RESIDENT & FELLOW SECTION

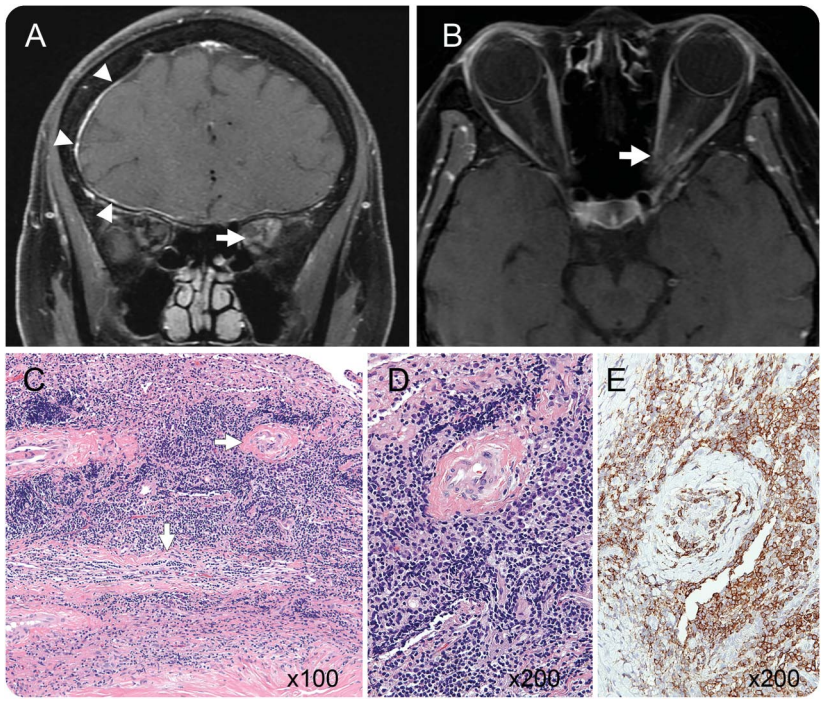
Section Editor
John J. Millichap, MD

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

Collin J. Culbertson, MD
Seth C. Lummus, DO, MS
Carl A. Gold, MD, MS

Correspondence to Dr. Culbertson: collinc@stanford.edu

Figure MRI and histology of the dural biopsy



(A, B) T1-weighted postgadolinium sequences reveal diffuse pachymeningeal enhancement (arrowheads) and asymmetric enhancement of the left optic nerve sheath (arrows). (C, D) Standard hematoxylin & eosin sections of the dural biopsy show numerous lymphocytes and plasma cells infiltrating thick-walled blood vessels (arrows). (E) Cells are confirmed to be lymphoid with CD45 immunohistochemistry.

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.[1] The patient improved clinically and radiographically with prednisone and rituximab.

AUTHOR CONTRIBUTIONS

Collin J. Culbertson: literature review, manuscript preparation, radiology images. Seth C. Lummus: pathology images. Carl A. Gold: manuscript preparation and editing.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

REFERENCE

1. Yokoseki A, Saji E, Arakawa M, et al. Hypertrophic pachymeningitis: significance of myeloperoxidase anti-neutrophil cytoplasmic antibody. Brain 2014;137:520–536.

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From the Departments of Neurology & Neurological Sciences (C.J.C., C.A.G.) and Pathology (S.C.L.), Stanford University, CA.