Teaching Neuroimage: Crowned Dens Syndrome, an Acute Attack of Calcium Pyrophosphate Deposition Disease Mimicking Acute Meningitis

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A 88-year-old man presented with acute onset of severe neck pain, meningismus, headache, and fever (up to 38°C). Blood tests showed raised inflammatory markers, but CSF analysis was not suggestive of CNS infection (table). Autoimmune antibodies were negative. Cervical-spine-CT showed calcifications of the transverse ligament of atlanto-axial joint with inflammatory changes revealed by cervical-spine-MRI (figure). Clinical presentation, biochemical and radiological findings were all consistent with crowned dens syndrome. Symptoms and inflammatory markers promptly decreased after a brief course of anti-inflammatory treatment. Recognition of this syndrome is important to differentiate it from other infectious or autoimmune diseases and avoid unnecessary treatment.

Table: Laboratory data on admission and at discharge 10 days later

<table>
<thead>
<tr>
<th>Variable</th>
<th>On admission</th>
<th>At discharge</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Blood</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>White-cell count (× 10⁹/l)</td>
<td>30.5</td>
<td>10.75</td>
<td>4.8 – 10.8</td>
</tr>
<tr>
<td>Neutrophils (× 10⁹/l)</td>
<td>25.73</td>
<td>8.98</td>
<td>1.50 – 6.50</td>
</tr>
<tr>
<td>Lymphocytes (× 10⁹/l)</td>
<td>1.00</td>
<td>1.15</td>
<td>1.20 – 3.40</td>
</tr>
<tr>
<td>Monocytes (× 10⁹/l)</td>
<td>3.76</td>
<td>0.59</td>
<td>0.30 – 0.60</td>
</tr>
<tr>
<td>C-reactive protein (mg/dl)</td>
<td>26.4</td>
<td>3.42</td>
<td>&lt;0.5</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate (mm/h)</td>
<td>65</td>
<td>23</td>
<td>1 – 20</td>
</tr>
<tr>
<td>Glucose (mg/dl)</td>
<td>141</td>
<td>–</td>
<td>70-110</td>
</tr>
<tr>
<td>Rheumatoid factor (KIU/L)</td>
<td>Negative</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>Anti-cyclic citrullinated peptide antibody (RU/ml)</td>
<td>Negative</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td><strong>Cerebrospinal fluid</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Red-cell count (per μl)</td>
<td>Absent</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>White-cell count (per μl)</td>
<td>2</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>Protein (mg/dl)</td>
<td>54</td>
<td>–</td>
<td>10-45</td>
</tr>
<tr>
<td>Glucose (mg/dl)</td>
<td>99</td>
<td>–</td>
<td>40-70</td>
</tr>
<tr>
<td>Gram’s stain</td>
<td>No bacteria seen</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>Viral and bacterial multiplex PCR assay</td>
<td>Negative</td>
<td>–</td>
<td></td>
</tr>
<tr>
<td>CSF Cultures</td>
<td>Negative</td>
<td>–</td>
<td></td>
</tr>
</tbody>
</table>

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References

Ethics approval: the study was performed in accordance with the ethical standards statement.
Informed consent: the patient provided written informed consent.
Figure

Title: Imaging Features of an Acute Attack of Calcium Pyrophosphate Deposition Disease (CPPD) in the Atlantoaxial Joint.

Legend: (A) Axial CT image of the atlantoaxial joint shows curvilinear calcification of the transverse ligament (long arrow). Coronal CT image (B) demonstrates crown-shaped calcium deposits (short arrow) around the odontoid process. Spine-MRI shows inflammatory tissue (arrow) surrounding the odontoid dens characterized by high signal on sagittal fat-suppressed T2-weighted image (C), low signal on sagittal T1-weighted image (D) and enhancement on post-contrast sagittal T1-weighted images (E).
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