

Teaching NeuroImages: Idiopathic hypertrophic pachymeningitis

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Figure 1 Coronal brain MRI



Coronal T1-weighted MRIs show smooth and diffuse pachymeningeal enhancement with involvement of the bilateral optic canals (arrows).

A 39-year-old woman presented with acute, painless left monocular vision loss in the context of 6 months of right peripheral facial weakness and prior right optic neuropathy. MRI showed asymmetric, bilateral smooth pachymeningeal enhancement with involvement of the optic canals (figure 1). Autoimmune, inflammatory, and neoplastic testing including CSF and serum immunoglobulin G4 levels were unremarkable. Dural biopsy revealed chronic lymphohistiocytic pachymeningitis without granulomatous

inflammation (figure 2), consistent with idiopathic hypertrophic pachymeningitis (IHP). Clinical and radiographic responses were achieved with steroids and methotrexate. IHP is a diagnosis of exclusion characterized by headache and cranial neuropathies.¹ MRI T1-weighted images show smooth, thickened, enhancing pachymeninges.²

AUTHOR CONTRIBUTIONS

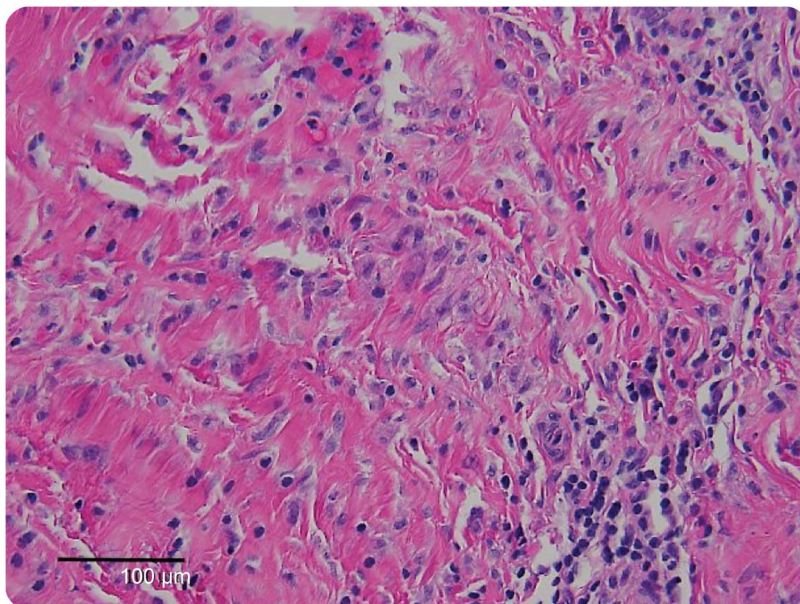
Andrea Wasilewski: article concept, acquisition of data, manuscript drafting. Lawrence Samkoff: manuscript drafting.

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Figure 2 Pathologic slide of dural biopsy



Hematoxylin & eosin-stained pathologic slide of dural biopsy shows chronic lymphohistiocytic pachymeningitis without granulomatous inflammation or necrosis.

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DISCLOSURE

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

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