Review Article

GMSMJ Greater Mekong Subregion Medical Journal

Diabetic Striatopathy: A Rare Microvascular Neurological Diabetic Complication

Kaset Chimplee, M.D.[1]

[1]Department of Internal Medicine, School of Medicine, Mae Fah Luang University, Chiang Rai 57100, Thailand

Received 25 May 2021 • Revised 27 June 2021 • Accepted 30 July 2021 • Published online 1 September 2021

Abstract:

Diabetic striatopathy is a rare neurological diabetic complication which has an incidence of 1:100,000 population. This report presents a female with poorly controlled type 2 diabetes and hypertension, who presented with right sided choreoathetosis. No other neurological abnormalities were found on physical examination. Brain imaging showed hyperdensity of the left caudate and left lentiform nuclei on non-contrast CT scan. The patient made a complete recovery from the abnormal movements following tight glucose control and introduction of trihexyphenidyl and haloperidol. A follow up CT scan 6 months following presentation revealed resolution of the previous striatal abnormalities.

Keywords: Diabetic striatopathy, Choreoathetosis, Hemichorea/hemiballism, Corpus striatum

Introduction

Type 2 diabetic patients can present with abnormal movement called choreoathetosis or hemichorea/hemiballism. This movement disorder is known to develop in association with stroke, Wilson's disease, neoplasm, infection and thyrotoxicosis[1]. However, this choreoathetosis was possibly directly related to complications of diabetes. As already known, one of the microvascular neurological complications of diabetes is diabetic neuropathy, in which patients present with glove and stocking numbness or painful sensations affecting their limbs. There is also another microvascular neurological diabetic complication called "Diabetic Striatopathy". This case report presents one case of poorly controlled type 2 diabetes, that presented with abnormal movements and abnormal brain imaging.

Case presentation

A 76-year-old Thai woman with history of type 2 diabetes and hypertension presented with fatigue, dizziness and vomiting. Her regular medications had consisted of glipizide and metformin for her diabetes and amlodipine and enalapril for high blood pressure. She had never received insulin therapy for glucose control. For unknown reasons she had stopped her oral hypoglycemic agents and all of her other medications for 3 months, prior to development of the presenting symptoms. On physical examination, she had normal mental status without any focal neurological deficit. Biochemical investigations revealed random blood glucose 408 mg/dL, creatinine 1.75 mg/dL (estimated GFR 28 mL/min), Hb 12.2 g/dL, Hct 34.3%, WBC 11.7×10^9/L. Non-contrast CT scan of the brain showed

Corresponding author: Kaset Chimplee, M.D.
School of Medicine, Mae Fah Luang University, Chiang Rai 57100, Thailand
E-mail: kaset.chi@mfu.ac.th

GMSMJ 2021; 1 (3): 151-156

relative hyperdensity at the left caudate nucleus and left lentiform nucleus without surrounding brain edema. Diffuse brain atrophy was also seen (Figure 1A-C). Because of the very high glucose level, the patient received insulin treatment. Her blood glucose at 4 weeks follow up had declined to 120 mg/dL. However, 2 weeks later, 6 weeks following her first appointment, she came to the hospital with pain and numbness of her left arm with uncontrolled movements of her right shoulder and right arm. These abnormal movements had persisted all day and night for 3 days. She did not have muscle weakness or facial weakness. Furthermore, her speech was normal, and she did not have headache, dizziness or vomiting. On physical examination her blood pressure was 175/82 mmHg, she had normal mental status and normal cranial nerves examination. Obvious continuous low amplitude choreoathetoid movements of the right shoulder, arm and wrist were noted. There was normal muscle power with no sensory deficit. No evidence of muscle rigidity nor bradykinesia was observed. Non-contrast CT brain revealed obviously increased hyperdensity of both the left caudate nucleus and left lentiform nucleus. without peripheral brain swelling, compared with the previous imaging. The CT findings were compatible with non-ketotic hyperglycemic hemichorea (diabetic striatopathy), (Figure 2A-C). She was commenced on trihexyphenidyl for the abnormal movements with no therapeutic effect so haloperidol was added in 2 weeks later. Her blood glucose at follow-up visit was 124 mg/dL, HbA1c 7.3%. Medical treatment for diabetes included Insulin (NPH) and glipizide. Fourteen weeks after treatment, the choreoathetosis disappeared and her glycemic profile remained within the normal range. (fasting plasma glucose 101 mg/dL and HbA1c 6.8%). Follow up CT scan done 6 months later showed resolution of the previous hyperdensities of the left caudate and lentiform nuclei (Figure 3A-C).

