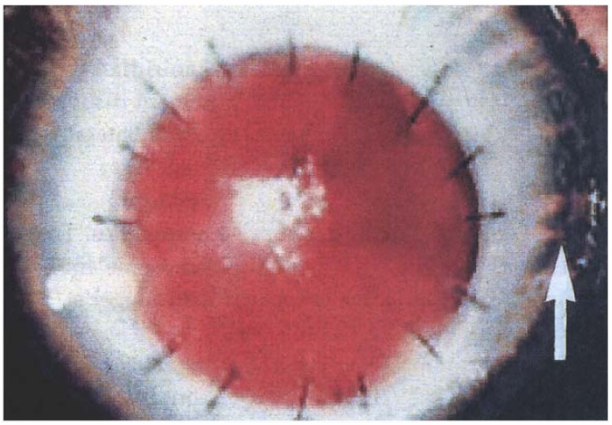

Fig. 2. *Photograph of the right eye of the son, taken through the operating microscope, showing the penetrating keratoplasty and aniridia, with the ciliary processes shown against the red reflex (arrow).*

However, her only child, a 5-year-old boy, had bilateral aniridia, associated with congenital glaucoma, buphthalmos and secondary corneal opacity. This young patient received bilateral glaucoma implant surgery, and subsequently bilateral penetrating keratoplasties (Fig. 2), all within his first year of life. At 2 years of age, his right eye suffered from corneal graft rejection and retinal detachment. Even with repeated surgeries, blindness still ensued. His left eye developed endogenous endophthalmitis secondary to an episode of *Streptococcus* pneumonia, eventually necessitating enucleation.

Comment

Associations between aniridia and other anterior segment dysgeneses are known. A mutated aniridia gene has been reported in a family with Rieger's anomaly.[5] Furthermore, Sey mice with a *PAX6* mutation could have clinical features of aniridia, corneal opacifications, keratolenticular adhesions and anterior synechiae suggestive of Peters' or Rieger's anomalies.[5] These may suggest that Rieger's anomaly and/or syndrome may result from loss of function of one copy of *PAX6*.

We investigated the sequence alterations of the coding exons (exons 4 to 13) of the *PAX6* gene of these two patients by polymerase chain reaction and direct DNA sequencing. No *PAX6* mutations were found, suggesting that both patients might have full *PAX6* functions. The occurrence of Rieger's syndrome and aniridia in the same family is interesting and unreported previously. It is unclear whether the phenotypic features of these two patients are associated with defects of the same or different genes. Further genetic investigations would be worthwhile.

Supported in part by (1) a direct grant of the Medical Panel, the Chinese University of Hong Kong, (2) Mrs Annie Wong Eye Foundation, Hong Kong, and (3) Mr W.K. Lee Eye Foundation, Hong Kong.

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Sir,

Granulomatous orbital myositis

Orbital myositis is the most common cause of inflammatory enlargement of the extraocular muscles after dysthyroid eye disease.[1] The inflammation of the muscles is idiopathic and the condition is considered a variety of orbital pseudotumour.[2,3]

Clinically, orbital myositis is characterised by acute pain, exacerbated by eye movement.[4] Ocular and periocular inflammation are absent.[5] Radiological confirmation of the diagnosis involves demonstration of enlargement of the extraocular muscles, which may either involve or spare the tendons of insertion.[6,7] The condition is often self-limiting and tends to resolve spontaneously within 3–6 weeks.[8] However, treatment is indicated to alleviate discomfort, hasten recovery, limit post-inflammatory sequelae and prevent later recurrences. Systemic corticosteroids are the mainstay of treatment, but recurrences are common and may require long-term steroid therapy.[9]

We report a case of a 9-year-old girl with orbital myositis, characterised by granulomatous inflammation, which has rarely been described.

Case report

A 9-year-old girl presented to her general practitioner with mild bilateral ocular discomfort. Four days later she developed vertical diplopia, followed the next day by bilateral proptosis, which rapidly progressed over the next 3 days.

