

**Sir,
Infraorbital nerve enlargement due to IgG4-related disease**

IgG4-related disease is a recently defined condition which may result in multi-organ involvement.¹ Cranial nerve involvement has rarely been reported. We describe a case of infraorbital nerve enlargement (IONE) due to IgG4-related disease.

Case report

A 60-year-old Caucasian man was referred to the adnexal clinic for biopsy of an asymptomatic lump in the region of his right infraorbital foramen. In the preceding 14 years, he underwent multiple biopsies for bilateral submandibular gland enlargement and right cervical lymphadenopathy, which were reported as reactive lymphoid hyperplasia with a suspicion of Hodgkin's disease.

Apart from a palpable mass related to the right infraorbital foramen, ocular examination was unremarkable with normal visual acuity and no proptosis. CT and MRI scans (Figure 1) demonstrated enlargement of the infraorbital nerve along its entire course, extending into the pterygopalatine fossa.

An incisional biopsy of the mass was initially reported as reactive lymphoid hyperplasia but histological review and immunohistochemistry revealed a high proportion of IgG4-positive plasma cells (Figure 2), consistent with the diagnosis of IgG4-related disease; this finding was repeated on review of the previous cervical node and submandibular gland biopsies. Serum IgG4 was grossly elevated and total body CT excluded visceral involvement.

Although steroid treatment was considered, the patient was simply observed as he was diabetic and the lesion was completely asymptomatic, without causing a cosmetic blemish. Four years from presentation, he remains stable under observation.

Comment

IgG4-related disease is characterized by a diffuse or mass-forming inflammatory reaction rich in IgG4-positive plasma cells, often associated with a raised serum IgG4.¹ Cranial nerve involvement, including IONE, has rarely been reported as part of the disease.²⁻⁴ In a recent study, the incidence of IONE was significantly higher in patients with IgG4-related orbital disease than in patients with non-IgG4-related lymphoproliferative orbital disorders.⁵ The diagnosis of IgG4-related disease should be considered in a patient presenting with IONE, especially if multiple sites in the head and neck are