Discussion

"Diabetic striatopathy" was also known as hyperglycemic non-ketotic hemichorea/hemiballism or diabetic hemichorea/hemiballism or chorea hyperglycemia basal ganglia syndrome. Hemichorea and hemiballism are defined as random involuntary continuous jerking movements, involving one side of the body in which chorea, characteristically being described as more distal and of less amplitude than ballism[1]. Therefore, diabetic striatopathy is described as a hyperglycemic state accompanied by one of these conditions 1) chorea/ballism 2) striatal hyperdensity, as seen on non-contrast CT scan and as hyperintensity on a T1 weight MRI brain scan[2].

The incidence of diabetic striatopathy is recorded as 1:100,000 population incidence and is composed of Asian (71.6%), European (8.5%), American (4%). Mean age was 67.6 years old, with a female to male ratio of 1.7:1[2]. Mean age of onset was 70 and 96 percent of cases were found in DM type 2 but only 3.4 percent was found in DM type 1. Fifty-four percent were associated with poorly controlled diabetes and 55 percent had high blood pressure[3].

Hemichorea/hemiballism can involve symptoms in the face and trunk, in addition to the arm and leg, and showed bilateral involvement in less than 10 percent of cases.[2] Patients usually had hyperglycemia at presentation (random blood glucose 306-414 mg/dL, HbA1c 13.1-14.5%)[2-5]. As found in our case, the patient also had high blood glucose, 408 mg/dL, when striatal abnormality was detected on the CT scan. However, choreoathetosis was diagnosed 6 weeks later when the CT scan showed more obvious hyperdensity of both the left caudate nucleus and left lentiform nucleus.

On imaging of diabetic striatopathy, non-enhanced CT scan shows striatal (caudate nucleus and lentiform nucleus) hyperdensity and MRI scan reveals striatal (caudate nucleus, putamen and globus pallidus) hyperintensity on T1 weight with hypointensity on T2 weight[4]. Nevertheless, there is discrepancy between clinical hemichorea/hemiballism and striatal involvement as seen on CT and MRI. Furthermore, discrepancies, between CT and MRI findings, are related to differences in the location of the striatal abnormalities. Sensitivity of CT scan to detect striatal abnormality in diabetic striatopathy was 78.5 percent compared with 95.5 percent when using MRI, but CT scan can be useful to detect abnormalities on negative scans using MRI[4]. Using CT scan, the locations of striatal abnormality, 78.6% were found in putamen, 47.6% in the caudate nucleus and 27.8% in the globus pallidus. After treatment of hyperglycemia, complete or partial resolution of striatal abnormalities were seen on MRI and CT scan[2].

The median time of resolution of striatal hyperintensity seen on MRI was 180 days and was 60 days on CT scan[2]. However, abnormalities on imaging can persist, as in a case of untreated diabetic striatopathy, reported by Lucassen[4], in which symptoms persisted for 4 years, where MRI showed severe atrophy of the caudate nucleus. Homaida[6] has suggested that differential diagnosis of striatal hyperintensity, seen on CT scan or MRI, could be 1) petechial hemorrhage 2) mineral deposition 3) myelin destruction 4) infarction with astrocytosis.

The mechanism of pathogenesis of diabetic striatopathy could be formulated as shown in figure 4.

Abe[7] performed a needle biopsy of the corpus striatum in one case of diabetic striatopathy, a 50-year-old Japanese man found to have low intensity areas in the right putamen and caudate nuclei on CT scan. Histopathology of his striatal specimen is shown in table 1.

In summary histopathology of diabetic striatopathy, can be characterized as striatal abnormalities due to 1) occlusive vasculopathy 2) patchy necrosis 3) prominent neovascularization. Treatment of diabetic striatopathy consists mainly of correction of hyperglycemia which results in partial or complete resolution of clinical symptoms of hemichorea/hemiballism and reduction of striatal abnormalities on neuroimaging. Anti-chorea medication includes haloperidol, tetrabenazine, risperidone, clonazepam, used as single medications or in combination. With high efficacy, haloperidol has become the most frequently used medication[1,4,5,6,8,9]. The median recovery time after commencement of anti-chorea medication was 14 days[3].

Conclusion

Diabetic striatopathy is a rare microvascular neurological diabetic complication which can be seen in poorly controlled diabetic patients, especially in the elderly Asian females. Such patients commonly present with hemichorea/hemiballism. Diagnosis is composed of the triad of 1) unilateral involuntary movements and 2) contralateral striatal abnormality on imaging and 3) hyperglycemia. Pathology of diabetic striatopathy is microangiopathy that is confined to the corpus striatum. Patients have a good prognosis with complete clinical remission after successful treatment of hyperglycemia.

Acknowledgement

The author would like to thank to Roger Timothy Callaghan MB, ChB. a family physician and lecturer, for his suggestion on English writing following careful reading of the manuscript and Kwinnart Wongsirodkul, M.D. a radiologist

and lecturer for her suggestion of appropriate CT scan cuts. The author also has special thanks to the patient for permission to publish her clinical details presented in this report.

References

1. Özgür A, Esen K, Kaleağası H, Yılmaz A, Kara E. Diabetic striatopathy in a patient with hemiballism. Emergency radiology 2015; 22: 347-9.
2. Chua CB, Sun CK, Hsu CW, Tai YC, Liang CY, Tsai IT. "Diabetic striatopathy": clinical presentations, controversy, pathogenesis, treatments, and outcomes. Scientific reports 2020; 10: 1-11.
3. Cosentino C, Torres L, Nuñez Y, Suarez R, Velez M, Flores M. Hemichorea/hemiballism associated with hyperglycemia: report of 20 cases. Tremor and other hyperkinetic movements 2016; 6: 402-5.
4. Lucassen EB, Delfyett WT, Stahl MC. Persistent hemichorea and caudate atrophy in untreated diabetic striatopathy: A case report. Case reports in neurology 2017; 9: 299-303.
5. Lupescu IC, Lupescu IG, Arbune A, Toron B, Dulamea AO. A few thoughts on diabetic striatopathy-case report and short review. Romanian Journal of Neurology 2020; 19: 41-5.
6. Homaida M, Kanodia AK, Young N, Yu WM. Diabetic striatopathy: a rare condition and diagnostic dilemma. BMJ Case Reports 2021;14: DOI:10.1136/bcr-2020-240141
7. Abe Y, Yamamoto T, Soeda T, Kumagai T, Tanno Y, Kubo J, et al. (2009). Diabetic striatal disease: clinical presentation, neuroimaging, and pathology. Internal Medicine 2009; 48:1135-41.
8. Chutpiboonwat P, Chutinet A, Tontiwutikul B, Snabboon T. (2020). Diabetic Striatopathy. Acta Médica Portuguesa 2020; 33:13.
9. Tocco P, Barbieri F, Bonetti B, Barillari M, Marangi A, Tinazzi M. Hemichorea-hemiballismus in patients with non-ketotic hyperglycemia. Neurol Sci 2016; 37: 297-8.

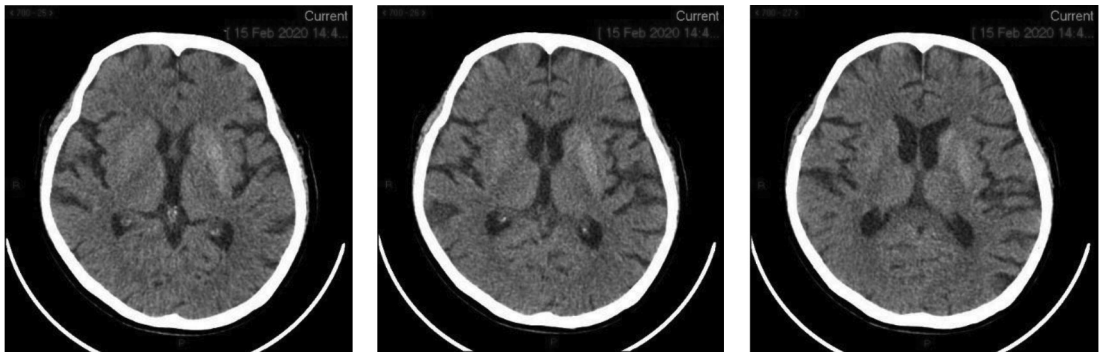

Figure 1 A-C Non-contrast CT scan at first presentation

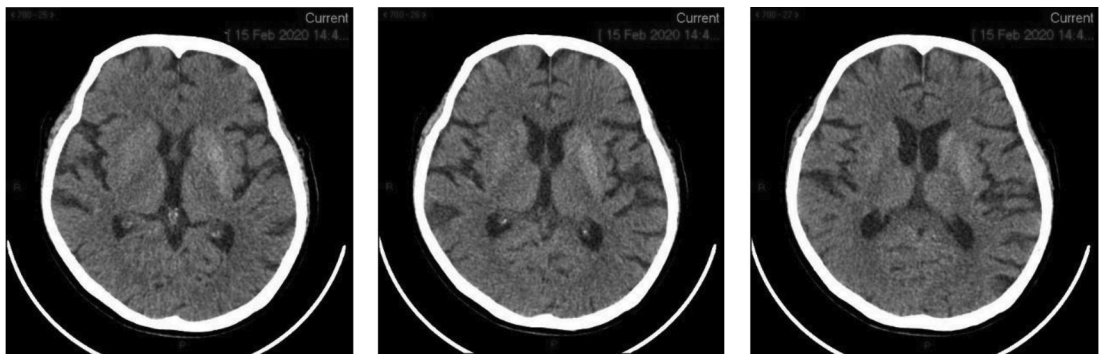

Figure 2 A-C Non-contrast CT scan at presentation with choreoathetosis, 6 weeks from the first presentation

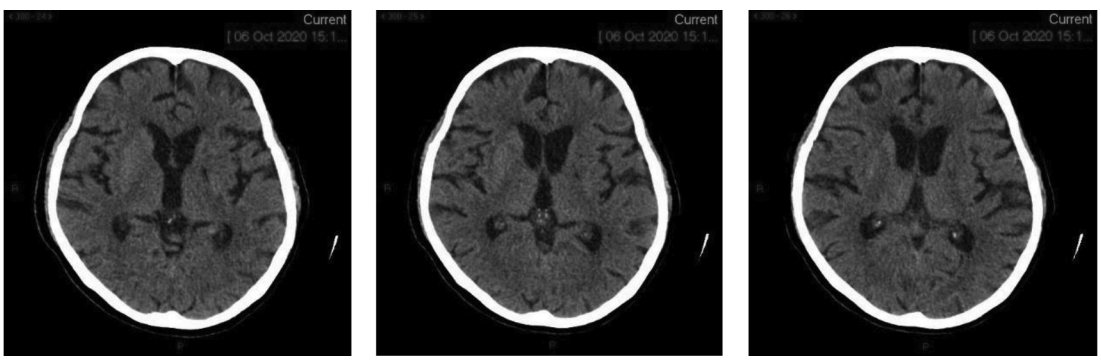

Figure 3 A-C Non-contrast CT scan at follow up period, 6 months after treatment

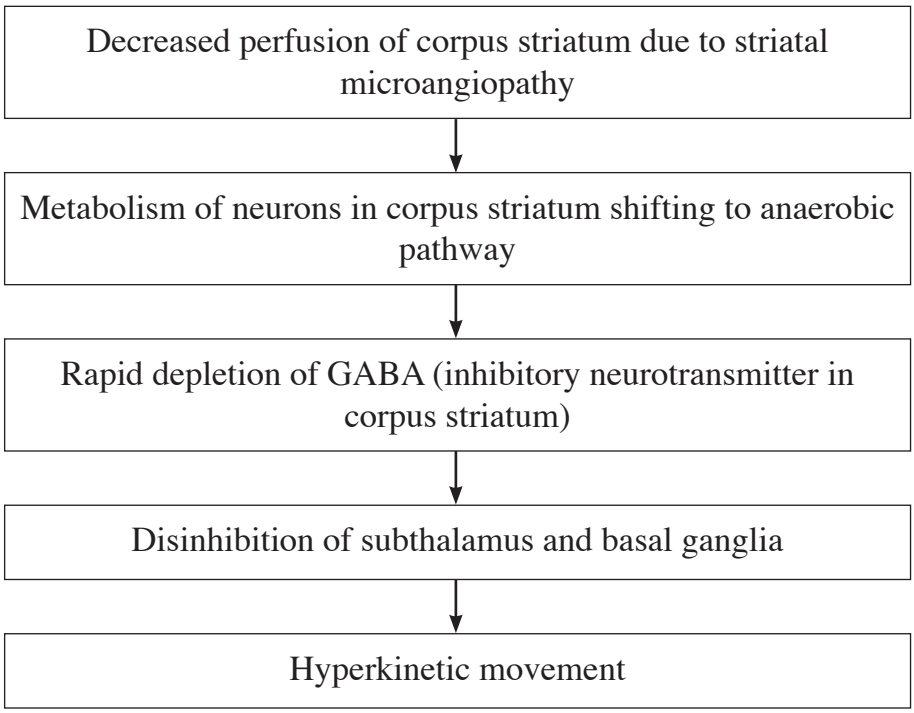

Figure 4 Hypothesized mechanisms in pathogenesis of diabetic striatopathy[1,2]

Table 1 Histopathology of corpus striatum in diabetic patient with striatal abnormalities on CT scan[7]

- Marked thickening media with hyaline changes of vessel wall
- Obliteration of arteriolar lumen
- Focal cell infiltration, red blood cell extravasation
- Lymphocyte infiltration and macrophage invasion
- Capillary proliferation and neovascular formation