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CASE REPORTS

Chorea, Hyperglycemia, Basal Ganglia Syndrome in Diabetes – Two Rare Case Reports

S. DWARAK[a], K. PRABHAKAR[a], Mahesh KAKANALE[a], Anitha ASWATHANARAYANA[a], B. R. SINDHU[a]

[a]Department of General Medicine, Sri Devaraj Urs Medical College, India

ABSTRACT

Neurological manifestations of diabetes are protean. Hyperglycaemia induced involuntary movements (HIIM) are uncommon manifestations of diabetes seen especially in those with poor glycaemic control. Of these, basal ganglia syndrome (C-H-BG) is a rare entity which can mislead the unwary. Awareness and prompt recognition of C-H-BG syndrome is crucial, since correction of hyperglycaemia can lead to favourable clinical and radiological improvement.

In this article, we report two patients presenting with C-H-BG syndrome on a background of poor glycaemic control. The clinical features of these two elderly diabetic females were characterized by hemichorea-hemiballismus in the first patient and generalised bilateral chorea/ballism in the second one. Radiologic findings associated with C-H-BG syndrome were characterized by hyperattenuation and hyperintensity on T1-weighted magnetic resonance imaging (MRI) at bilateral basal ganglion regions in both cases. Strict glycaemic control formed the corner stone of treatment for the two cases. The prognosis of nonketotic hyperglycemia-induced hemichorea-hemiballism is good and depends on the prompt recognition and optimal glycaemic control in diabetic patients.

Keywords: chorea, hyperglycemia, basal ganglia syndrome.

INTRODUCTION

Neurological manifestations of diabetes are protean. Stroke and peripheral neuropathy are the commonest macro- and microvascular complications involving the central and peripheral nervous system, respectively (1). Hyperglycemia induced involuntary movements (HIIM) are uncommon manifestations of diabetes seen especially in those with poor glycaemic control. Of these, basal ganglia syndrome (C-H-BG) is a rare entity which can mislead the unwary (2). Awareness and prompt recognition of C-H-BG syndrome is crucial since correction of hyperglycaemia can lead to favourable clinical and radiological improvement.

In this article, we report two patients presenting with C-H-BG syndrome on a background of poor glycaemic control.

Address for correspondence:
Dr. S. Dwarak, Junior Resident
Sri Devaraj Urs Medical College, Tamaka, Kolar – 563101, Karnataka, India
Tel.: 9789225092
Email: dwarak.acf@gmail.com

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Case 1

A 56-year-old normotensive diabetic lady presented with a one-week history of involuntary movements of left arm and leg. Initially, she noticed her left hand being fidgety which gradually progressed in intensity to wide flinging movements. The patient was diagnosed with type 2 diabetes mellitus five years prior to the date of presentation and had been non-compliant with her diabetic medications during the last month. There was no preceding history of stroke, fever, trauma or intake of any drugs. On physical examination, the woman was afebrile, conscious and oriented. Her blood pressure was 130/80 mm Hg and she had a regular pulse of 80/minute. She was noted to have left hemichorea with a ballistic component. Initial biochemistry revealed blood glucose 435 mg/dL, normal arterial pH (7.37) and negative urine ketones. Her glycated haemoglobin level was 13%. Complete hemogram, serum electrolytes including serum sodium, potassium, calcium, urea and creatine were normal. Viral serology for HIV, hepatitis B, hepatitis C, liver function tests, lipid profile, thyroid function tests, 2D ECHO and ANA were also normal. MRI brain showed diffuse asymmetric hyper intensities involving basal ganglia and right putamen and caudate nuclei on T1 weighted and T2 weighted images (Figure 1). These lesions were characteristic of chorea, hyperglycaemia, basal ganglia syndrome (C-H-BG). Subsequently, the patient was initiated on a twice daily regimen of mixed insulin preparation and glycaemic control was optimized. She had complete remission of signs and symptoms after control of blood sugar levels within five days.

