

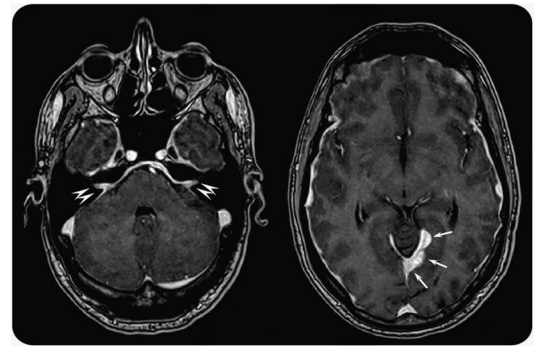
Teaching NeuroImages: Multifocal neurologic involvement as the only manifestation of IgG4-related disease

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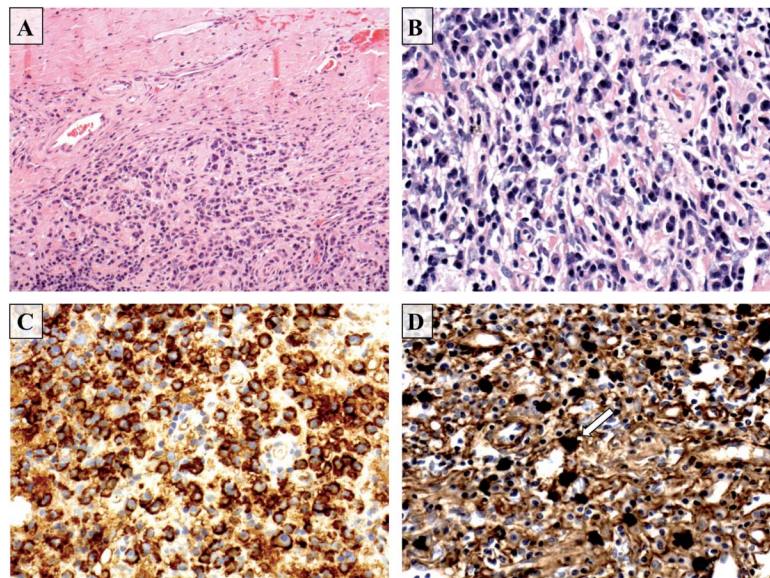
A 46-year-old man with progressive visual loss underwent brain MRI showing multifocal nodular pachymeningeal thickening involving optic nerve meninges bilaterally and internal acoustic meatus dura mater (figure 1). Diffuse meningiomas was diagnosed and radiation therapy was given with symptom stabilization. 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.¹ Rituximab was unsuccessful. IgG4-related disease is a fibroinflammatory, multiorgan condition characterized by tumefactive lesions and lymphoplasmacytic infiltrates rich in IgG4-positive

Figure 1 Brain MRI



Contrast-enhanced T1-weighted axial images showing diffuse dural thickening with internal acoustic meatus involvement (arrowheads) and with nodular appearance, more evident at the tentorium level (arrows).

Figure 2 Histopathologic findings



(A) Section of meninges with plasma cell infiltration, hematoxylin & eosin (H&E) 200 \times . (B) A stronger magnification display plasma cells with no atypical features, H&E 400 \times . (C) Positive plasma cells, CD138 staining, 400 \times . (D) Positive plasma cells, immunoglobulin G4 staining (see arrow), 400 \times .

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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,²⁻⁴ it should be considered in the differential diagnosis of tumor-like intracranial lesions and hyperthrophic pachymeningitis.

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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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