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Case Report

Diabetic striatopathy unusual presentation with ischemic stroke—A case report and literature review ☆

Omar Y. Alkhaja, MD, Ali Alsetrawi, MD, Tareq AlTaei, MD, Mohammed Taleb, MD*

Department of Radiology, Salmaniya Medical Complex, Rd No 2904, Manama, Bahrain

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ABSTRACT

Diabetic striatopathy (DS) is a rare condition present as a combination of hemichorea or hemiballismus with striatal computed tomography (CT) or magnetic resonance imaging (MRI) changes in the presence of a severely hyperglycemic state. Varying severity and manifestations of DS were reported throughout the literature. However, the exact pathogenesis and mechanism remain unclear. In this case report, we are presenting an unusual case of DS combined with acute ischemic stroke. A 74-year-old male presented with the main complaint of acute left arm and leg weakness, and was found to have significantly high blood sugar and evidence of DS in combination with stroke on CT and MRI. Eventually, he was labeled as a case of combined DS and ischemic stroke.

Introduction

Diabetic striatopathy (DS) is a hyperglycemic state that manifests as hemichorea or hemiballismus, in addition to the presence of striatal hyperdensity changes on computed tomography (CT) or high signal changes on T1-weighted magnetic resonance imaging (MRI) contralateral to the symptoms side [1,2]. DS was known as hyperglycemic non-ketotic hemichorea and hemiballism, and Bedwell first identified it in the 1960s [3]. It is a rare condition, with less than 200 cases reported until 2021 [4]. The prevalence of DS was estimated in the literature as 1 in every 100,000 [5].

In this case report, we are presenting a patient who came without the classic symptoms of DS. Instead, his main complaint was stroke-like symptoms of ipsilateral limb weakness without any movement disorder. However, with further workup, classical CT and MRI imaging features of DS were detected in association with multiple areas of acute infarction seen in the DWI images. We also present significant volume loss and gliosis of the affected caudate after the resolution of the basal ganglia hyperdensity on CT scan follow-up.

Abbreviations: DS, diabetic striatopathy; CT, computed tomography; MRI, magnetic resonance imaging.

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* Corresponding author.
E-mail address: Omar.alkhajah@gmail.com (O.Y. Alkhaja).

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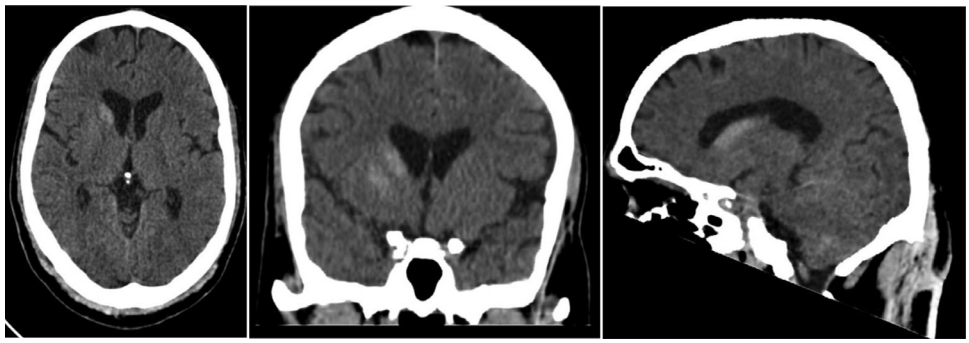

Fig. 1 – Axial, Coronal, and sagittal cuts of plain brain CT scan: demonstrating right basal ganglia hyperdensity.

Case report

In this report, we present a 74-year-old Caucasian Bahraini male, who presented to the emergency department with the main complaint of acute left arm and leg weakness. He is a known case of type 2 diabetes mellitus, hypertension, and coronary artery disease with a coronary artery stent one month before presentation. His main complaint was acute ipsilateral left arm and leg weakness, along with slurred speech for almost one day before his presentation to the emergency department at Salmanya Medical Complex in Bahrain. There was no history of headache, visual disturbance, loss of consciousness, or previous similar episodes. The patient denied any prior history of chorea or abnormal limb movement. He was on insulin glargine, insulin aspart, hydrochlorothiazide, acetylsalicylic acid, rabeprazole, clopidogrel, ezetimibe, trimetazidine, bisoprolol fumarate, and atorvastatin. The patient stated that he was not fully compliant with his medications.

