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

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Contributions:

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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}

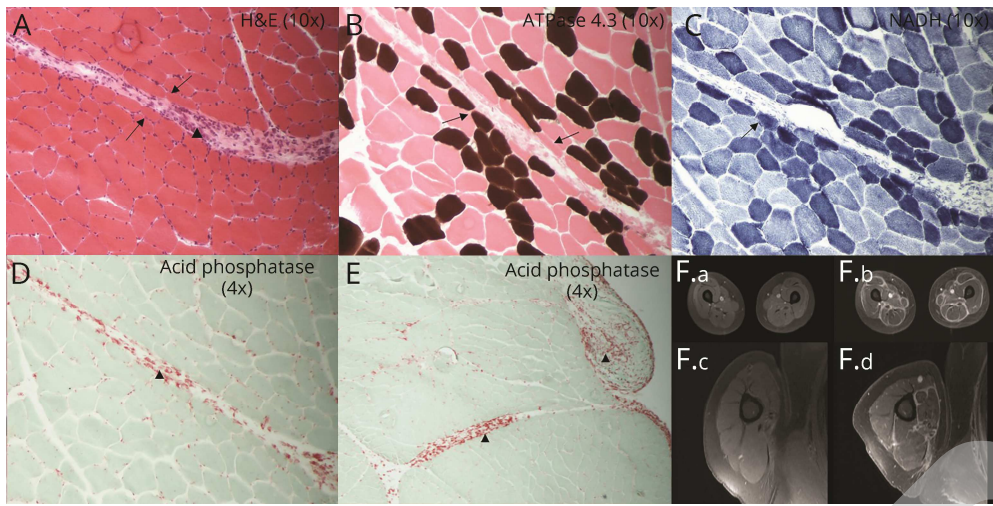
Teaching Slides - <http://links.lww.com/WNL/B481>

References:

- 1) Matas-García A, Milisenda JC, Selva-O'Callaghan A, et. al. Emerging PD-1 and PD-1L inhibitors-associated myopathy with a characteristic histopathological pattern. *Autoimmun Rev.* 2020 Feb;19(2):102455. doi: 10.1016/j.autrev.2019.102455. Epub 2019 Dec 12. PMID: 31838162
- 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

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

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