poorly compliant with the insulin, but had no history of hyperglycemic hyperosmolar nonketotic coma or diabetic ketoacidosis. She specifically denied hospitalization related to her diabetes or her hypertension. She did not measure her blood pressures at home, but in her 7 visits to our facilities her systolic blood pressure ranged between 152 and 160 mm Hg, suggesting poorly controlled hypertension as well. She reportedly had a history of hyperlipidemia. Family history was unremarkable, and was specifically negative for any movement disorders. Social history was positive for an approximately 10 pack-year history of tobacco use in the past but she had been tobacco-free for 15 years at the time of presentation.

General neurological examination revealed a normal mental status and cranial nerves. On motor examination, there was nearly continuous low-amplitude choreoathetosis of the left wrist and fingers, without evidence of bradykinesia or rigidity (See online suppl. video S1; for all online suppl. material, see www.karger.com/doi/10.1159/000484201). Extraocular movements were entirely normal. There were no abnormal movements of the mouth or tongue. There was no myoclonus or dystonia. She had slightly reduced DTRs and vibratory sensation, consistent with her known peripheral neuropathy. Gait was normal. Prior workup included an unremarkable EEG and infectious, autoimmune, metabolic, and paraneoplastic labs, only notable for HbA1c of 14%. An MRI of the brain had been obtained within a month of symptom onset (Fig. 1a) and was read at an outside facility as having “basal ganglia calcification.” This image demonstrates nonenhancing hyperintensity on T1 and hypointensity on T2 (FLAIR) and gradient-echo T2*-weighted images of the right putamen and caudate nucleus, with dramatic sparing of the internal capsule consistent with diabetic striatopathy.

Follow-up imaging at presentation to our clinic 5 years after symptom onset showed resolution of the T1 hyperintensity but severe atrophy of the right caudate nucleus (Fig. 1b).

Quetiapine was not tolerated due to sedation. Movements improved significantly but did not entirely resolve with the addition of 10 mg olanzapine to 1.5 mg clonazepam daily. Unfortunately, she was lost to follow-up before trials of risperidone, haloperidol, or tetrabenazine could be undertaken.

Discussion

The precise pathophysiology of diabetic striatopathy is not well understood but biopsy studies suggest the lesion is a vasculopathy with gliosis restricted to the striatum [1]. Post-mortem findings have been somewhat inconsistent but include reactive astrocytosis, patchy ischemic necrosis, petechial hemorrhage, vascular proliferation, and arteriolar changes somewhat akin to the pathology of diabetic proliferative retinopathy [1, 4, 5]. The mechanism by which these changes produce chorea is unknown, but a small-scale SPECT study implicates a relative vascular perfusion defect in the striatum as seen in Huntington's disease [6].

The exact nature of the characteristic reversible appearance on T1-weighted MRI is also uncertain. Methemoglobin, lipid, protein, and inorganic minerals can all produce T1 shortening, but given the immediate appearance and reversibility of the signal it is unlikely to represent mineralization or blood products.

Most reported cases that include follow-up imaging suggest that the associated T1 hyperintensity resolves along with the movement symptoms as hyperglycemia is corrected (although radiographic resolution was found to lag clinical improvement) [2, 7]. In a large case series, 16 of 52 patients who were treated only by controlling the hyperglycemia had complete resolution of their chorea, and the remainder generally improved with standard treatments targeting hyperkinetic movement [7]. Other reported cases in which the chorea became permanent were characterized by an antecedent hyperglycemic crisis requiring hospitalization [3], which our patient did not have. Our patient is a non-Asian female with type 2 diabetes. Some studies suggest that diabetic striatopathy is more prevalent in Asian and female patients with type 2 diabetes than in other populations for unclear reasons [1, 7].

This case is unusual in that the patient's hyperglycemia was never intensively treated and appears to have persisted at a high level. While the characteristic resolution of the T1 MRI changes was observed in our patient, sufficient structural damage apparently occurred such that near-complete atrophy of the caudate was observed on follow-up imaging 5 years after onset. This observation supports the concept of diabetic striatopathy as a structurally destructive lesion whose pathological driver is hyperglycemia.