Upon arrival at the emergency department, the patient was afebrile, his blood pressure was 100/60 mmHg, his heart rate of 75 beats per minute, and he had normal oxygen saturation on room air. Further, physical examination revealed slurred speech, however, intact comprehension and orientation. He had 3/5 motor power in the left upper and lower limbs. However, intact sensation to soft and sharp and pain stimuli. His right upper and lower limbs were normal. Cranial nerves examination was normal apart from hypoglossal cranial nerve function weakness evidenced by weak tongue motor function—otherwise, no other significant abnormalities.

Laboratory examination revealed highly elevated blood sugar, with negative ketones in the urine. His random blood glucose level was 18.3 mmol/L, and his Hemoglobin A1C was 109 mmol/mol. Other laboratory investigations were as follows, LDL cholesterol of 5.15 mmol/L, triglycerides of 3.1 mmol/L, sodium blood level was 140 mmol/L, potassium blood level was 4.3 mmol/L, calcium level of 2.25 mmol/L and chloride level of 106 mmol/L. Moreover, syphilis blood tests and tuberculosis tests were done and were all negative. In addition, an emergency CT scan showed mainly unilateral hyperdense right caudate and lentiform nucleus, otherwise, no other significant abnormality was noted (Fig. 1).

Later on, MRI was ordered and revealed multiple areas of acute lacunar infarction in the DWI and ADC images involving the right MCA cortical branches, the deep white matter of the centrum semiovale and corona radiata at the deep border zone (between deep white matter branches of the MCA and the ACA). Additionally, subacute lacunar infarction was noted in the left medial temporal lobe and occipital lobe (Fig. 2). Also, a re-demonstration of the previously seen changes in the CT scan of the right caudate nucleus and basal ganglia in the form of low T2 signal intensity and high signal intensity in T1 changes at the region (Fig. 3). No definite evidence of restricted diffusion or blooming artifacts was seen in gradient and SWI images to suggest hemorrhage. In addition, an MR angiogram was also done and showed multifocal narrowing of bilateral MCA - M1 and both PCAs (Fig. 4).

Accordingly, in keeping with the overall scenario of this patient and his imaging findings, he was admitted under the impression of DS in association with an acute cerebrovascular accident. Blood sugar correction, anticoagulation therapy, and further management interventions were initiated. The patient was discharged after 2 weeks of admission in a better overall condition, and his slurred speech improved; however, his left limbs were not fully recovered. On further follow-up, the patient's clinical condition partially improved since discharge, with only slight left lower limp weakness remaining. However, his radiological findings did not improve. Follow-up after 4 months with a plain CT scan revealed ex-vacuo dilatation of the right lateral ventricle in keeping with previously noted findings. Additionally, a wedge-shaped hypodensity in the right occipitotemporal lobe associated with loss of grey-white matter differentiation and effacement of adjacent sulci was noted (Fig. 5A). Similarly, plain CT scan follow-up after 9 months revealed progressive findings in keeping with cerebrovascular gliosis of the previously noted infarctions (Figs. 5B and C).

Discussion

Diabetic striatopathy is a modern concept for a condition referred to as hyperglycemic non-ketotic hemichorea and hemiballismus 6 decades ago [6]. It is associated with

