membrane and corneal endothelium for nonmechanical preparation and transplantation of corneal endothelium. The positional spikes in the thick autologous corneal flap covering the thin deep corneal allograft may increase postoperative rotational stability of the flap, and in combination with the corneal surface remaining almost untouched may reduce postoperative corneal astigmatism as compared with penetrating keratoplasty. The decreased amount of grafted allogenic tissue in comparison to penetrating keratoplasty may decrease the rate of immunologic graft reactions. Femtosecond laser surgery of the cornea warrants further investigation for nonmechanical posterior lamellar keratoplasty for corneal endothelium transplantation.

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Sir, Wegener's granulomatosis of the orbit in a 5-year-old child

Wegener's granulomatosis (WG) is a rare disorder of unknown aetiology, characterised by a necrotising vasculitis and granulomatous infiltration of the respiratory and renal systems.¹ Ocular manifestations are reported to occur in up to 77% of patients and include conjunctivitis, episcleritis, peripheral ulcerative keratitis, scleritis, uveitis, choroiditis, and central retinal artery occlusion.^{2–6} Orbital involvement is seen in 18–22% of patients with WG.⁷ The disease typically manifests in the fourth and fifth decades, although its occurrence at ages ranging from 7 to 75 years has been reported.^{8–10}

We report the case of a 5-year-old girl presenting with unilateral upper-lid oedema, conjunctival injection, and restriction of eye movements, who was subsequently diagnosed as having WG. To our knowledge, this is the youngest reported case of WG in the literature.

Case report

A 5-year-old girl was referred to the Aristotle University eye clinic with a 4-week history of right-upper-lid swelling, ocular and facial pain, red eye, and probably some limitation of eye movements. As she also had a cold and a temperature of 38°C, she has been treated with topical and systemic antibiotics on the basis of presumed orbital cellulitis, but this did not improve her condition.

On examination, she was found to have right-upper-lid oedema, conjunctival injection, mild restriction of ocular movements, and a palpable hard mass on the upper temporal quadrant corresponding to the lacrimal gland site (Figure 1a). Her visual acuities were 6/6 in both eyes, and the remaining ocular examination, including fundoscopy through dilated pupils, was unremarkable. Paediatric evaluation also did not reveal anything apart from a mildly raised temperature of 37.5°C. She had no significant past ocular and medical history of note.

An MRI was ordered, which showed diffuse swelling in the right orbit at the lacrimal gland region with normal extraocular muscle and optic nerve imaging and no other abnormalities (Figure 2). Her chest X-ray, abdominal MRI, urine analysis, and blood tests were normal apart from a raised erythrocyte sedimentation rate (ESR) of 55 mm in the first hour, and a C-reactive protein (CRP) of 182 mg/l. An orbital biopsy was undertaken that showed mixed granulomatous and active inflammation, and focal areas of fat necrosis and disruption. There was mixed

Figure 1 (a) Right-upper-lid oedema at presentation, and (b) resolution of oedema following treatment.

Figure 2 Diffuse swelling in the right orbit.

inflammatory infiltration consisting of lymphocytes, polymorphonuclear neutrophils, and macrophages around and within the walls of small vessels, but no evidence of fibrinoid necrosis of the vessel wall.

The child was initially treated with prednisolone that slowly improved the symptoms. By this stage, the results of her c antineutrophil cytoplasmic antibody (c-ANCA) test were available and found to be positive to a 1:160 dilution. Her serum was also found to be strongly positive for antiproteinase 3 (PR3) antibodies. These

results in association with the MRI and biopsy findings led to the diagnosis of Wegener's granulomatosis of the orbit. Further paediatric evaluation failed to show any systemic or renal involvement. She continued on a gradually reducing course of oral steroids and within 2 months, her ocular symptoms and signs resolved (Figure 1b).

Discussion

WG may occur in two forms, classical and limited. The latter specifically excludes renal disease and carries a better prognosis.^{3,11} Ocular manifestations are common in both typical and limited WG, noted in 40–77% of patients, and thought to be a result of a necrotising obliterative vasculitis.^{2–6} The diagnosis of WG, as in the case presented, should be based on the correlation of histopathologic, ANCA serologic, and clinical and imaging features.

WG is rare in childhood,^{8,9} and orbital involvement has only been reported twice previously at the ages of 7 and 9 years.^{10,12} To our knowledge, this is the youngest case in the literature and demonstrates that WG should be considered in the differential diagnosis of orbital disease even in very young children.

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