Temporomandibular Joint Arthritis in Juvenile Dermatomyositis and Pediatric Mixed Connective Tissue Disease

Peter Weiser¹, Stephen Johnson², Robert M. Lowe¹, Randy Q. Cron¹

¹ UAB Department of Pediatrics, Division of Rheumatology, Birmingham, AL; ²The Ohio State University, Columbus, OH, USA

Corresponding Author: Peter Weiser, UAB Department of Pediatrics, Division of Rheumatology, Birmingham, AL, USA
e-mail: pweiser@peds.uab.edu

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Abstract

Inflammation of the temporomandibular joint (TMJ) is a frequent finding in juvenile idiopathic arthritis and can lead to facial deformity. Our retrospective cohort review aimed to address presence of similar TMJ arthritis in myositis associated connective tissue disorders.

Electronic health records of patients at the Children's of Alabama, Birmingham, AL, USA with TMJ arthritis on contrasted MRI examination and diagnosed with juvenile dermatomyositis (JDM) or mixed connective tissue disease (MCTD) between January 2010 to March 2012 were reviewed for subjective and objective findings of TMJ involvement. During that period, four patients with JDM and three patients with MCTD were found to have imaging proof of TMJ arthritis. Two of the four JDM patients and two of the three MCTD patients did not report pain at rest nor with mastication and only one of the JDM patients with pain had jaw deviation on mouth opening. Peripheral arthritis was present in the two JDM patients with TMJ pain and in all MCTD patients. The two JDM patients with no TMJ pain nor deviation nor peripheral joint involvement had even low disease activity score as per physician global assessment score.

Children with myositis associated connective tissue disease can also have concurrent TMJ arthritis and as in JIA it can have a silent progression. Peripheral arthritis seems to be associated with TMJ arthritis, but larger studies are needed for incidence, prevalence and possible progression evaluation and for indications of therapy. We suggest that evaluation of the TMJ should be part of the diagnostic work-up for pediatric arthritis-associated autoimmune connective tissue diseases with or without active peripheral arthritis.

Key words: Kawasaki disease, echocardiography, coronary artery involvement

Introduction

The temporomandibular joint (TMJ) is frequently affected in juvenile idiopathic arthritis (JIA). Depending on cohorts, up to 80 percent of JIA patients with peripheral arthritis have been shown to have synovitis of the TMJ on MRI [1]. It has also been found in pediatric Sjögren disease and sarcoidosis [2], and our report is focusing on TMJ arthritis in myositis-associated pediatric rheumatologic diseases which also share peripheral ar-
Arthritis as a common feature.

Juvenile dermatomyositis (JDM) is the most common chronic childhood inflammatory myopathy [3]. It primarily manifests as weakness of the proximal muscle groups, heliotrope rash, and Gottron papules, while one third of patients present with arthritis as well [4]. Mixed connective tissue disease (MCTD) is an autoimmune condition heralded by vasculopathy leading to Raynaud syndrome, myositis, erosive arthritis, and potentially multiorgan involvement.

Based on our experience with JIA, maximal interincisal opening (MIO) measurements have been part of the standard physical exam when patients present with peripheral arthritis. In addition, subjective complaints of jaw pain or objective mandibular asymmetry in any referred patient would result in MIO assessment. Considering the possible devastating outcomes of TMJ arthritis in growing children (e.g., micrognathia), further imaging assessment by MRI of the TMJ with and without intravenous contrast is pursued if the mouth opening is <40 mm, a midline incisor deviation is present on the opening, there is tenderness in the TMJ area on examination, or there is asymmetry in the appearance of the lower jaw.

An Institutional Review Board-approved retrospective review of electronic medical records at the Children’s of Alabama Rheumatology Clinic (Birmingham, Alabama) from January 2010 to March 2012 identified four JDM and three pediatric MCTD patients who had MRI-proven arthritis of the TMJ (i.e., synovial enhancement after intravenous contrast administration with or without erosions). Patients with peripheral arthritis both in JDM and MCTD showed TMJ synovitis on MRI (Figure 1), while two of the JDM patients did not have any peripheral joint symptoms or findings on physical examination. Patient number 4 with JDM was found to have previously not commented TMJ synovitis during a retrospective review of her brain MRI (obtained 10 months prior for headaches and vision difficulties), which had been triggered by a newly noted small mouth opening. Patient number 5 with JDM was screened based on the findings of the prior patient. Neither child had any complaints regarding their jaw. The primary demographics, clinical characteristics, and imaging results of the cohort can be found in Table 1.