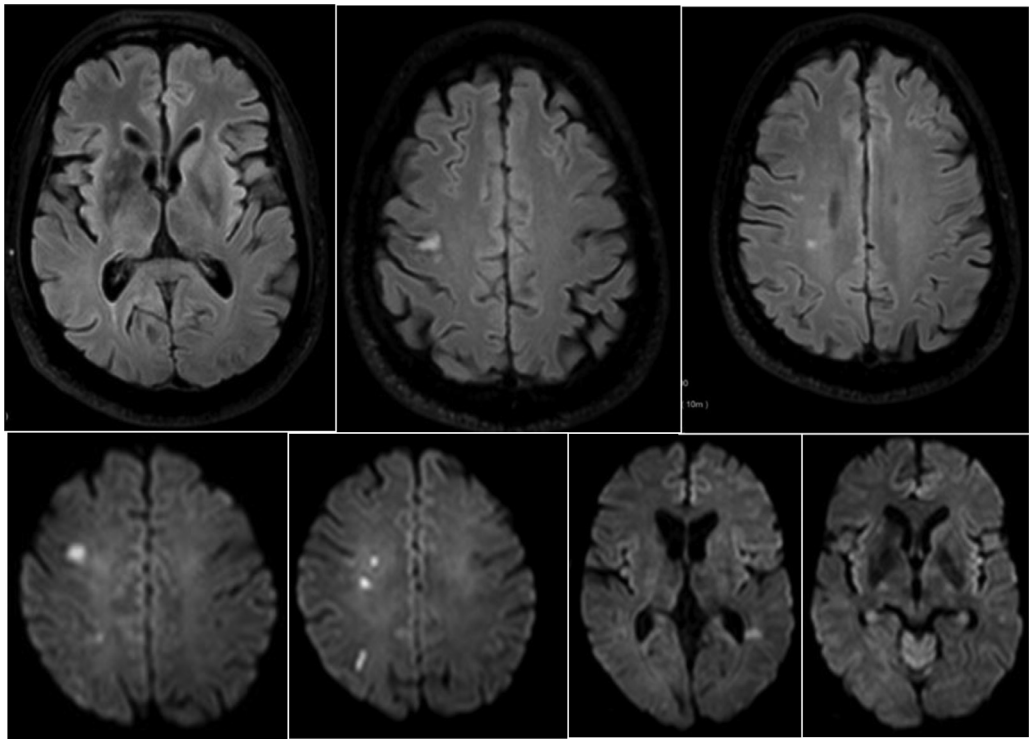

Fig. 2 – Flair and DWI sequence of brain MRI: demonstrating multiple areas of acute cortical lacunar infarctions.

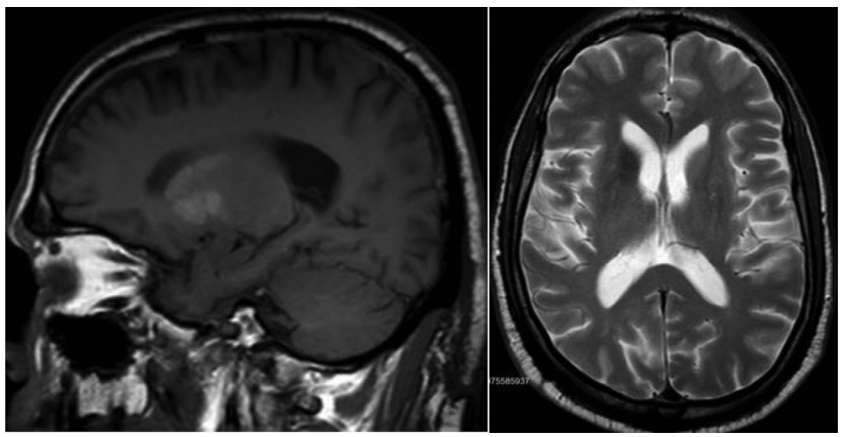

Fig. 3 – T1 and T2 sequence of brain MRI respectively: demonstrating high signal intensity in T1 and low T2 signal intensity and changes at the region of the right basal ganglia.

extremely elevated blood sugar, which occurs mostly without the production of ketones, and it is mostly associated with type 2 diabetes mellitus [4]. However, some cases of DS were detected in type one diabetic patients, and some had positive ketone in their urine [6]. Nevertheless, several studies have also reported this condition in newly diagnosed patients with diabetes [1–8].

DS is a rare condition that affects 1 in every 100,000 [5]. Uncontrolled long-standing diabetes, old age, Caucasian ethnicity, and female gender are risk factors observed in many studies and reports [1]. An observation of 20 patients conducted in 2016 revealed a majority of Asian ethnicity and female gender associated with DS, in addition to an age average of 67.8 years [9]. Similarly, 53 cases observed in 1960 by Bedwell revealed that females are more susceptible to developing this condition than males, with a 1.7-1 ratio. In addition, he identified the average age for this condition as 71.1 years old and found that 91% of the patients were of Asian ethnicity [10].

