Neuromyelitis optica (NMO) diagnosed as syringinx on MRI

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Objective

- To review the original spinal cord MRI interpretations on patients with a subsequent diagnosis of Neuromyelitis Optica (NMO) and assess their clinical course.

Background

- NMO, also known as Devic’s disease, is a severe idiopathic inflammatory demyelinating disease of the CNS that preferentially affects the optic nerves and spinal cord causing attacks of painful lesions (optic neuritis and paralytic transverse myelitis) with a frequent relapsing course [1]. Optic neuritis, transverse myelitis, and a remitting-relapsing course are also common features of multiple sclerosis (MS), which is considered the prototypical inflammatory demyelinating disease.

- Wingerchuk et al. proposed diagnostic criteria for NMO in 1999 to aid in the distinction between NMO and MS. The diagnostic criteria were revised [2][3] following the discovery of serum NMO-IgG, a disease-specific autoantibody targeted against aquaporin-4 (AQP4). These criteria are sensitive and specific for the diagnosis of NMO [2,3].

- Longitudinally extensive transverse myelitis (LETM) refers to an inflammatory demyelinating lesion of the spinal cord that spans three or more vertebral segments on T2-weighted spinal magnetic resonance imaging (MRI) and is classically associated with NMO, but longitudinally extensive spinal lesions can be seen in other causes of non-traumatic transverse myelopathy (Table 4).

- In the acute phase of transverse myelitis (TM), focal demyelination of the spinal cord causes diffuse cord inflammation, edema, and central cavitation. Imaging of the spinal cord in NMO shows a progressive, cavitory, cavitation spanning multiple contiguous spinal cord levels with increased signal intensity and contrast enhancement.[6,7] (Figures 1-4)

- When NMO presents as acute transverse myelitis, prior to optic nerve involvement, LETM with cord edema and enhancement on MRI can be difficult to differentiate from other idiopathic spinal processes and can be misdiagnosed as a syrinx, thus delaying appropriate treatment. Early identification of cavitation in order to initiate appropriate treatment and prevent relapses.

- We clinically noted that 6 out of 25 patients diagnosed with NMO had presented with an original diagnosis of syrinx in our tertiary care center.

Methods

- Chart review of 25 patients with the diagnosis of NMO that were seen at our Tertiary Care Multiple Sclerosis Clinic from 2011 to 2012 and reviewed the initial radiologic interpretation of each case.

- Literature search in PubMed, Medscope and Google Scholar to review the related literature.

- Keywords: Neuromyelitis optica, Devic’s disease, magnetic resonance imaging, syrinx, cavitation, myelitis and aquaporin-4.

Table 1: Data for all study patients

<table>
<thead>
<tr>
<th>Disease</th>
<th>Number (n)</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>NMO</td>
<td>15 (50.0%)</td>
<td></td>
</tr>
<tr>
<td>MS</td>
<td>12 (40.0%)</td>
<td></td>
</tr>
</tbody>
</table>

Table 2: Data for study patients diagnosed with syringinx on MRI

<table>
<thead>
<tr>
<th>Disease</th>
<th>Number (n)</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>NMO</td>
<td>13 (86.7%)</td>
<td></td>
</tr>
<tr>
<td>MS</td>
<td>3 (13.3%)</td>
<td></td>
</tr>
</tbody>
</table>

Table 3: Clinical, laboratory and imaging findings in different causes of Longitudinally Extensive Spinal Cord Lesions

<table>
<thead>
<tr>
<th>Cause of LETM</th>
<th>Clinical Presentation</th>
<th>Neurologic Symptoms</th>
<th>CT/MRI Findings</th>
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<tbody>
<tr>
<td>NMO</td>
<td>Ataxia, diplopia, dysautonomia</td>
<td>Lower extremity weakness</td>
<td>Cauda equina syndrome</td>
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Table 4: Causes of Longitudinally Extensive Spinal Cord Lesions (LETM)

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Conclusion

- There are multiple causes of transverse myelitis with a subsequent spinal cord demyelination causing cord edema and necrosis that produces the characteristic MRI findings of a longitudinally extensive cord lesion (LETM)—involving 3 or more vertebral segments. The detection of a contrast-enhancing lesion on T1-weighted images at least 3 vertebral segments in length is an essential component of a diagnosis of NMO.

- In our study there were 6 of 20 patients that were found to have an original diagnosis of syrinx by radiologic interpretation. In figures 1-4, MRI imaging shows longitudinal extensive lesions with increased signal intensity and central necrosis.

- This appearance of an expansive cavitory lesion spanning multiple contiguous spinal cord levels with increased signal intensity and contrast enhancement can be misdiagnosed as a syrinx (Figure 7).

- All of the patients that were originally diagnosed with a syrinx were subsequently diagnosed with NMO based on their clinical presentation, the imaging features of longitudinally extensive transverse myelitis on MRI, positive serum NMO-IgG antibody and disease course.

- Patients with acute or sub-acute onset of bilateral weakness accompanied by bowel and bladder dysfunction should be reviewed for LETM.

- A radiologic assessment of syrinx should be considered highly suspicious for LETM and the subsequent development of NMO.

References