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Case report: CT and MRI in diabetic straital disease with Hemiballism-hemichorea

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Abstract
Diabetic striatopathy is an uncommon and life-threatening manifestation of *Diabetes mellitus*. Individuals of Asian descent, females, and the elderly are till now reported to be commonly affected by this disease. The affected patients present with sudden onset hemiballism-hemichorea which is likely caused by nonketotic hyperglycemia. Hemiballism-hemichorea is involuntary continuous random appearing movement involving one half of body. Diagnosis is usually based on imaging findings and blood analysis. In this case report, our aim is to present imaging findings in an adult man with diabetes mellitus presenting with sudden onset of hemiballism-hemichorea.

Keywords: Diabetic striatopathy, Nonketotic hyperglycemia, Hemiballism-hemichorea, CT, MRI

Introduction
A 50 yr old professional tailor presented with slurring of speech and abnormal movement of his left upper limb for about 5 days. He had already been diagnosed with Type 2 *Diabetes mellitus* (T2D) before and had taken medication, albeit in an irregular manner, for the last 5 years. He is a smoker. On first examination, no other significant findings were found, except for a continuous abnormal movement of the left upper limb.
A computer tomography (CT) scan revealed hyperdense right basal ganglia, including the caudate nucleus and putamen. A random blood sugar (RBS) test revealed a a glucose concentration of over 500mg/dL and 13.4% glycosylated hemoglobin (HbA1c), indicating and confirming the previous T2D diagnosis. No evidence of ketones was found in the urine, which is not out of line with the patient suffering from T2D; it simply suggests the man does not suffer from diabetic ketoacidosis (DKA) or similar complications.
In magnetic resonance imaging (MRI), an altered signal intensity was found that involved the right caudate head and putamen; specifically, they were hyperintense on T1W1 and hypointense on T2W1 and fluid-attenuated inversion recovery (FLAIR).
The above findings with associated uncontrolled hyperglycemia and sudden onset of hemiballism-hemichorea suggested the diagnosis of diabetic striatopathy. This condition is characterized by a hyperactivity in the corpus striatum on T1W1, associated with contralateral motoric problems. As Diabetes is well-known to cause neurological problems, the existence of diabetic striatopathy does not seem unexpected, though it would be interesting to know why only specific regions in the brain are affected at first [1].

Discussion
A hyperactive putamen in line with motor activity affection
Diabetic striatopathy (DS) is a unique, rare and potentially life-threatening complication of uncontrolled T2D. It can accompany diabetic patients with *non-ketotic hyperglycemia* (NKH). This rare cause of hemichorea-hemiballism (HH) is characterized by unilateral, involuntary, poorly patterned movements developing over a period of hours; the average age of onset is 72 years. This is predominantly seen in elderly patients with poorly controlled T2D, as it appeared to be the case with this patient, albeit with 50 years, he was distinctly younger than 72 years. The younger age is reminiscent of other cases that have been more recently found. For example, in a study with n = 20 participants suffering from NKH, the average age of onset of hemichorea-hemiballism is almost 68 years, while 5 patients are less than 60 years old, with one individual even of 29 years.[2] As visible from CT and MRI scans, the putamen on the contralateral side to movement defects was affected in all cases.

In some of the cases analyzed, movement problems were bilateral, suggesting lateral specificity may not be a defining factor of the observed condition; this suggests that defects in the putamen and basal ganglia may be the result of general neuropathies under diabetic conditions. This also fits with the observation from this and other case studies that HH occurs after poorly controlled dietary behaviors, although Cosentino *et al*. could not find a significant relationship the control of diabetes and the occurrence of the condition, suggesting that multiple factors play a role in the development of DS [2].

Women are more commonly affected than men, and the majority of reports mention a predilection for individuals of Asian descent, which may indicate a propensity for DS on the genetic level [3]. It is possible that the genes that are significantly responsible for precipitating the condition are linked to the X chromosome, although genetic studies for hemichorea-hemiballism in conjunction with NKH are lacking. As hemichorea-hemiballism is a rare occurrence, the bias towards patients that are female and of Asian descent could also simply be due to unspecific effects, e.g. a low and as yet unrepresentative number of recorded cases. In principle, the condition does occur in other ethnicities as well.2 More studies, including genomic analyses, are necessary to determine whether there is any meaningful influence of genetic background.

