Teaching NeuroImages: Myeloperoxidase–anti-neutrophil cytoplasmic antibody–positive hypertrophic pachymeningitis

A 49-year-old woman with chronic epistaxis presented with painless left monocular vision loss. Notable findings on examination of the left eye included visual acuity of 20/200 and relative afferent pupillary defect. MRI of the brain revealed enhancement of the left optic nerve sheath and diffuse dural thickening (figure). Laboratory workup yielded only lymphocytic pleocytosis (12 white blood cells) and positive serologies for perinuclear anti-neutrophil cytoplasmic antibodies (ANCA) and anti-myeloperoxidase (MPO) antibodies. Dural biopsy showed multifocal dense lymphoplasmacytic infiltration with vasculitis and reactive fibroplasia. A diagnosis of MPO-ANCA-positive hypertrophic pachymeningitis was made. This phenotypic variant of granulomatosis with polyangiitis is typically restricted to the CNS and upper airway; treatment involves immunosuppression. The patient improved clinically and radiographically with prednisone and rituximab.

AUTHOR CONTRIBUTIONS

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REFERENCE
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Collin J. Culbertson, Seth C. Lummus and Carl A. Gold
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