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

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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
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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.¹²

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

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<thead>
<tr>
<th>Name</th>
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<tbody>
<tr>
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