Teaching Video NeuroImage: Aurora and Dusk of the Lentiform Fork Sign in a Patient With Reversible Dystonia

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The lentiform fork sign (LFS) is an uncommon imaging finding. Accepted hypotheses include vasogenic edema and metabolic acidosis affecting the lentiform nuclei. MRI imaging in a 46-year-old female undergoing regular hemodialysis showed three stages of the LFS over nine months. Her initial neurological examination revealed severe dystonia [Video 1]. Laboratory testing revealed mild compensated metabolic acidosis with nearly normal bicarbonate and anion gap [Table 1]. MRI sequence scans demonstrated symmetric hyperintensities of the BG
and hyperintense rim delineating the lentiform] [Figure 1]. This case demonstrates that LFS can exist without metabolic acidosis. Vasogenic edema is considered a likely underlying mechanism.

Video 1-http://links.lww.com/WNL/B646
Teaching Slides-http://links.lww.com/WNL/B647

References:

Video 1 legend: In segment 1 of the video, the patient showed severe dystonic movements with *geste antagoniste*, dysarthria, and was unable to remain seated. In Segment 2, the patient showed an improvement in the dystonic movements. The patient can now sit in an upright and comfortable position, and the *geste antagoniste* disappeared. There are mild to moderate signs of dystonia and bradykinesia, discretely worst on the left side.
Figure 1, Brain MRI and the three stages of LFS: 2020/02 - Brain MRI axial T2-FLAIR and STIR-FSE A.a, B.a, C.a no evidence of the LFS; 2020/08 - A.b, B.b, C.b bilateral, symmetric hyperintensities of the caudate, putamen, and thalamus, with the LFS, the hyperintense rim delineating the lentiform nucleus. (white arrowheads) and, 2020/10 - A.c, B.c, C.c show an impressive reduction of the bilateral hyperintensities, and the LFS just vanished (white arrowheads).
<table>
<thead>
<tr>
<th>Laboratory</th>
<th>2020/02</th>
<th>2020/08</th>
<th>2020/10</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glucose (mg/dL)</td>
<td>269</td>
<td>245</td>
<td>246</td>
</tr>
<tr>
<td>Sodium level (Na+) (meq/L)</td>
<td>137,0</td>
<td>129,9</td>
<td>133,3</td>
</tr>
<tr>
<td>K+ (meq/L)</td>
<td>3,7</td>
<td>6,67</td>
<td>4,91</td>
</tr>
<tr>
<td>ion Ca++ (mmol/L)</td>
<td>1,11</td>
<td>1,22</td>
<td>1,17</td>
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<tr>
<td>Bicarb level (HCO3) (meq/L)</td>
<td>20,4</td>
<td>19,9</td>
<td>24,7</td>
</tr>
<tr>
<td>Chloride level (Cl-) (meq/L)</td>
<td>96</td>
<td>97</td>
<td>98</td>
</tr>
<tr>
<td>pH</td>
<td>7.35</td>
<td>7.398</td>
<td>7,45</td>
</tr>
<tr>
<td>pO2 (mmHg)</td>
<td>53,1</td>
<td>66,4</td>
<td>159,5</td>
</tr>
<tr>
<td>pCO2 (mmHg)</td>
<td>39,1</td>
<td>33</td>
<td>39,8</td>
</tr>
<tr>
<td>BE (meq/L)</td>
<td>-5,0</td>
<td>-4,30</td>
<td>-0,6</td>
</tr>
<tr>
<td>Anion gap (meq/L)</td>
<td>20,6</td>
<td>13</td>
<td>10,6</td>
</tr>
<tr>
<td>Lactate (meq/L)</td>
<td>1,2</td>
<td>5,95</td>
<td>2,65</td>
</tr>
<tr>
<td>Hgb (g/d)</td>
<td>9,7</td>
<td>9,6</td>
<td>11,6</td>
</tr>
<tr>
<td>Hematocrit (%)</td>
<td>31,5</td>
<td>28,0</td>
<td>34,0</td>
</tr>
<tr>
<td>Creatinine (mg/dL)</td>
<td>4,93</td>
<td>7,91</td>
<td>3,79</td>
</tr>
<tr>
<td>Urea (mg/dL)</td>
<td>66</td>
<td>159</td>
<td>30</td>
</tr>
</tbody>
</table>

Table 1: The central laboratory result
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