ABSTRACT

Diabetic Basal Ganglia Striatopathy is a rare and life-threatening manifestation of diabetes mellitus occurring more frequently in people of Asian descent, females, and the elderly. The usual presentation is of a Hemiballismus - hemichorea caused by non ketotic hyperglycemia the symptoms of which are reversible with correction of hyperglycemia.

Chorea, Hyperglycemia, Basal Ganglia Syndrome (C-H-BG) can be a delayed onset manifestation of involuntary movements in the form of chorea or ballismus in patients with uncontrolled hyperglycemia which usually occurs weeks to months after an episode of non ketotic hyperglycemia.

Here we present a case report of a 65 year old diabetic patient with poorly controlled glycemic status with complaints of involuntary movements of left upper and lower limbs of 4 days duration. At the time of presentation he had normal blood sugar values with a very high HbA1c value not measurable in laboratory parameters. MRI Brain showed hyperintense signals in both basal ganglia on T2 weighted and FLAIR images which are isointense on T1 images. After exclusion of other possible causes a diagnosis of Chorea, Hyperglycemia, Basal Ganglia Syndrome (C-H-BG) was made. Strict glycemic
control was maintained. Patient treated supportively with haloperidol. Patient improved by 3 months of treatment

INTRODUCTION

The most common cause of hemichorea-hemiballism (HCHB) syndrome is the vascular insult in contralateral striatum or subthalamic nucleus, however it can occur secondary to a variety of causes like tumors, encephalitis, neurodegenerative disorders, drugs, chronic subdural hemorrhage and metabolic disorders such as non-ketotic hyperglycemia.

CASE REPORT

A 65 year old male patient with history of T2DM of 20 years duration presented to our casualty with complaints of acute onset of involuntary movements of both proximal and distal muscles of left upper and lower limbs for a duration of 4 days. He denied any complaints of diplopia, seizures, blurred vision, headache, fever or weakness of limbs. He had a past history of admission for diabetic ketoacidosis which was corrected from a local hospital. On examination patient was conscious and oriented to place, time and person. His Respiratory, Cardiovascular, Abdomen examination were within normal limits. His nervous system examination was normal and revealed no focal neurological deficits.

<table>
<thead>
<tr>
<th>Hemoglobin</th>
<th>13.4 g/dl</th>
<th>RBS</th>
<th>150mg/dl</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total count</td>
<td>7600/ microlitre</td>
<td>Renal function (mg/dl)</td>
<td>42 /0.8</td>
</tr>
<tr>
<td>Differential count</td>
<td>L24M4N72</td>
<td>Serum electrolytes (mmol/L)</td>
<td>134/3.7</td>
</tr>
<tr>
<td>PLATELET</td>
<td>3.6 lakhs/ microlitre</td>
<td>Total bilirubin/ Direct Bilirubin (mg/dl)</td>
<td>0.8/0.1</td>
</tr>
<tr>
<td>MCV/MCH</td>
<td>86 fl/30pg</td>
<td>Total Protein/ Albumin (g/dl)</td>
<td>7.2/3.6</td>
</tr>
<tr>
<td>MCHC</td>
<td>31g/dL</td>
<td>SGOT/SGPT (IU/L)</td>
<td>35/33</td>
</tr>
<tr>
<td>ESR</td>
<td>30mm/1st hour</td>
<td>ALP (IU/L)</td>
<td>112</td>
</tr>
</tbody>
</table>

CHBG syndrome is the most common cause of unilateral chorea in type 2 diabetes mellitus patients with the underlying mechanism of non-ketotic hyperglycemia. The characteristic imaging manifestation is T1 shortening in the contralateral corpus striatum. Therefore, it should always be included in the differential diagnosis of chorea, especially in elderly patients.
His laboratory investigations showed RBS-150 mg/dl, with normal hemogram, Liver function, Renal function tests and Thyroid function tests.

However his HbA1c was very high which could not be measured by laboratory parameters.

