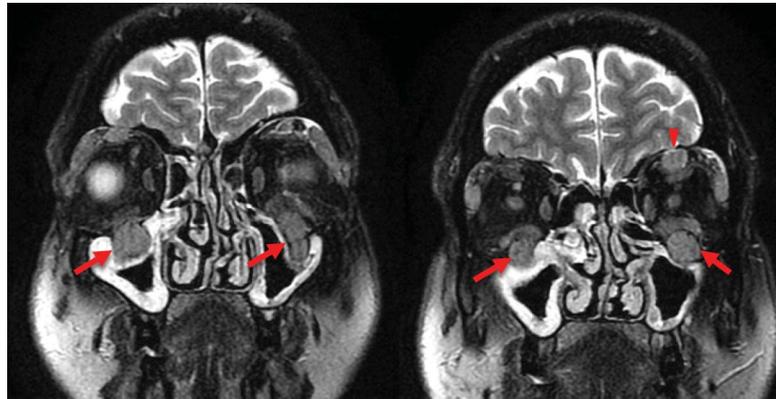


Teaching NeuroImages: IgG4-related orbital disease and enlargement of the trigeminal nerve branches

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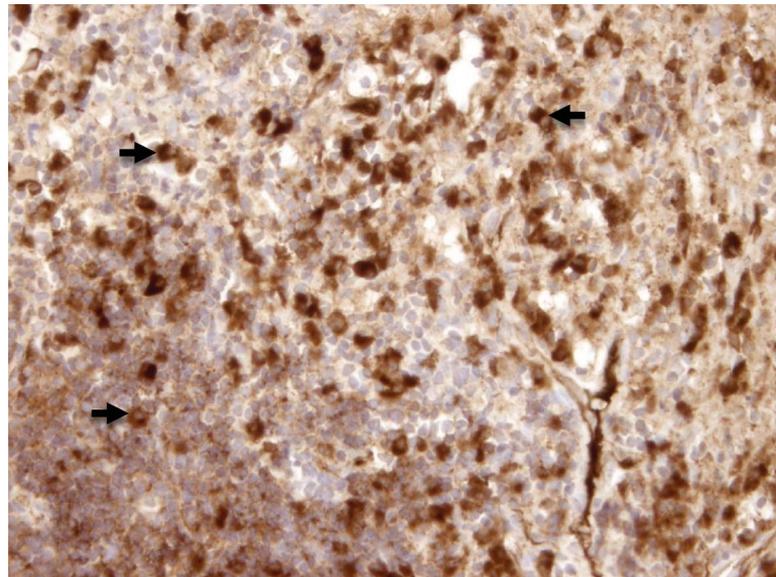
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Figure 1 Brain MRI findings



STIR fat-saturated T2-weighted coronal MRI demonstrates thickening of bilateral infraorbital (arrow; adjacent to the enlarged inferior rectus) and left frontal (arrowhead; near the swollen left superior rectus and levator complex) nerves.

Figure 2 Pathologic findings of the biopsy from the left infraorbital canal



Immunoglobulin G 4 immunohistochemistry shows numerous plasma cells stained positive (arrows) (immunoperoxidase, original magnification $\times 200$).

A 54-year-old man had a 5-year history of painless bilateral eyelid swelling, proptosis, and diplopia. MRI showed enlargement of extraocular muscles, lacrimal gland, and divisions of the trigeminal nerve (figure 1). Biopsy from

the left infraorbital canal demonstrated lymphoplasmacytic infiltrate and fibrosis. Mean number of immunoglobulin G (IgG)4+ cells was 155 per high-power field and ratio of IgG4+/IgG+ cells was 89% (figure 2).

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Serum IgG4 concentration was 878 mg/dL (normal range 4–86 mg/dL). Symptoms resolved with steroids. The patient relapsed during tapering and azathioprine was added. IgG4-related disease is characterized by IgG4-positive cells and lymphocyte infiltration into various organs, including orbital tissues. Thickening of branches of the trigeminal nerve is highly suggestive of IgG4-related disease.^{1,2}

AUTHOR CONTRIBUTIONS

Dr. R. Deschamps: corresponding author; drafting/revising the manuscript for content, including medical writing for content. Drs. L. Deschamps, Vignal, Putterman, Galatoire, and Gout: drafting/revising the manuscript for content, including medical writing for content.

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DISCLOSURE

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