A 46-year-old man with progressive visual loss underwent brain MRI showing multifocal nodular pachymeningeal thickening involving optic nerve meninges bilaterally and internal acoustic meatus dura mater (figure 1). Diffuse meningiomatosis was diagnosed and radiation therapy was given with symptom stabilization. Four years later, left hearing loss and right hypoacusia occurred, with slight transient improvement after high-dose dexamethasone. Serum immunoglobulin (Ig)G4 was increased. CSF analysis showed increased protein, oligoclonal IgG, plasma cells, and lymphocytes. Cerebral biopsy showed meningeal plasma-cell granuloma with IgG4-positive polyclonal plasma cells and B-lymphocyte infiltration (figure 2). IgG4-related disease was diagnosed. Rituximab was unsuccessful. IgG4-related disease is a fibroinflammatory, multiorgan condition characterized by tumefactive lesions and lymphoplasmacytic infiltrates rich in IgG4-positive
plasma cells that may affect every organ; serum IgG4 may be elevated. Although IgG4-related disease with exclusive multifocal CNS localization is extremely rare and often misdiagnosed, it should be considered in the differential diagnosis of tumor-like intracranial lesions and hyperthrophic pachymeningitis.

STUDY FUNDING
No targeted funding reported.

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

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Teaching NeuroImages: Multifocal neurologic involvement as the only manifestation of IgG4-related disease
Silvia Imbergamo, Marta Campagnolo, Renzo Manara, et al. Neurology 2013;80:e40-e41
DOI 10.1212/WNL.0b013e31827f08ae

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