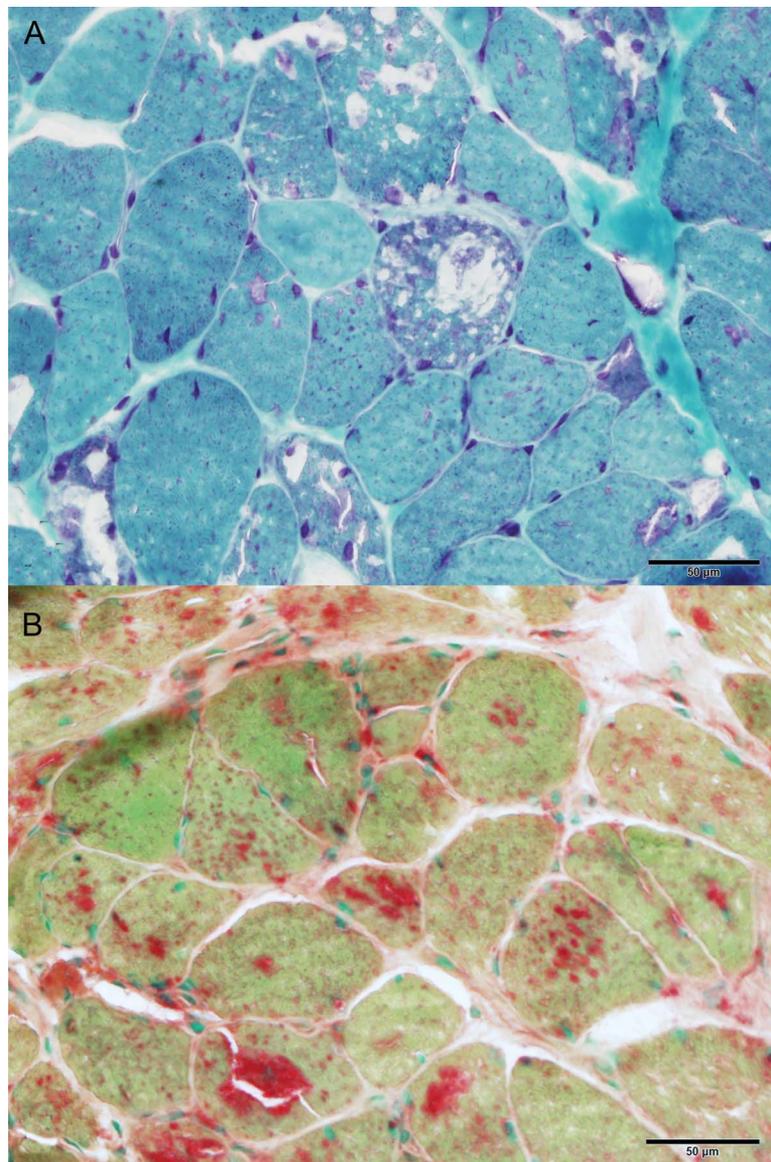


Teaching NeuroImages: Hydroxychloroquine-induced vacuolar myopathy

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Figure Frozen sections of the quadriceps muscle



(A) The trichrome stain shows numerous fibers harboring single or multiple vacuoles with or without granular material. (B) Nearly all vacuoles are strongly acid-phosphatase-positive. Bars = 50 µm.

A 58-year-old woman with long-standing mixed connective tissue disorder had proximal leg weakness for 4 months. She had been treated with 400 mg/day of hydroxychloroquine and varying doses of prednisone over 15 years. Creatine kinase was 600 U/mL. MRI of

quadriceps showed edema and its biopsy revealed myriad acid-phosphatase-positive autophagic vacuoles indicating increased lysosomal activity (figure). Hydroxychloroquine induces autophagy by reducing lysosomal acidity.¹ Autophagic vacuolar myopathy can

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be seen with chloroquine or colchicine therapy or in inherited disorders (α -glucosidase deficiency, Danon disease, and X-linked myopathy with excessive autophagy).¹ Hydroxychloroquine myopathy usually presents with mild to moderate proximal weakness and rarely causes severe weakness and respiratory failure.² Our patient's weakness improved after discontinuing hydroxychloroquine.

AUTHOR CONTRIBUTIONS

Partha S. Ghosh: drafting/revising the manuscript, study concept or design, analysis or interpretation of data. David Swift: analysis or interpretation of data. Andrew G. Engel: drafting/revising the manuscript, analysis or interpretation of data, study supervision.

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