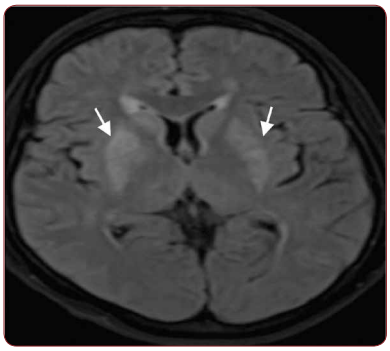

FIGURE 1. Diffuse asymmetric hyper intensities involving bilateral basal ganglia including putamen and caudate nuclei (white arrows) on T1 weighted MRI (right more than left)

Case 2

A 55-year-old normotensive woman, recently diagnosed with type 2 diabetes mellitus, presented with involuntary non-rhythmic movements of both arms and legs (on the left side more marked than the right side) of three days duration. The patient reported that the involuntary movement initially started in her left arm and then progressed, involving her left leg within six hours. The next day, she had a similar sequence of events occurring in her right upper and lower limb. The patient had no history of weakness of the limbs, altered sensorium, fever or trauma, and these involuntary movements resolved during her sleep. Fifteen days prior to the current presentation, she was newly diagnosed with diabetes mellitus with high glycaemic levels. Her random blood sugar level was 525 mg/dL on her previous visit, with no acidosis and normal serum osmolality. Because her haemoglobin A_{1c} was 15%, she was initiated on multiple daily injections of short and intermediate acting insulins. However, she was poorly compliant with the treatment regimen.

Physical examination findings revealed bilateral upper and lower extremity choreiform and ballistic movements which were observed on the left side more than the right side. The patient had normal sensorium and higher mental functions. Complete cranial nerve examination was performed and found to be normal. The patient was also observed to have normal muscle tone, strength and deep tendon reflexes in all four limbs. Sensory system examination and Babinsky's sign were normal.

Initial biochemistry revealed a random blood glucose of 158 mg/dL, normal arterial pH and negative urine ketones. Complete hemogram and serum electrolytes, including serum sodium, potassium, calcium, urea and creatine, were also normal. Viral serology for HIV, hepatitis B, hepatitis C, liver function test, lipid profile, thyroid function tests, 2D ECHO and ANA were normal too. MRI imaging showed diffuse hyper intensities involving bilateral basal ganglia – putamen and caudate nuclei on T1 weighted and T2 weighted images (Figure 2). These lesions were characteristic of chorea, hyperglycaemia and basal ganglia syndrome (C-H-BG). Subsequently, she was re-initiated on regular human insulin preparation and glycaemic control was opti-

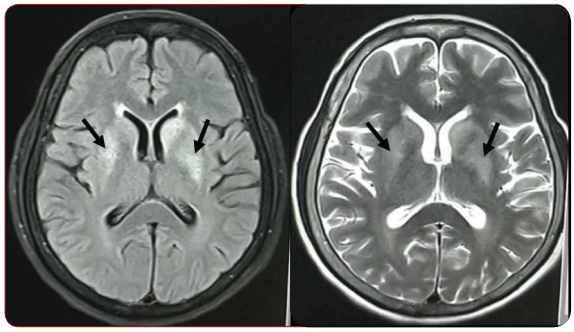

FIGURE 2. Diffuse hyper intensities involving bilateral basal ganglia including putamen and caudate nuclei (black arrows) on T1 weighted and T2 weighted MRI

mized. She has been also administered a short course of olanzapine 10 mg to control movements, as they were disabling. She had complete remission of signs and symptoms after optimization of blood sugar levels within 14 days .

Table 1 summarises the clinical, laboratory and radiological characteristics of both cases. ❑

DISCUSSION

The clinical features of these two elderly females were characterized by hemichorea-hemiballismus in the first patient and generalised bilateral chorea/ballism in the second patient on a background of poorly controlled glycaemic status.

The first patient had high sugar levels on presentation, while the second one had normal sugar levels on presentation but long term poor glycaemic control, as showed by her high glycated haemoglobin A1C and recent admission for high sugar levels.

Radiologic findings associated with both patients with C-H-BG syndrome were characterized by hyperattenuation on computed tomographic (CT) scans and asymmetrical hyperintensity on T1 weighted magnetic resonance imaging (MRI) at bilateral basal ganglion regions in both the cases.

Strict glycemic control formed the corner stone of treatment in both cases. Dopamine receptor antagonist like olanzapine was effectively used to control choreiform movements.

