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

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Figure Immune Checkpoint Inhibitor–Related Fasciitis and Myositis With Perifascicular Atrophy

Perifascicular atrophy noted on H&E (A), ATPase (B), and nicotinamide adenine dinucleotide (C) stains (arrow) with fascial, perimysial, and perivascular inflammatory infiltrates on H&E (A, arrowhead) and acid phosphatase (D, E, arrowheads). Precontrast and postcontrast MRI of the lower extremities (F.a, F.b) and upper extremities (F.c, F.d) with fascial and muscle enhancement.

Case Summary

A 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 fluorodeoxyglucose-avid lymph nodes and muscles; lymph node biopsy revealed granulomatous inflammation suggesting immune checkpoint inhibitor-related inflammatory reaction. Biceps biopsy demonstrated perifascicular atrophy (PA) and fascial/perimysial perivascular inflammation (Figure).

Histopathologic findings of immune checkpoint inhibitor-related myositis initially included necrosis, macrophagy, and endomysial inflammation. The spectrum has expanded to include perimysial perivascular inflammation and PA tendency.^{1,2}

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Appendix Authors

Name	Location	Contribution
Timothy Fullam, MD	Department of Neurology, The University of Kansas Medical Center, Kansas City	Analysis of biopsy, review of neuroimaging, and drafting the manuscript

Appendix (continued)

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

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