isolated cervical arthritis as the sole manifestation of familial Mediterranean fever: A case report

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Background: Familial Mediterranean Fever (FMF) is an autosomal recessive disease affecting mainly eastern Mediterranean populations. Fever and abdominal pain are the 2 most prevalent features. The most common arthritis manifestation of FMF is acute self-limiting monoarthritis. 5% of FMF patients develop chronic erosive arthritis. FMF mutation M694V has been associated to an increased risk of spondylarthritis.

Objectives: We report the case of a child with cervical spine inflammation as the sole presentation of FMF.

Methods: A boy born to non-consanguineous parents presented at the age of 4 with complete blockage of his neck. He was described to have experienced protracted limp at the age of 1 with complete spontaneous remission, and possible recurrent fever in the first 2 years of life. Family history is negative. His physical exam revealed a painful and completely blocked neck in all movements, and stumpy fingers. He had failure to thrive and a big belly without hepatosplenomegaly. Cognitive development was normal.

Results: Given this atypical isolated inflammation of the cervical spine, genetic testing for FMF was conducted. We identified 2 typical mutations (M694V and M694I) confirming the diagnosis of FMF. Colchicine treatment was started.

Conclusions: To the best of our knowledge, this is the first report of FMF masquerading as neck arthritis. Early spondylarthropathy is a possibility, but this unusual neck inflammation might be an isolated arthritis associated to FMF. Based on this clinical presentation, in the setting of atypical arthritis, diagnosis of FMF is to be raised in at-risk ethnicities, even in the absence of familial history and common clinical signs.

REFERENCES:

Disclosure of Interest: None declared

AB1118

PROCALCITONIN DIFFERENTIATES INFECTION FROM ACTIVE DISEASE IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS (JIA)


Background: Patients with JIA often present with signs and symptoms suggestive of infection. However, differentiation of infections from non-infectious presentation in routine clinical care is challenging. Procalcitonin (PCT) is a serum biomarker elevated in the setting of bacterial infection, but whether it can reliably differentiate infection from disease flare in patients with JIA is unknown.

Objectives: To test the hypothesis that PCT levels will differ between active JIA, quiescent JIA, bacteremic patients and healthy controls.

Abstract AB1118 – Table 1. Patient Characteristics

<table>
<thead>
<tr>
<th>Age (median, IQ range)</th>
<th>Male Gender</th>
<th>Race</th>
<th>Caucasian/ White</th>
<th>AA/Black</th>
<th>Asian</th>
<th>Other Hispanic</th>
<th>Ethnicity</th>
<th>Private Insurance</th>
</tr>
</thead>
<tbody>
<tr>
<td>9.0 (2.4–12.8)</td>
<td>5 (41.7%)</td>
<td>7 (58.3%)</td>
<td>1 (8.3%)</td>
<td>3 (25.0%)</td>
<td>7 (58.3%)</td>
<td>3 (20.0%)</td>
<td>1 (8.3%)</td>
<td>7 (58.3%)</td>
</tr>
<tr>
<td>14.5 (9.9–17.4)</td>
<td>3 (20.0%)</td>
<td>14 (93.3%)</td>
<td>0 (0.0%)</td>
<td>0 (0.0%)</td>
<td>14 (93.3%)</td>
<td>6 (37.5%)</td>
<td>0 (0.0%)</td>
<td>14 (93.3%)</td>
</tr>
<tr>
<td>14.4 (13.9–15.5)</td>
<td>6 (37.5%)</td>
<td>12 (75.0%)</td>
<td>3 (18.8%)</td>
<td>1 (6.3%)</td>
<td>12 (75.0%)</td>
<td>3 (20.0%)</td>
<td>1 (6.3%)</td>
<td>12 (75.0%)</td>
</tr>
<tr>
<td>1.1 (0.8–1.8)</td>
<td>3 (20.0%)</td>
<td>2 (50.0%)</td>
<td>3 (20.0%)</td>
<td>2 (50.0%)</td>
<td>2 (50.0%)</td>
<td>1 (6.3%)</td>
<td>2 (50.0%)</td>
<td>2 (50.0%)</td>
</tr>
</tbody>
</table>

Abstract AB1118 – Table 2. Laboratory Data

<table>
<thead>
<tr>
<th>WBC (median, IQR)</th>
<th>ESR</th>
<th>Normal-10 (median, IQR)</th>
<th>CRP</th>
<th>Normal-1 (median, IQR)</th>
<th>PCT</th>
<th>Bacteremic patients (median, IQR)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>8.9 (4.4–14.4)</td>
<td>6.0</td>
<td>[4.0–40.0]</td>
<td>0.27</td>
<td>[0.12–6.48]</td>
<td>0.00</td>
<td>[0.00–0.00]</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>7.7 (1.6–16.7)</td>
<td>8.0</td>
<td>[5.0–10.0]</td>
<td>0.31</td>
<td>[0.12–1.85]</td>
<td>0.00</td>
<td>[0.00–0.00]</td>
<td>0.067</td>
</tr>
<tr>
<td>6.7 (1.7–14.1)</td>
<td>8.0</td>
<td>[5.0–10.0]</td>
<td>0.44</td>
<td>[0.12–1.85]</td>
<td>0.00</td>
<td>[0.00–0.00]</td>
<td>0.067</td>
</tr>
<tr>
<td>13.1 (12.1–12.1)</td>
<td>43.0</td>
<td>[20.0–66.0]</td>
<td>16.63</td>
<td>[7.76–25.68]</td>
<td>5.78</td>
<td>[4.26–52.00]</td>
<td>0.067</td>
</tr>
</tbody>
</table>

Methods: From 10/16–4/17, consecutive children 6 months – 18 years with a) active untreated JIA b) quiescent JIA and c) healthy pre-surgical candidates were recruited from a musculoskeletal specialty hospital. JIA was defined according to ILAR criteria. Patients with active JIA despite treatment were excluded, to avoid

Disclosure of Interest: None declared

Abstract AB1117 – Figure 1

Conclusions: To the best of our knowledge, this is the first report of FMF masquerading as neck arthritis. Early spondylarthropathy is a possibility, but this unusual neck inflammation might be an isolated arthritis associated to FMF. Based on this clinical presentation, in the setting of atypical arthritis, diagnosis of FMF is to be raised in at-risk ethnicities, even in the absence of familial history and common clinical signs.

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