Teaching NeuroImages: Immune Checkpoint Inhibitor–Related Fasciitis and Myositis With Perifascicular Atrophy

Author(s):
Timothy Richard Fullam, M.D.¹; Nathan McGraw, M.D.¹; Matthew Grainger¹; Mazen M. Dimachkie, M.D.¹; Swathy Chandrashekhar, M.D.¹

Corresponding Author:
Timothy Richard Fullam
trfullam23@gmail.com

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Affiliation Information for All Authors: 1. The University of Kansas Medical Center, Kansas City, KS

Contributions:
Timothy Richard Fullam: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data
Nathan McGraw: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data
Matthew Grainger: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data
Mazen M. Dimachkie: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data
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Case Summary:

58-year-old man with melanoma treated with ipilimumab/nivolumab presented with pain, limited joint mobility and proximal weakness without oculobulbar weakness, dyspnea, or rash. EMG demonstrated positive sharp waves, fibrillations and myopathic units. Extremity MRI showed diffuse fascial and mild muscle enhancement consistent with fasciitis/mild myositis (figure). Creatinine kinase was normal. PET-CT demonstrated diffusely FDG-avid lymph nodes and muscles; lymph node biopsy revealed granulomatous inflammation suggesting ICI-related inflammatory reaction. Biceps biopsy demonstrated perifascicular atrophy (PA) and fascial/perimysial perivascular inflammation (figure).

Histopathologic findings of ICI-related myositis initially included necrosis, macrophagy and endomysial inflammation. The spectrum has expanded to include perimysial perivascular inflammation and PA tendency. ¹,²

Teaching Slides - http://links.lww.com/WNL/B481

References:


2) Touat M, Maisonobe T, Knauss S, et. al. Immune checkpoint inhibitor-related myositis and myocarditis in patients with cancer. Neurology Sep 2018, 91 (10) e985-e994; DOI: 10.1212/WNL.0000000000006124

Figure: Immune Checkpoint Inhibitor Related Fasciitis and Myositis with Perifascicular Atrophy

Perifascicular atrophy noted on H&E (A), ATPase (B), and NADH (C) stains (arrow) with fascial, perimysial, and perivascular inflammatory infiltrates on H&E (A, arrowhead) and Acid Phosphatase (D,E, arrowheads). Pre- and post-contrast MRI of the lower extremities (F.a, F.b) and upper extremities (F.c, F.d) with fascial and muscle enhancement.
Appendix 1. Authors

<table>
<thead>
<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Timothy Fullam, M.D.</td>
<td>Department of Neurology, The University of Kansas Medical Center</td>
<td>Analysis of biopsy, review of neuroimaging, drafting of manuscript</td>
</tr>
<tr>
<td>Nathan McGraw, M.D.</td>
<td>Department of Neurology, The University of Kansas Medical Center</td>
<td>Analysis of biopsy, critical review of manuscript</td>
</tr>
<tr>
<td>Matthew Grainger</td>
<td>Department of Neurology, The University of Kansas Medical Center</td>
<td>Preparation and processing of muscle biopsy, critical review of manuscript</td>
</tr>
<tr>
<td>Mazen M. Dimachkie, M.D.</td>
<td>Department of Neurology, The University of Kansas Medical Center</td>
<td>Analysis of biopsy, critical review of manuscript</td>
</tr>
<tr>
<td>Swathy Chandrashekhar, M.D.</td>
<td>Department of Neurology, The University of Kansas Medical Center</td>
<td>Patient management, analysis of biopsy, critical review of manuscript</td>
</tr>
</tbody>
</table>
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