Pachymeningeal enhancement on magnetic resonance imaging in granulomatosis with polyangiitis

Granulomatosis with polyangiitis (GPA) is a multisystemic, antineutrophil cytoplasmic antibody (ANCA)–associated small vessel vasculitis. The orbit and ocular tissues are implicated in approximately a third of cases.[1] Common manifestations are conjunctivitis, episcleritis, orbital inflammation, and uveitis.[1]

In this report we present a characteristic imaging finding in orbital GPA with postcontrast pachymeningeal enhancement associated with orbital inflammation on T1-weighted, fat-suppressed magnetic resonance imaging (MRI) imaging.

A 42-year-old man was referred to the ophthalmology department with a 1-month history of bilateral watery, red eyes; diplopia; and headache. The presentation was on the background of a 7-month history of a lower back ulcer being managed as pyoderma gangrenosum with prednisolone and cyclosporine.

Visual acuities were 6/9 right and 6/6 left. There was a right relative afferent pupillary defect and impaired colour vision as measured with Ishihara plates (4 out of 13). He had bilateral proptosis of 22 mm associated with upper and lower lid swelling (Fig. 1). Movement of the right eye was limited in all directions of gaze with accompanying diplopia except on downgaze. There was bilateral conjunctival injection, and fundus examination demonstrated choroidal folds in the right eye.

MRI demonstrated enhancement of the pachymeninges of the frontal lobes bilaterally, middle cranial fossa, anterior falx cerebri, and left parietal lobe. T1-weighted, postcontrast, fat-suppressed MRI demonstrated bilateral, inferior extraconal enhancing infiltrates in the orbits involving the periosteum (Fig. 2). The mass in the right orbit appeared to extend to the orbital apex. Additionally there was mild thickening of the inferior recti bilaterally.

Cytoplasmic ANCA was positive with a proteinase 3 titre of 80 IU/mL, which in conjunction with the clinical and radiological findings was highly suspicious of GPA. He was admitted to hospital with a view to undergo orbital biopsy but developed central nervous system–associated vasculitis and so was treated urgently with pulsed 1 g intravenous (IV) methylprednisolone for 3 days and 1 g rituximab. Despite treatment he further deteriorated soon after, with increasing proptosis and loss of vision in his right eye to perception of light only, and so was treated again with a further 3-day pulse of 1 g IV methylprednisolone followed by 500 mg IV cyclophosphamide. He then received a further dose of 1 g IV rituximab as well as 5 further fortnightly doses of 500 mg IV cyclophosphamide in addition to a tapering dose of oral prednisolone. At the last review, vision in the right had improved to 6/21 (6/18 with pinhole) with an associated dense, inferior altitudinal visual field defect. He remains under the care of the immunology team with a current immunosuppressive regime of mycophenolate 1000 mg twice daily and prednisolone 5 mg daily.

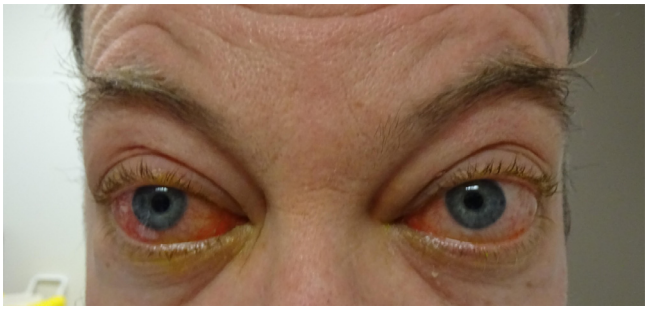

Fig. 1—Photograph on presentation demonstrating proptosis, chemosis conjunctival injection, and lid swelling.

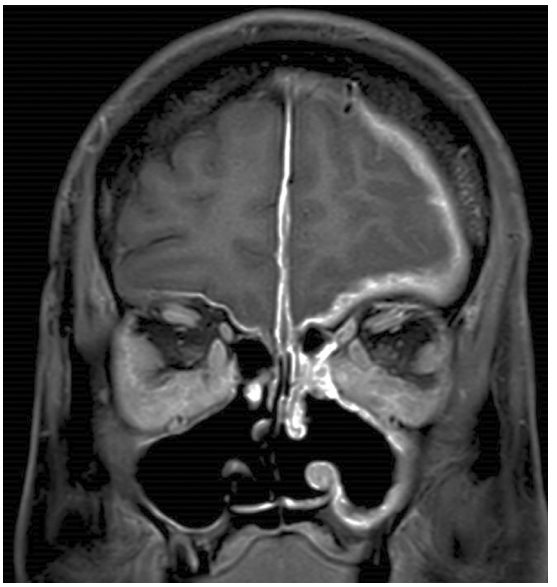

Fig. 2—Coronal T1-weighted, fat-suppressed magnetic resonance imaging scan demonstrating enhancement of the pachymeninges and falx cerebri and inferior extraconal enhancing infiltrates in both orbits involving the periosteum.

Orbital inflammatory disease shares several etiologies with pachymeningitis such as sarcoidosis and IgG4-related disease. The presence of both concurrently is unusual and is highly suggestive of GPA.[2]

Imaging findings typical of orbital GPA include obliteration and infiltration of fat planes and erosion of bone. The theory behind these findings is of contiguous spread of inflammation from the paranasal sinuses to the orbit. It has been suggested that central nervous system involvement, as seen in this case, is also a consequence of the same mechanism.[2,3]

GPA is the most commonly reported specific diagnosis in the literature on orbital inflammation associated with pachymeningeal enhancement.[2] In a study of 59 patients with orbital ANCA-associated vasculitis, 56 of whom had GPA, 12 were found to have contiguous

pachymeningitis.[4] Pachymeningeal involvement was typical of refractory disease and poorer prognosis in this series. In a retrospective series of 6 patients with orbital inflammation and pachymeningitis, 4 had GPA and 2 had tuberculosis.[2]

The mechanism for periorbital postcontrast enhancement on MRI may be secondary to development of periostitis from granulomatous and vasculitic involvement of the sinus mucosa and periosteum.[3] Long bone biopsies in patients with GPA have demonstrated periostitis and vasculitis and correlate with new bone growth on x-ray.[5]

Clinicians should be aware of this imaging finding typifying GPA when evaluating orbital inflammation, particularly when thickening and inflammation of the periorbita is documented.

Bobak Bahrami, Valerie Juniat, Garry Davis, Dinesh Selva
University of Adelaide and South Australian Institute of Ophthalmology, Adelaide, Australia

Correspondence to:
Bobak Bahrami, MBBS, PhD; bobak.bahrami@gmail.com.

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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Infectious keratitis as the presenting sign of giant cell arteritis

Giant cell arteritis (GCA) is a medium-to-large vessel vasculitis that usually presents with headache, loss of vision, jaw claudication, and scalp tenderness. We report a case of biopsy-consistent GCA presenting as an infectious bacterial keratitis. To our knowledge, this is the first such case reported in the English-language ophthalmic literature.

A 76-year-old white woman presented with progressive loss of vision and acute onset of several weeks' duration of pain and redness in her left eye (OS) with associated headache and neck pain (Fig. 1A). On examination, the best-corrected visual acuity was 20/400 in the right eye (OD) and no light perception in the left eye (OS). She had a left relative afferent pupillary defect. Motility examination was full in both eyes (OU). The left eye had conjunctival injection and a purulent corneal ulcer with discharge that was mixed with blood. The left pupil was fixed and dilated with neovascularization of the iris (rubeosis iridis) (NVI) and an intraocular pressure of 50 mm Hg. The right eye had a dense cataract, intraocular pressure of 20 mm Hg OD, and an unremarkable posterior pole examination. No scalp tenderness or temporal artery nodularity was found.

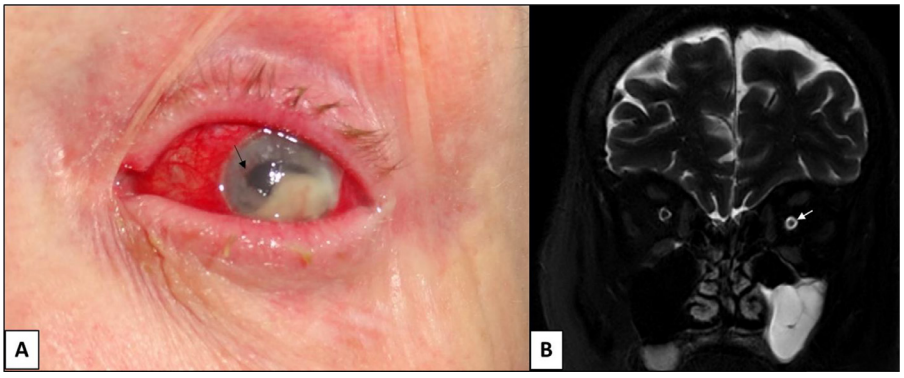

Fig. 1—Clinical presentation. (A) Notice the chemosis and hyperemia of the conjunctiva of the left eye. The cornea shows a tan-white infiltrate from 2:00 to 7:00 clock hours. The pupil is oval and irregular. The iris shows a neovascularized stroma near the pupil margin from 9:00 to 11:00 clock hours (arrow). (B) Coronal T2-weighted magnetic resonance imaging of the brain shows asymmetric abnormal T2 hyperintense signal within the left optic nerve (arrow).