The differential diagnosis for a patient with acute sporadic chorea would include drug-induced, vascular, autoimmune, infectious, metabolic, paraneoplastic, and other miscellaneous etiologies. Hyperglycaemia was the only meta-

Parameters (normal lab values)	Case 1	Case 2
Age	56	55
Sex	Female	Female
Duration of diabetes	Five years	15 days (recently diagnosed)
Treatment	Tablets of Metformin 500 mg OD and Glimepride 2 mg OD	Nil
Clinical features	Left upper and lower limb hemichorea/ hemiballismus	Generalised (bilateral) chorea/ballismus
Duration of symptoms	Seven days	3 days
Initial random glucose levels (< 200 mg/dL)	435	158
Glycated haemoglobin (<6.5%)	13%	15%
Blood urea (20-40mg/dL)	22	24
Serum creatinine (0.7–1.4 mg/dL)	0.6	0.6
Serum sodium (135–145 mEq/L)	135	138
Serum potassium (3.5–5 mEq/L)	4.5	3.8
WBC (4.5–11.0×103/UL)	8.4×10^3	7.2×10^3
Haemoglobin (12.0–15.0 g/dL)	11	12.8
Haematocrit (36.0–47.0%)	34	38
Platelets (150–450×103/UL)	358	
Chloride (96–106 mEq/L)	98	100
CO_2 (21–32 mmol/L)	25	22
Calcium (8.5–10 mmol/L)	9.2	9.4
Albumin (3.4–5.0 g/dL)	3.5	3.8
Protein (6.4–8.2 g/dL)	6.5	6.2
AST (15–37 IU/L)	22	32
ALT (12–78 IU/L)	26	25
Alkaline phosphatase (50–136 IU/L)	65	55
TSH (0.35–5.0 MIU/L)	3.5	2.3
ANA	Negative	Negative
MRI Findings	Diffuse asymmetric hyper intensities involving bilateral basal ganglia including putamen and caudate nuclei on T1 weighted MRI (right more than left)	Diffuse hyper intensities involving bilateral basal ganglia including putamen and caudate nuclei on T1 weighted and T2 weighted MRI
Time for resolution of dyskinesia	Five days	14 days

TABLE. Clinical, laboratory and imaging characteristics of patients with C-H-BG Syndrome

bolic factor operative in both of our cases and we could rule out other causes. Chorea/ballism associated with non-ketotic hyperglycaemia, though an uncommon manifestation of hyperglycaemia, can be the first presenting sign of undiagnosed diabetes mellitus or can develop after weeks or even months of poor glycaemic control, as seen in case 1. In case 2, the development of C-H-BG syndrome was more of a legacy effect of poor glycaemic control. Case reports of similar phenomenon have been published (1). It is not entirely clear how hyperglycaemia leads to this syndrome. Proposed hypothesis for hyperglycaemia as a cause of these movement disorders involve hyperglycaemia induced intracellular hyperosmolality and ischemia in the putamen with the additive effect of anaerobic metabolism leading to gamma-aminobutyric acid (GABA) depletion (2). This may also result in disinhibition of the thalamocortical pathway, leading to motor cortical hyperexcitability (3). Neuroradiological involvement of putamen is characteristic. In most cases, the head of the caudate nucleus is also involved, and rarely globus pallidus. Lesions have been reported to be unilateral in the majority of cases, as seen in our cases bilateral involvement (3, 4). The characteristic finding on a T1 weighted MRI is high signal intensity in the contralateral putamen if there is hemichorea/hemiballismus, or bilateral putamen if there is bilateral chorea/ballismus without signs of mass effect, oedema, or volume loss. In contrast, the findings in T2 weighted images are variable, with signal characteristics ranging from hyper- and iso- to hypointensity.

Computed tomography (CT) can also reveal hyperattenuation in the striatum contralateral to the affected side or both sides. An abnormal signal can encompass the globus pallidus and up to the medial part of the cerebral peduncle in the midbrain along the striatonigral pathway (5). Magnetic resonance spectroscopy may reveal low N-acetylaspartate to creatinine ratio and high choline-to-creatinine ratio and associated lactate peak (6). Positron emission tomography (PET) may reveal reduced glucose metabolism in the basal ganglia. ❑

CONCLUSION

The two cases described here have shown that chorea, hyperglycaemia, basal ganglia syndrome (C-H-BG) is an uncommon adverse event of nonketotic hyperglycaemia or long term poor glycaemic control in elderly patients with type 2 diabetes. It is associated with contralateral or bilateral striatal radiological abnormality and typically T1 hyperintensity on MRI. The prognosis of non-ketotic hyperglycaemia-induced hemichorea-hemiballism is good and depends on the prompt recognition and optimal glycaemic control in diabetic patients. Rapid clinical improvement is the rule with adequate control of diabetic status though radiological signs may take longer time of more than one year to resolve (7). ❑

Conflicts of interest: none declared.
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