Teaching NeuroImages: Idiopathic hypertrophic pachymeningitis

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. MRI T1-contrasted images show smooth, thickened, enhancing pachymeninges.

AUTHOR CONTRIBUTIONS
Andrea Wasilewski: article concept, acquisition of data, manuscript draftsing. Lawrence Samkoff: manuscript drafting.
Hematoxylin & eosin-stained pathologic slide of dural biopsy shows chronic lymphohistiocytic pachymeningitis without granulomatous inflammation or necrosis.

REFERENCES

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