DS is classically present with sudden involuntary and uncontrollable jerky limb movements, mostly limited to a single limb or unilateral limb with less commonly bilateral limb involvement [4,11], and to a lesser extent associated with facial muscles involvement like jaw or tongue [1,4]. In more severe cases, patients may present with ballism, which is more

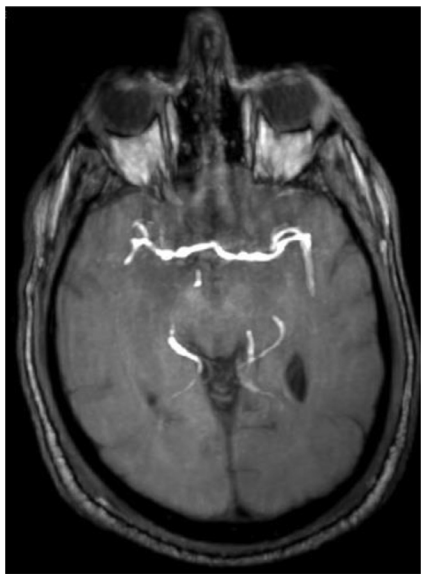

Fig. 4 – MR angiogram: demonstrating multifocal narrowing of bilateral MCA—M1 and both PCAs.

proximal and frequent than chorea. In very rare severe cases, patients could present with more aggressive neurological complications like seizures, slurred speech, decreased level of mentation, or coma [1,7,12].

Most of the pathological hyperkinetic movement or chorea reflect an insult to the basal ganglia that could result from many pathologies detected through CT or MRI images, such as; Huntington, Wilson disease, or related to autoimmune conditions or paraneoplastic syndromes in malignancies [1,13]. Some toxins could also be responsible for bilateral basal ganglia lesions and can be associated with altered mental status or even coma, such as; carbon monoxide, cyanide, and methanol toxification [14]. Moreover, hyperglycemia and hyperammonemia can lead to basal ganglia hyperdensity in CT images and MRI hyperintensity [14]. In addition, vascular hemichorea results from either ischemia or hemorrhage to the highly vascular basal ganglia, a major cause of sudden chorea or hemiballismus [13].

Although the exact pathophysiology behind DS remains unclear, however, several studies investigated this phenomenon and suggested multiple theories to explain this condition. One of which stated that severe hyperglycemia induces hyperviscosity of the blood flow, which is thoughts to be related to vascular insufficiency that could result in cytotoxic edema with deposition of paramagnetic material, thus interfering with neurotransmitter metabolism and cause transient dysfunction of the basal ganglia [1,13]. Chorea was proposed to develop because of dysfunction of γ aminobutyric acidergic projection neurons resulting from the ischemic insult, which in turn led to the loss of basal ganglia tonic inhibition and thus caused chorea [13].

In contrast, in this case, our patient was an elderly Caucasian male, who did not present with the classical manifestations of hemichorea or hemiballism mentioned earlier. Instead, he presented with stroke-suggestive signs and symptoms represented as unilateral hemiparesis in the left upper and lower limbs associated with slurred speech. Accordingly, given the history of previous myocardial heart ischemia this patient has, stroke was highly probable. However, the initial CT scan images showed unilateral striatal hyperdense right caudate and lentiform nucleus, which is further suggestive of basal ganglia pathology, and based on the severely hyperglycemic stat of this patient, other possible deferential raised, such as diabetic striatopathy. With further imaging investigation, the patient was found to have a combination of both acute cortical lacunar infarctions with evidence of basal ganglia predisposition suggested by the CT images and redemonstrated by MR images in the form of low signal intensity changes, with no evidence suggesting hemorrhage. Consequently, this patient was diagnosed with DS in the presence of stroke.

Conversely, Lancellotti et al. [15] presented a case of DS associated with ischemic stroke with presenting

