Osmotic Demyelination Syndrome Presenting with Chorea

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Abstract
Osmotic demyelination syndrome is an acute shift in serum osmolality causing demyelination, which may be due to rapid correction of hyponatremia, hyperglycaemia, hypokalemia and ketoacidosis.

We present a case of 55yr old female and a known diabetic presented with the choreic movements involving left upper and lower limb for 2 days without any weakness.Her blood sugar was 428mg/dl at the time of admission with wide fluctuations.Her CT Brain showed hyperdensity in the right basal ganglia and the MRI brain showed hyperintense lesion in T2 weighted images showing features suggestive of osmotic demyelination. Patient's sugar levels improved with Insulin and chorea controlled with carbamazepine and sodium valproate.

Osmotic demyelination can occur not only due to rapid correction of hyponatremia but also due to wide fluctuations in the blood sugar levels causing swinging of serum osmolality.

Introduction
Osmotic demyelination syndrome is an acute shift in serum osmolality causing demyelination, which may be due to rapid correction of hyponatremia, hyperglycaemia, hypokalemia and ketoacidosis. The presentation of osmotic demyelination syndrome is usually seizures, altered mentation, dysphagia, dysarthria. Here is the interesting case report of osmotic demyelination which presented as chorea

Case Report
A 55 yr old female presented with the H/o involuntary movements involving the left upper and lower limb for 2 days. She has no history suggestive of weakness of limbs, dyspnoea, fever, loss of consciousness or arthralgia. Patient is a Known case of Type 2 DM under treatment for 10 years irregularly. On examination patient is conscious, depressed, afebrile of blood sugar values showed fluctuations (Table 2).

Her CT brain showed a slight hyperdense lesion on the right caudate and lentiform nucleus. MRI study of brain showed unilateral hyperintense lesion of right basal ganglion on T2 weighted images suggestive of osmotic demyelination. No evidence of hemorrhage or mass effect or midline shift (Figures 1 and 2).

Patient was started on IV fluids, Inj Human Insulin R/B – 15/10-0-15/10 and dose titrated accordingly. T. Sodium Valproate 200 mg two times a day was given along with T.

Haloperidol 1.5 mg two times a day. Blood sugar was controlled from day 5 and her involuntary movements started to disappear from day 8 and completely stopped on day 10. CT brain repeated after 2 weeks of glycemic control (using insulin) showed decrease in hyperdensity with no abnormal enhancement in right caudate and lentiform nucleus.

Discussion
Osmotic demyelination is characterized by acute demyelination caused by rapid shifts in serum osmolarity. The rapid shift in osmolarity causes breakdown of blood brain barrier causing easy access of myelinocytic factor and organic and inorganic osmolytes to injure the axons. The myelinocytic factors are considered to be complement proteins and immunoglobulins. Sites of predilection for osmotic demyelination are pons, cerebellum, lateral geniculate body, external capsule, hippocampus, putamen, cerebral cortex, thalamus and caudate nucleus. An interesting question is that Why certain areas of the brain are more injured than others ? Normally white matter is rich in myelin and grey matter is rich in capillaries. The areas which are admixed with both myelin and capillaries are at high risk.

Conditions which can exacerbate osmotic demyelination are hepatic failure, renal failure, adrenal insufficiency, burns, post-operative states and transplantation. Rapid correction of chronic hyponatremia causes imminent osmotic demyelination. In hyperglycemia, the fluctuating sugar values and hence

Table 1: Baseline investigations

<table>
<thead>
<tr>
<th>Hb g/dl</th>
<th>TC mm</th>
<th>ESR mm</th>
<th>Urea mg/dl</th>
<th>Creatinine mg/dl</th>
<th>Glucose mg/dl</th>
<th>Na mEq/L</th>
<th>K mEq/L</th>
<th>Urine acetone</th>
</tr>
</thead>
<tbody>
<tr>
<td>12</td>
<td>12000</td>
<td>5/10</td>
<td>15</td>
<td>0.8</td>
<td>420</td>
<td>140</td>
<td>4</td>
<td>neg</td>
</tr>
</tbody>
</table>

'Hb, g/dl; TC, mm; ESR, mm; Urea, mg/dl; Creatinine, mg/dl; Glucose, mg/dl; Na, mEq/L; K, mEq/L; Urine acetone

Table 2: Daily fasting blood glucose values of the patient

<table>
<thead>
<tr>
<th>Day1</th>
<th>Day2</th>
<th>Day3</th>
<th>Day4</th>
<th>Day5</th>
<th>Day6</th>
<th>Day7</th>
<th>Day8</th>
<th>Day9</th>
<th>Day10</th>
</tr>
</thead>
<tbody>
<tr>
<td>406</td>
<td>310</td>
<td>480</td>
<td>278</td>
<td>174</td>
<td>140</td>
<td>138</td>
<td>152</td>
<td>118</td>
<td>105</td>
</tr>
</tbody>
</table>

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the osmolarity, is the triggering point for demyelination.

Usual manifestation of osmotic demyelination syndrome are seizures, altered mentation, dysphagia, dysarthria, locked in syndrome and coma. However chorea can be a manifestation if the demyelination involves the caudate nucleus, as in this case.

Prevention of rapid osmotic shift is by meticulous correction of the electrolytes, in this case meticulous correction of hyperglycemia to prevent rapid swings and fluctuations. Strict glycemic control without fluctuation of sugar value is highly recommended.

Treatment includes treating the manifesting symptoms as in this case movement disorder by T. Haloperidol and T. Sodium Valproate. However the control of movement disorder is not easy which can take days and even weeks.

Acknowledgement

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References