Recurrent Optic Neuritis in Children

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Background

• Recurrent episodes of optic neuritis (ON) occur in children without MS or NMO.
• There is very little literature on this group.
• Presentation characteristics, long-term prognosis and the best treatment plan are unknown.

Objectives

• To describe the clinical characteristics of pediatric patients with recurrent optic neuritis.

Methods

• A retrospective analysis of prospectively collected data from May 2011 to April 2014 on all patients in the Pediatric MS and Other Demyelinating Diseases Database.
• Inclusion criteria:
  • <18 yr at onset of first demyelinating attack
  • More than one attack of optic neuritis
• Exclusion criteria:
  • Diagnosis of MS or NMO.

Results

• 20 patients (8 girls and 12 boys) met inclusion criteria and were followed for a mean duration of 13.5 months.
• 9 patients had only idiopathic attacks of ON without other neurologic symptoms.
• 3 had ADEM and isolated ON (ADEM-ON)1
• 3 did not fit either of these classifications.
• Although 37 (66%) of the attacks were unilateral, over the follow-up period, 14 (70%) patients had at least 1 attack affecting both optic nerves.

<table>
<thead>
<tr>
<th>Attack Characteristics</th>
<th>Idiopathic recurrent ON (n = 9)</th>
<th>ADEM-ON (n = 8)</th>
<th>Other (n = 3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lowest visual acuity (median and range)</td>
<td>20/200 (20/20, NLP)</td>
<td>20/40 - 20/50 (20/20, LP)</td>
<td>Finger counting (20/400, LP)</td>
</tr>
<tr>
<td>Abnormal visual field</td>
<td>1/3 (constricted, central scotoma)</td>
<td>1/4 (constricted)</td>
<td>1/3 (varied)</td>
</tr>
<tr>
<td>Papillitis</td>
<td>4/5</td>
<td>0/5</td>
<td>2/3</td>
</tr>
<tr>
<td>Pain with eye movement</td>
<td>6/9</td>
<td>4/7</td>
<td>2/3</td>
</tr>
</tbody>
</table>

Diagnostic Studies

• Proportion of orbit MRIs with T2 lesion and/or enhancement | 11/16 (69%) | 9/14 (64%) | 12/12 (100%) |
• Proportion of orbit MRIs with optic nerve enlargement | 9/17 (53%) | 4/9 (44%) | 8/11 (73%) |
• Brain lesions on any MRI | 5/8 (63%) | 8/8 (100%) | 2/3 (67%) |
• Spine lesions on any MRI | 1/8 (13%) | 2/8 (25%) | 1/3 (33%) |
• CSF pleocytosis | 5/6 (83%) | 8/8 (100%) | 2/3 (67%) |
• CSF oligoclonal bands | 1/7 (14%) | 1/6 (17%) | 0/3 (0%) |
• IgG index elevation | 0/5 (0%) | 0/6 (0%) | 1/3 (33%) |

Treatment and Outcome

• Treated with steroids at any time | 8/9 (89%) | 6/8 (75%) | 3/3 (100%)
• Treated with DMT at any time | 4/9 (44%) | 3/8 (38%) | 2/3 (67%)
• Optic disc pallor at follow up | 5/9 (56%) | 5/8 (63%) | 2/3 (67%)
• 6 months attack free at last follow-up | 7/8 (88%) | 7/8 (88%) | 1/2 (50%)
• 20/20 vision at follow-up | 5/9 (56%) | 7/8 (88%) | 2/3 (67%)

* Treatments included azathioprine, mycophenolate and glatiramer acetate in the Recurrent ON group, Interferon beta 1a, fingolimod, and glatiramer acetate in the ADEM-ON group, IVIG and mycophenolate in the Other group.

LP = light perception only, NLP = no light perception, DMT = disease modifying therapy

Discussion & Conclusions

• Of children with recurrent ON who do not meet diagnostic criteria for MS or NMO, many have a disease course consistent with idiopathic recurrent ON or ADEM-ON.
• Inter-attack interval can be very long, similar to outcomes associated with our ADEM-ON group, suggesting that long term monitoring is indicated.
• Many of the MRI abnormalities in the Idiopathic group included punctate and non-specific abnormalities.
• Cortical and diffuse MRI abnormalities were seen in some “Other” patients, such as 1 patient ultimately diagnosed with CNS vasculitis.
• In the majority of cases, regardless of etiology, visual outcomes were good with at least 70% of eyes recovering to baseline 20/20 vision. This is similar to outcomes associated with MS-ON.2
• Some children with idiopathic recurrent ON may have Chronic Relapsing Inflammatory Optic Neuropathy (CRION), as described in adults.3

References


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