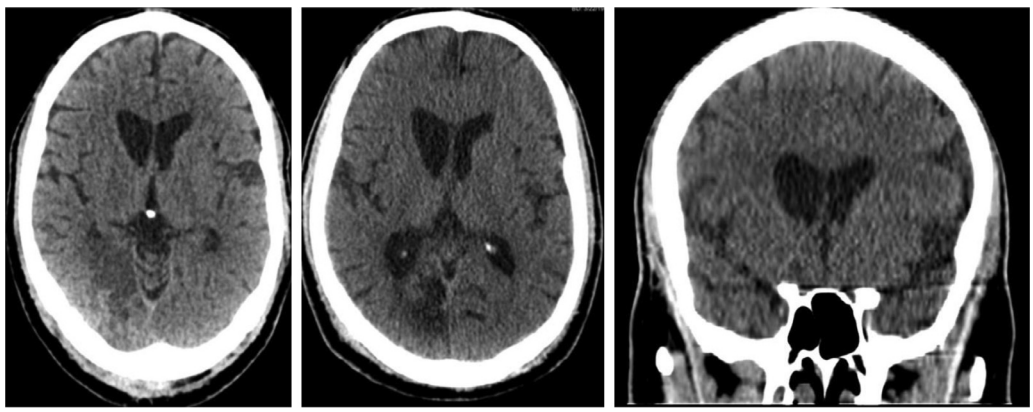

Fig. 5 – (A) Axial cut of plain CT brain - after 4 months. (B, C) Axial and coronal cuts of plain CT brain respectively—after 9 months follow-up: demonstrating ex-vacuo dilatation of the right lateral ventricle and cerebrovascular gliosis of the previously noted infarctions.

symptoms of hemichoreoathetosis for 48 hours. Their patient was treated with insulin, and platelet anti-aggregation, which consequent in dramatic cognitive and motor improvement. Their patient experienced multiple recurrences of similar symptoms within 24 months. However, he showed complete remission with strict sugar control after 24 months. Moreover, Carrion DM and Carrion AF [16] reported a patient who presented with diabetic striatopathy with no evidence of stroke symptomatically or radiologically. Their patient was treated and achieved complete remission. However, he represented with the same complaint one week later. On the fifth day of his new admission, a repeated CT scan showed an ischemic stroke involving the same presentation territory.

Management of DS is mainly through hyperglycemia control, and many cases showed dramatic improvement after normalizing blood sugar [1]. Adding a small dose of atypical antipsychotic to control chorea in more severe cases was suggestive and improved the overall outcome of the disease management [4,17]. The prognosis of the disease is highly dependent on the presentation severity and, to a lesser extent time interval between the beginning of the symptoms to the treatment intervention [1]. Although symptoms of medical intervention time are not as significant as the presentation severity, early detection and intervention showed a better outcome in many cases [18,19]. Thus, clinical insight of DS and any other pathologies accompanying it is essential. The median time recovery for patients with milder symptoms of diabetic striatopathy and the minimum time for CT follow-up resolution was found to be 2 days and ten days respectively, in a systemic review conducted in 2020, which is much lesser than those with more severe presentations. In our case, the patient showed partial symptomatic remission within 9 months of follow-up. However, significant volume loss and gliosis of the caudate lobe after the resolution of the unilateral basal ganglia hyperdensity on the CT scan.

Given the wide variety of symptoms, neuroimaging presentations, prognosis, different treatment strategies effectiveness, symptomatic remission time, resolution time of neuroimaging anomalies, and incidence of symptom recurrence of diabetic striatopathy, more investigation of this condition is warranted, along with more awareness among clinical physicians to be aware it and its variations.

Conclusion

In conclusion, although DS is a rare condition, it could present with different scenarios and need different management interventions depending on the presenting context. Thus clinical and radiological physicians need to be alert of its possibility in the presence of chronic or newly diagnosed diabetic patients, as its timely-bound condition and prognosis partially depend on the time window of detection and management. Nevertheless, the radiologist needs to pay attention to the distinct features of this condition, and should not dismiss other entities, such as acute infarction in patients with atypical presentation.

Patient consent

The patient was consented for the publication purposes of his clinical case as per hospital policy and the ethical committee protocol.

This form state that the patient gave his full permission for the publication, reproduction, broadcast, and other use of photographs, recordings, and other audio-visual material in all editions of the above-named product and in any other publication (including books, journals, CD-ROMs, online and internet), as well as in any advertising or promotional material for such product or publications. Thus, he declared that he has no claim on the ground of breach of confidence or any other ground in any legal system against—(Salmaniya Medical complex)—and its agents, publishers, successors and assigns in respect of such use of the photograph(s) and textual material (case histories).

He agrees to release and discharge the authors named above, and any editors or other contributors and their agents, publishers, successors, and assigns from any and all claims, demands, or causes of action.

This statement was written by the corresponding author to confirm that an informed written consent was obtained from the patient for publication purposes. A copy of the original patient consent is stored in the patient record at the sponsoring hospital.

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