Conclusion

To our knowledge, this is the first report of persistent hemichorea and caudate atrophy in the setting of untreated diabetic striatopathy. Overall, chorea-hyperkinesia caused by hyperglycemia is treatable and carries a good prognosis. As this case illustrates, however, it may lead to permanent abnormal movements and structural brain changes if the hyperglycemia is left uncorrected. This finding suggests that the pathology of diabetic striatopathy, whether vasculopathic or otherwise, represents an ongoing process that requires active

intervention to achieve a favorable outcome. The increasing prevalence of diabetes worldwide makes awareness of even relatively rare complications important for clinicians in multiple disciplines.

Statement of Ethics

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Disclosure Statement

Funding Sources and Conflict of Interest

No specific funding was received for this work. The authors declare that there are no conflicts of interest relevant to this work.

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Author Contributions

E.B.L.: manuscript conception, writing of the first draft, review, critique. W.T.D.: interpretation of radiological imaging, manuscript review, critique. M.C.S.: manuscript conception, writing of the first draft, review, critique, and writing of the final draft.

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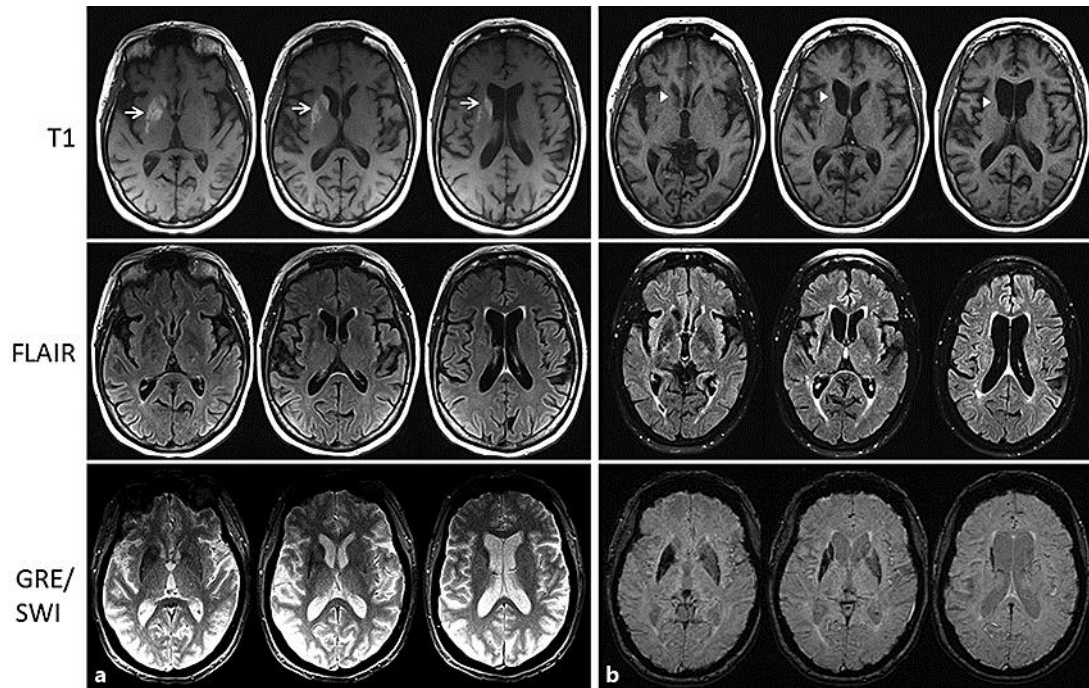


Fig. 1. MRI images at symptom onset (**a**) and at 5 years (**b**). Initial images show unenhanced T1 hyperintensity restricted to the right striatum (arrows). The high T1 signal has resolved in follow-up imaging, but the caudate head has atrophied severely (arrowheads).