MRI Brain showed hyperintense signals in both basal ganglia on T2 weighted and FLAIR images which were isointense on T1 images (fig1)

Patient was treated with Haloperidol, subcutaneous insulin, other supportive measures with strict glycemic monitoring. He improved symptomatically and was discharged and kept on follow-up. The hemichorea-hemiballismus improved gradually over a period of 3 months.

DISCUSSION

Generally, the blood sugar level is extremely high during hemichorea onset. However, in this case report, we present an unusual disease course, in which the involuntary movement occurred after hyperglycemia correction. In the literature, very few cases of delayed-onset hemichorea after hyperglycemia correction have been reported. Bizet et al presented a case report of a 66-year old woman with hyperosmolar hyperglycemic nonketotic syndrome. Three months after admission, she developed chorea for 1 week at the euglycemic state (blood sugar level: 84mg/dL).

The characteristic imaging finding of CHBG syndrome is T1 hyperintensity in contralateral corpus striatum.
The differential diagnosis of T1 hyperintense lesion in the basal ganglia is wide, including methemoglobin in intracranial hemorrhage, manganese deposition due to parenteral nutrition, copper accumulation in Wilson’s disease, hemorrhagic infarction, Japanese encephalitis, calcification, hamartoma (neurofibromatosis-1), hypoxic ischemic encephalopathy, acquired hepatocerebral degeneration and CHBG syndrome.

Although imaging findings are characteristic, the origin of T1 hyperintensity remains speculative. There are many theories like ischemia, metabolic acidosis, petechial hemorrhages, depletion of acetyl choline among which ischemia is considered to be the most plausible explanation. Advanced neuroimaging studies, such as magnetic resonance spectroscopy (MRS), single-photon emission computed tomography (SPECT), and 2-[18F] fluoro-2-deoxy-D-glucose-positron emission tomography (FDG-PET), have provided some possible explanations. MRS revealed relatively low N-acetylaspartate to creatine ratio, high choline to creatine ratio, and lactate peak, indicating neuronal damage, gliosis, and acute or chronic ischemic changes, respectively. Furthermore, SPECT suggested hypoperfusion in the striatum, and FDG-PET demonstrated reduced rates of cerebral glucose metabolism in the corresponding areas with abnormalities on T1-weighted MRI.

The molecular pathogenesis underlying hyperglycemic hemichorea remains unclear. Hyperglycemia induced ischemia appears to be the most common proposed mechanism. g-aminobutyric acid (GABA) is used as an alternate energy substrate during hyperglycemic crisis. GABA depletion leads to thalamic disinhibition and hyperkinesias. Vasculopathy is also hypothesized to be a possible mechanism. Long-term hyperglycemia causes hyperviscosity of blood, which leads to latent ischemia of the striatum and subsequent dyskinesia. It is partially supported by a striatal biopsy after hyperglycemia-related hemichorea, which showed neuron loss, gliosis, and reactive astrogliosis. Ketone bodies may play a role in hyperglycemia-related hemichorea. The absence of insulin leads to hyperglycemia and the release of free fatty acids from the adipose tissue, which are metabolized to ketone bodies (acetoacetate and b-hydroxybutyrate). Acetoacetate can be used as a GABA substitute to temporarily compensate for the hyperglycemia-induced GABA depletion. Thus, this finding reflects the rarity of hemichorea in patients with diabetic ketoacidosis. Delayed chorea may be due to temporarily compensate for the GABA depletion by high level ketone bodies initially.

CONCLUSIONS

Uncontrolled diabetes has many well-known adverse effects and clinical presentations. However, this case highlights the importance of recognizing its infrequent manifestations. Chorea-type movement, hyperglycemia, and basal ganglia changes found on magnetic resonance imaging in patients with uncontrolled T2DM are the hallmarks of C-H-BG. Although the pathophysiological process of this syndrome is unknown, it is vital for physicians to identify C-H-BG as one of the rare and reversible
complications of uncontrolled diabetes to ensure early diagnosis and management and good outcomes.

REFERENCES:


