10 Wade EC, Flynn HW, Olsen KR, Blumenkranz MS, Nicholson DH. Sub-retinal haemorrhage management by Pars plana vitrectomy and internal drainage. *Arch Ophthalmol* **108**: 973–978.
11 Handwerger BA, Blodi BA, Suresh R, Chandra, Olsen TW, Stevens TS. Treatment of submacular haemorrhage with low-dose intravitreal tissue plasminogen activator injection and pneumatic displacement. *Arch Ophthalmol J* 2001; **119**: 28–32.

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Sir,

Wegener's granulomatosis causing lid destruction: a further sight-threatening complication

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Wegener's granulomatosis (WG) is a presumed autoimmune disease causing systemic small-vessel vasculitis, necrotising granulomas of the respiratory system, and a focal necrotising glomerulonephritis. Cytotoxic/steroid combination treatment has improved the mean survival from 5 months to a 20-year survival of more than 80%.[1,2] There is a better prognosis with the renal-sparing form of this disease.

Ear, nose, and throat disease is the most common presenting feature of WG, and is found in 75% of patients at diagnosis. Ocular complications from WG occur in around 50% of patients.[2] We present a patient with an unusual, visually threatening and surgically challenging form of orbital WG.

Case report

A 71-year-old man with a 4-year history of extrarenal ANCA-positive WG was referred for an oculoplastic opinion in January 2002. He had initially presented with epistaxis, sinusitis, and left scleritis. In spite of treatment with Campath-1H monoclonal antibody and pulsed cyclophosphamide and methylprednisolone, disease progression occurred. He developed nasal septum and turbinate loss, bony destruction of the medial maxillary sinus walls and ethmoid sinus floors (Figure 1), and an oro-antral fistula. A progressive bilateral medial lower lid ectropion appeared in early 1999.

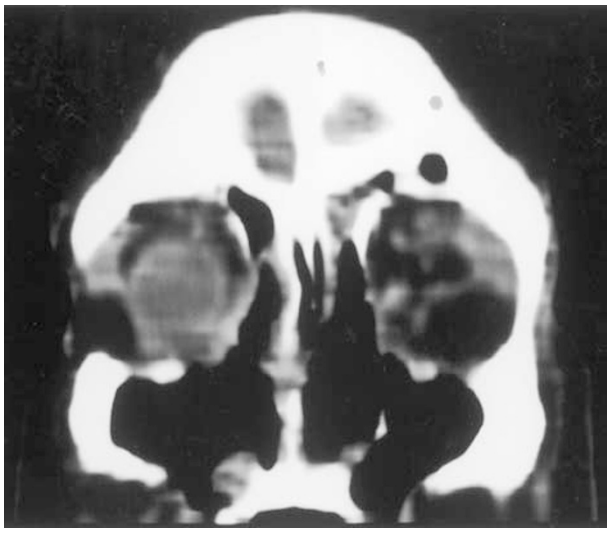

Figure 1 Destructive effects of WG: CT scan, reformatted coronal view. The left eye required enucleation following scleritis-induced perforation. Extensive sinonasal destruction with partial loss of the inferomedial orbital walls and nasal septum.

The left eye was enucleated in 2000, having become painful, blind, and phthisical following a scleritis-induced scleral perforation in August 1999. Abduction of the right eye had also become limited as a further consequence of his orbital WG.

Of primary concern was the progressive destruction of the medial canthal regions bilaterally, with resultant incomplete lid closure. This was causing signs of progressive (right) inferomedial corneal exposure and pannus formation. The bulbar conjunctiva was injected, but there was no active scleritis or marginal corneal thinning. The right lower lid medial attachment was limited to a residual thin strand of canthal tendon. Sclera and adjacent sinus/nasal cavity were visible through the resultant necrotic lid defect, and the surrounding lid skin was injected. The defect extended vertically to involve the upper lid medially, although the upper lid medial canthal attachments appeared secure. Surgery was proposed in view of the risk of progressive corneal damage and possible perforation of this patient's only eye, which had a corrected Snellen visual acuity of 6/12.

With concerns about lid vascular supply/healing potential of the affected lid area and surgery acting as a trigger to local disease flare-up, intervention was restricted to the minimal. The residual medial canthal attachment was divided and a permanent medial tarsorrhaphy was performed, leaving a limited palpebral fissure for axial vision (Figure 2).

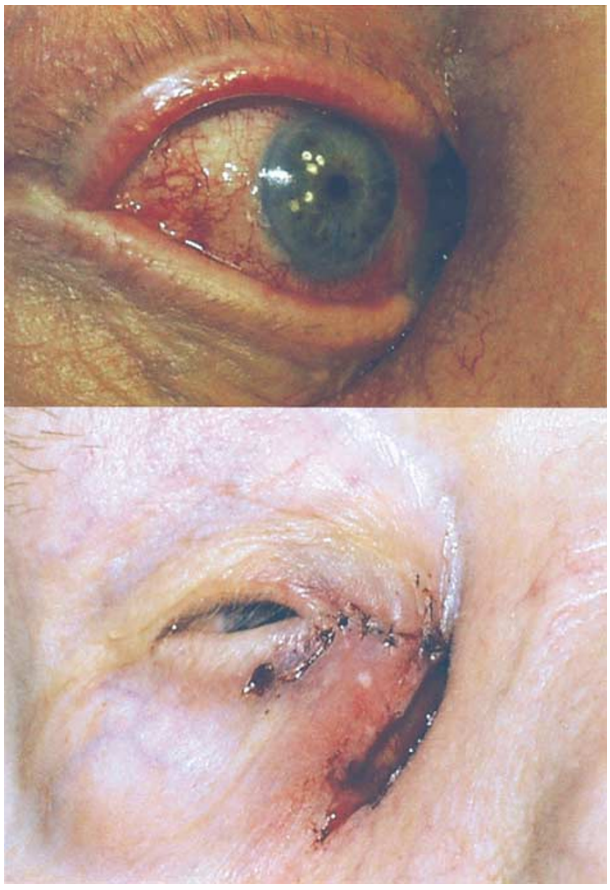

Figure 2 Pre(above) and post-right medial tarsorrhaphy (below): the extensive medial lid defect communicates with the adjacent sino/nasal cavity.

Since the surgery in early February 2002, the tarsorrhaphy has healed well, the corneal epithelial changes have regressed, the patient's acuity has improved to 6/9, and he is coping independently. He had initial visual difficulty with partial pupil occlusion because of the tarsorrhaphy and the altered tear film distribution it caused.

Comment

Ocular signs may be the first manifestation of WG. The *focal* form causes conjunctivitis, scleritis, episcleritis, peripheral ulcerative keratitis, uveitis, retinal vasculitis, and optic nerve atrophy. This is believed to result from focal vasculitis of the small vessels supplying the eye, and/or local granulomas and immune complex reaction and deposition.[1–3] *Contiguous* WG is thought to produce orbital/adnexal inflammation secondary to direct spread of the inflammatory process from the paranasal sinuses or nasopharynx. Pain, proptosis, motility restriction, eyelid oedema, forniceal ulceration (rarely), and recurrent nasolacrimal duct obstruction/dacroadenitis occur.[1,4] In view of the life-saving benefits of immunosuppressive therapy in WG, vigilance regarding this diagnosis has profound implications.

Similar WG-associated medial canthal destruction and eyelid fistulation has been reported.[1,4] Adequate postsurgical healing of the medially separated lower lid in our patient indicated sufficient blood supply from the lateral lid arterial feeder vessels: the anterior deep temporal, superficial temporal, lacrimal, and infraorbital arteries.[5] The inferior lid medial marginal and superficial skin vascular arcades will have been obliterated by the lid necrosis. The upper lid's marginal arcade artery runs deep to the medial canthal tendon before heading superficially to become the inferior marginal arcade vessel,[5] and its course will also have been interrupted by the necrotic tissue defect.

The bilateral medial lower lid ectropion changes developed prior to the appearance of the canthal fistulae, and were probably because of progressive destruction of the underlying supporting lid tissue and the inferomedial orbital walls. Ectropion may therefore be a useful indicator of advancing underlying bone loss and a predictor of the development of this unusual and sight-threatening consequence of Wegener's granulomatosis.

References

1 Woo TL, Francis IC, Wilcsek GA, Coroneo MT, McNab AA, Sullivan TJ. Australasian orbital and adnexal Wegener's granulomatosis. *Ophthalmology* 2001; **108**: 1535–1543.
2 Bullen CL, Liesegang TJ, McDonald TJ, DeRemee RA. Ocular complications of Wegener's granulomatosis. *Ophthalmology* 1983; **90**: 279–290.
3 Pulido JS, Goeken JA, Nerad JA, Sobol WM, Folberg R. Ocular manifestations of patients with circulating antineutrophil cytoplasmic antibodies. *Arch Ophthalmol* 1990; **108**: 845–850.
4 Jordan DR, Addison DJ. Wegener's granulomatosis: eyelid and conjunctival manifestations as the presenting feature in two individuals. *Ophthalmology* 1994; **101**: 602–607.
5 Tucker SM, Lindberg JV. Vascular anatomy of the eyelids. *Ophthalmology* 1994; **101**: 1118–1121.

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Sir,

An unusual macular involvement in pathological myopia

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Pathological myopia is one of the leading causes of blindness in the world. Progressive macular degeneration is the major complication and causes visual loss. These eyes generally have refractive errors greater than 6 dioptres, and the clinical findings are related to increased axial length.[1]

The degenerative changes in pathological myopia initially involve the choriocapillaris, Bruch's membrane, and retinal pigment epithelium (RPE).[2] Central visual function may be affected by breaks in Bruch's membrane, choroidal neovascular membranes and associated haemorrhages, serous and/or haemorrhagic detachments of the retinal pigment epithelium.

Herein, an unusual macular involvement in a case with pathological myopia is discussed with fluorescein angiography and optical coherence tomography (OCT) findings.

Case report

A 21-year-old healthy woman presented complaining of worsening visual acuity, especially in the right eye. The review of her medical story indicated that she began to wear glasses for high myopia when she was 5 years old, and she had been wearing contact lenses for 4 years. Her parents were first cousins and nobody was suffering from any eye disease in her family.

On February 1998, ophthalmic examination revealed that her best corrected visual acuity was RE 8/10 (-13.0 ($-2.0 \times 20^{\circ}$)) and LE 7/10 (-13.50 ($-3.75 \times 170^{\circ}$)). Anterior and posterior examinations were normal. On June 2001, ophthalmological examination demonstrated a visual acuity of 6/10 in the right eye and 8/10 in the left eye with contact lenses. External examination, pupils, intraocular pressure and slit-lamp examination were unremarkable in both eyes. Fundus examination of the right eye demonstrated a small white lesion centred on the fovea (Figure 1a).

Fluorescein angiography demonstrated a horizontal ovoid zone of hyperfluorescence surrounding a nonfluorescent centre in all phases in the right eye (Figure 1b). In the left eye, a similar appearance of the macula was obtained, but it was more indistinct than the right eye.

Horizontal and vertical OCT images obtained through the fovea demonstrated a sharply defined, full thickness defect of the highly reflective (red) band corresponding to the retinal pigment epithelium and choriocapillaris. An area of increased reflectivity was observed beneath this defect. The overlying retinal layers were normal (Figure 1c).

There were pigment epithelial defects in the left macular area, and OCT scans did not demonstrate any abnormality (Figure 2a,b).

A 30 Hz flicker response of ERG reflecting right macular function showed lower amplitude in comparison to the left eye. EOG responses were subnormal in both eyes (the Arden ratio was 1.40 OD and 1.52 OS) without significant light rise and dark trough.

Visual field examination was unremarkable. Axial length was 25.71 mm in the right eye and 25.91 mm in the left eye.

Discussion

Patients with progressive elongation of the eye develop thinning of the choroid and retinal pigment epithelium in the macular area. This may be associated with the development of tilting of the optic disc, peripapillary chorioretinal atrophy, posterior staphylomata, areas of atrophy in pigment epithelium and choroid, and lacquer cracks.[3] Progressive macular degeneration is the major cause of visual loss. Ruptures of Bruch membrane, subretinal neovascular membranes, haemorrhages, and serous detachment can occur in the macular area.

A rapid loss of central vision is usually caused by exudative and haemorrhagic macular detachment overlying areas of choroidal neovascularization. This may occur adjacent to a lacquer crack, in an area of geographic atrophy of RPE, or often in an area of generalized attenuation of RPE and choroid. The new vessels are small and located close to the central macular area. They characteristically appear as a faint grey