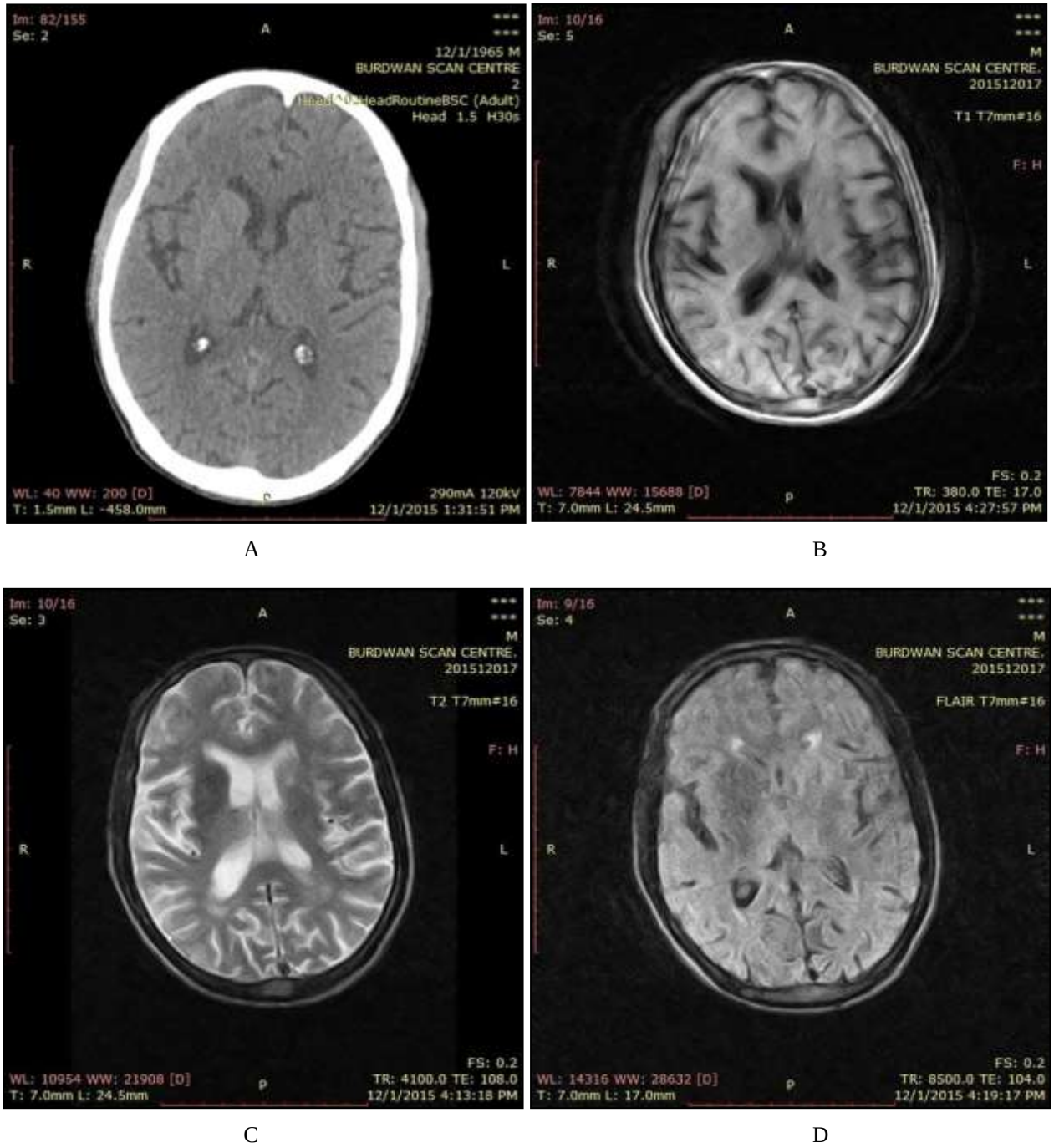
This patient's absence of ketonurea, together with elevated levels of blood glucose and HbA1C, and an absence of ketones in blood or urine, is in line with previous findings [4]. The movement problems on the left side of the body are in line with a hyperactive part of the brain - the putamen - on the right side, as the MRI and CT scan pictures also show. On the molecular level, involuntary movements as observed in this patient are thought to be the consequence of a hyperactive dopaminergic state due to GABA depletion in patients with NKH [5].

DS is a unique syndrome comprising of movement disorders with imaging abnormalities in the contra-lateral striatum. The characteristic imaging features include hyperdensity involving the striatum on plain CT which is not always well visible. Using an MRI, the most characteristic and reproducible finding is hyperintensity in the striatum on T1WI. This may be reflected by hypointensity on T2W and FLAIR images, attributed to petechial hemorrhage with hemosiderin deposition. When using diffusion weighted images, effects vary between remarkable and restricted diffusion. Proton MR spectroscopy may reveal acdecreased NAA/creatine ratio and an increased choline/creatine ratio.

Various theories have been proposed for the characteristic hyperintense MRI signal changes on T1W images in diabetic striatopathy, which include obliterative vasculopathy with prominent vascular proliferation involving the striatum; petechial haemorrhages involving the putamen, which may result from a compromise of the blood brain barrier due to underlying chronic focal cerebrovascular disease in diabetics; and, lastly, myelinolysis. In hemichorea-hemiballism, the contralateral basal ganglia are involved as seen in this case. In generalised chorea and ballismus, bilateral lesions may be seen.

The other important causes for T1 hyperintensity in the basal ganglia include Wilson’s disease, which can be differentiated from diabetic striatopathy by the presence of T2 hyperintensity involving basal ganglia and thalamus [6]. The other conditions include hepatic encephalopathy, manganese toxicity and carbon monoxide poisoning. Characteristic T1 hyperintensity with elevated blood sugar levels helps in clinching the diagnosis of diabetic striatopathy, which, on follow-up images, may show regression of imaging findings.

The importance of diagnosing this syndrome lies in the fact that it is a treatable condition and the patient’s symptoms can regress after correction of the blood glucose levels. However, as mentioned above, HH is a rare complication of Diabetes or hyperglycemia and may therefore not always be properly diagnosed, especially since it can occur amidst to an as yet undiagnosed T2D. It can be characterized by facial ticks and tremors of the arms and legs. Diagnosis-wise, it will show up in many cases in a similar manner as documented here on CT and MRI scans, yet this is not necessarily the case. Moreover, HH patients sometimes present with intermittent slow waves in an electro encephalogram. This suggests that HH has a complex etiology and can be caused by behavioral as well as genetic changes [7]. More research is required that links genetic background, neural function, behavior and nutrition, ideally in longitudinal studies, to find the causes of HH in NKH and other diabetic conditions. "There are no financial conflicts of interest to disclose."



A B

C D

Fig 1: (A) CT showing hyperdense right putamen and caudate head.
(B) T1WI showing hyperintense right putamen and caudate head.
(C) T2WI showing hypointense right putamen and caudate head.
(D) FLAIR image showing hypointense right putamen and caudate head.

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