There are several limitations of our cohort review, including a small number of patients and lack of widely accepted MIO values when selecting patients for further imaging evaluation. Nevertheless, our cohort points out that TMJ arthritis

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>TMJ Pain</th>
<th>Deviation mouth opening</th>
<th>MIO (mm)</th>
<th>Peripheral arthritis</th>
<th>Physician global assessment score (0-100)</th>
<th>TMJ arthritis by MRI</th>
<th>TMJ condylar erosions</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>15</td>
<td>F</td>
<td>MCTD</td>
<td>Yes</td>
<td>Yes</td>
<td>32</td>
<td>Yes</td>
<td>45</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>2</td>
<td>16</td>
<td>F</td>
<td>MCTD</td>
<td>No</td>
<td>Yes</td>
<td>36</td>
<td>Yes</td>
<td>45</td>
<td>Yes</td>
<td>Yes</td>
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<tr>
<td>3</td>
<td>12</td>
<td>F</td>
<td>MCTD</td>
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<td>No</td>
<td>48</td>
<td>Yes</td>
<td>20</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>F</td>
<td>JDM</td>
<td>No</td>
<td>No</td>
<td>30</td>
<td>No</td>
<td>10</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>1.5</td>
<td>F</td>
<td>JDM</td>
<td>No</td>
<td>No</td>
<td>31</td>
<td>No</td>
<td>10</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>6</td>
<td>10</td>
<td>F</td>
<td>JDM</td>
<td>Yes</td>
<td>No</td>
<td>46</td>
<td>Yes</td>
<td>18</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>7</td>
<td>5</td>
<td>M</td>
<td>JDM</td>
<td>Yes</td>
<td>Yes</td>
<td>18.5</td>
<td>Yes</td>
<td>46</td>
<td>Yes</td>
<td>No</td>
</tr>
</tbody>
</table>
can be found in patients who present predominantly with myositis, especially in those with peripheral arthritis. Considering the lack of reliable symptoms or signs on physical examination, TMJ MRI with and without contrast is both highly sensitive in identifying affected individuals prior to radiographic findings of bony damage [5], and highly specific [6] in identifying TMJ arthritis.

In the past, masticatory abnormalities in adults with MCTD have been described [7], and erosions on panoramic radiographs were shown in 19% (3/16). JDM patients’ reduced MIO has also been reported, but rather attributed to muscle weakness [8]. To our knowledge, there is only one case of pediatric MCTD with TMJ-proven arthritis in the medical literature [9], whereas our report is the first to document synovitis of the TMJ in children — not only with MCTD, but also with JDM.

The implications of TMJ arthritis in a child with JDM or MCTD are several-fold, just like in JIA. Joint destruction can affect facial proportions, bite and mastication. Even non-erosive TMJ arthritis with chronic pain can lead to decreased use and thus to improper bone development and remodeling during growth. As seen in JIA, TMJ arthritis can progress despite systemic therapy. Two of the seven patients in our cohort had TMJ condylar head erosions by MRI (Table 1). Since condylar resorption can lead to mandibular asymmetry and subsequently to facial deformity, based on data from JIA cohorts, intraarticular steroid injection might be suggested [10]. In JDM, concomitant systemic therapy with high-dose steroids could possibly effect TMJ disease progression, but MCTD patients are well known to have erosive peripheral arthritis despite aggressive therapy and therefore might benefit from intraarticular TMJ steroid injection, just like in JIA. Future clinical trials will likely be helpful in addressing the proper management.

In summary, TMJ arthritis is under-recognized, not only in JIA, but also likely in JDM and in pediatric MCTD. The latter patients with peripheral arthritis and/or decreased MIO, with or without deviation on the mouth opening, should undergo contrast-enhanced TMJ MRI evaluation to assess arthritis. The use of long-acting intraarticular corticosteroid injection could be considered when there is TMJ disease progression despite systemic therapy.

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**References**