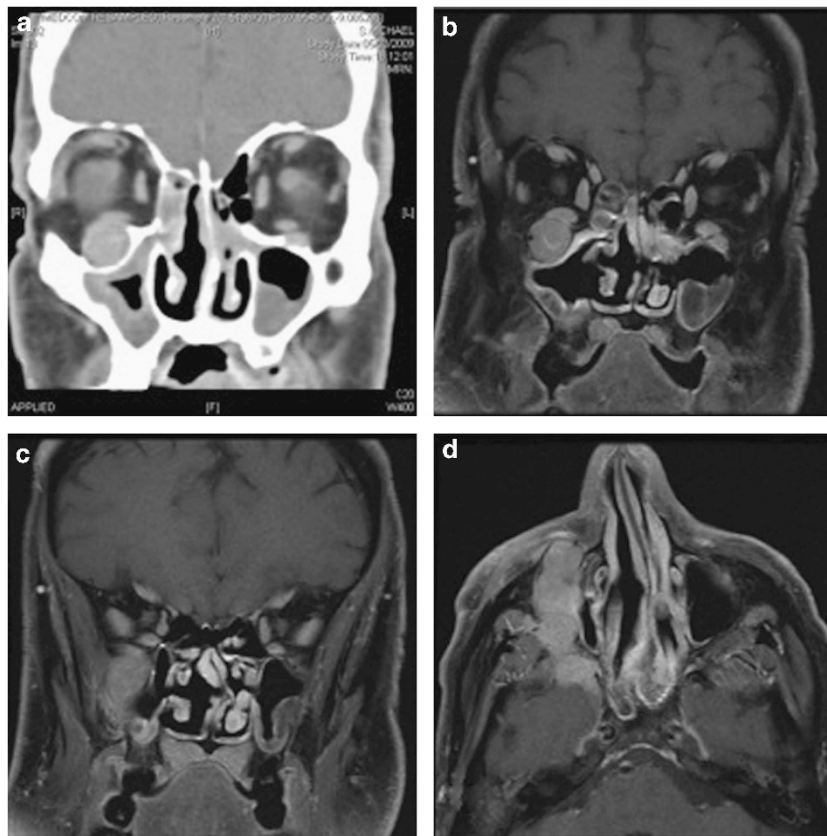


Figure 1 (a) Coronal CT scan and (b) T1-weighted coronal MRI scan demonstrating enlargement of the right infraorbital nerve, abutting the inferior rectus, with expansion of the infraorbital canal, and (c) extension into the pterygopalatine fossa. (d) The gross enlargement of the right infraorbital nerve is seen along its entire course on axial MRI.

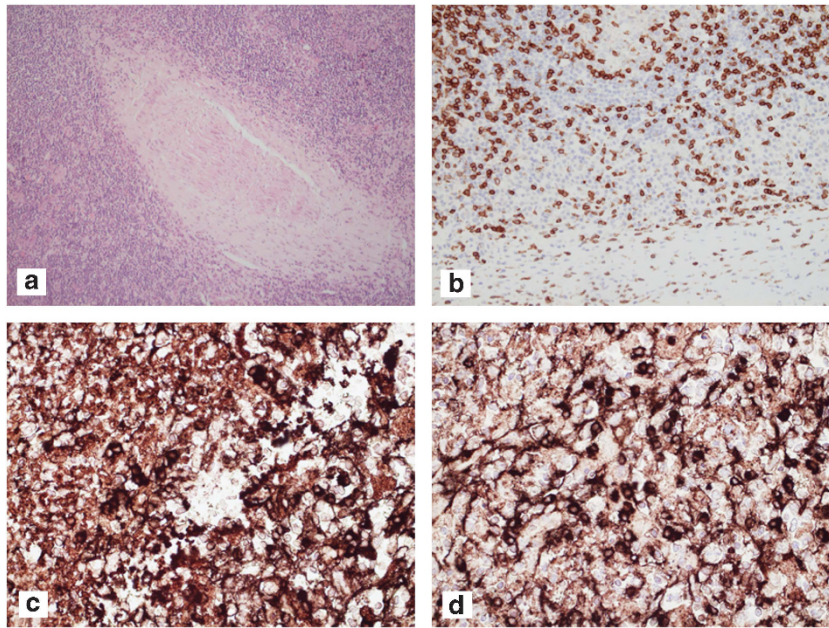


Figure 2 (a) Low power (H+E stain, $\times 10$ magnification) shows infraorbital nerve cuffed by chronic inflammation comprising lymphocytes, histiocytes, and plasma cells. (b) Immunostaining reveals that most of the cells are T cells (CD3 marker, $\times 20$ magnification). (c) Plasma cells are seen expressing generic IgG (IgG marker, $\times 40$ magnification) and (d) many of these plasma cells express IgG4 (IgG4 marker, $\times 40$ magnification).

involved by a lymphoproliferative disorder. The condition usually responds to systemic steroids but other immunosuppressive agents, for example, mycophenolate, methotrexate, and rituximab are also effective.

Conflict of interest

The authors declare no conflict of interest.

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Sir, Spectral domain optical coherence tomography features in niacin maculopathy

We report the spectral domain optical coherence tomography (SD-OCT) and fundus autofluorescence (FAF) findings in a case of niacin maculopathy. To the best of our knowledge, only time domain OCT has been reported; SD-OCT yields better resolution of the affected retinal layers in this unusual disorder.

Case Report

A 57-year-old male presented with blurred vision in the right eye over the past 2 weeks. He reported taking 2000 mg of niacin daily for 5 months after suffering from myalgia related to statin therapy for hyperlipidemia. Visual acuity was 20/40 in the right eye and 20/20 in the left eye. Intraocular pressure was normal and slit lamp