On examination unaided visual acuity measured 6/9 in the right eye and 6/6 in the left. There was bilateral proptosis of 4 mm and 2.5 mm in the right and left eyes respectively. The right eye manifested upward dystopia of 2 mm and hypertropia of 25 prism dioptres, associated with ptosis. There was almost complete bilateral external ophthalmoplegia but no pain on attempted eye movement. There was a subtle right relative afferent pupillary defect. There was no evidence of intraocular inflammation. Visual fields were full. Notable features were the complete lack of both pain and clinical evidence of inflammation.

CT scan of the orbits revealed gross enlargement of the extraocular muscles (Fig. 1), which was confirmed on ultrasound examination. The extraocular muscles were enlarged to greater than 4 times their normal size.

The risk of visual loss and the rapidity of progression of the disease led to the decision to perform a biopsy of the right inferior rectus muscle, prior to commencing steroid therapy. The tissue was fixed in buffered glutaraldehyde (2.5%) and processed through paraffin.

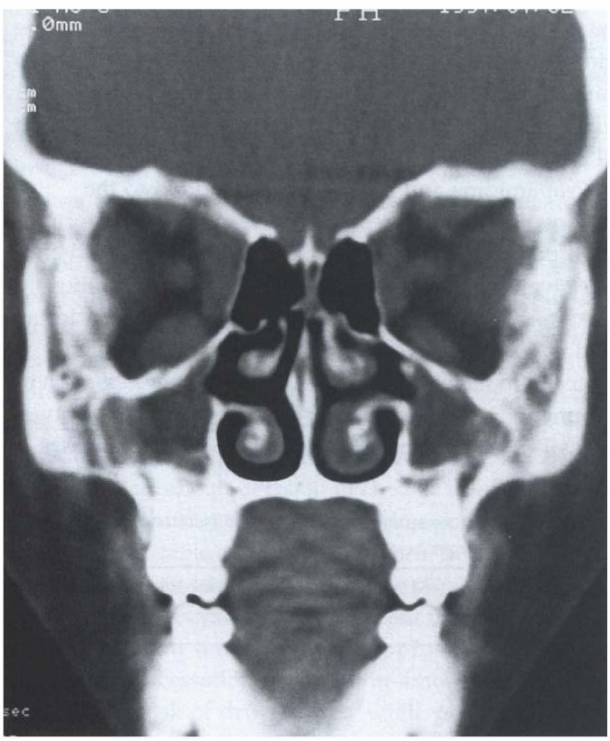

Fig. 1. *CT scan of the orbits, coronal view, demonstrating gross enlargement of the extraocular muscles, mucosal thickening in both maxillary antra and swelling of the left inferior turbinate.*

Sections were stained with H&E, PAS and Ziehl Neelsen stains. Microscopic examination by Prof. William R. Lee revealed a giant cell granulomatous reaction, without evidence of caseation or necrotising vasculitis (Fig. 2). The histopathological findings also did not have the compact pattern of sarcoidosis. Special stains did not reveal pathogenic organisms. Occasional muscle fibres contained several nuclei, indicating regeneration. There was no evidence of involvement of orbital fat. The pathological diagnosis was therefore a primary granulomatous myositis of unknown aetiology.

The serum anti-neutrophil cytoplasmic antibody level was elevated (ANCA titre = 1:40P), anti-nuclear antibody studies were negative and serum angiotensin converting enzyme measured 21 U/ml, which is within the normal adult range. Liver function tests and abdominal and thoracic imaging were all normal. Haematological examination revealed moderate thrombophilia and toxic granulation of neutrophils. The erythrocyte sedimentation rate was elevated at 50 mm in the first hour.

On the operating table immediately after the biopsy 250 mg prednisolone was administered intravenously in the light of the potential risk of acute muscle swelling as a sequel to the surgical trauma. This was followed by oral prednisolone 30 mg daily from the following day. Twenty-four hours later improvement was dramatic, with return of vision to 6/6 in both eyes, return of limited eye movements and reduction of proptosis to 2 mm in the right eye and 1.5 mm in the left. Four weeks later, visual acuity had improved to 6/5 in each eye and extraocular movements were full, with no proptosis or diplopia.

Two months following presentation, the ocular condition was stable and the steroids were slowly tapered off. A relapse of proptosis then occurred, which was controlled by increasing the steroid dose. The patient was then maintained on oral steroids, slowly tailing off over the ensuing 14 months, and has since remained stable, with no recurrence.

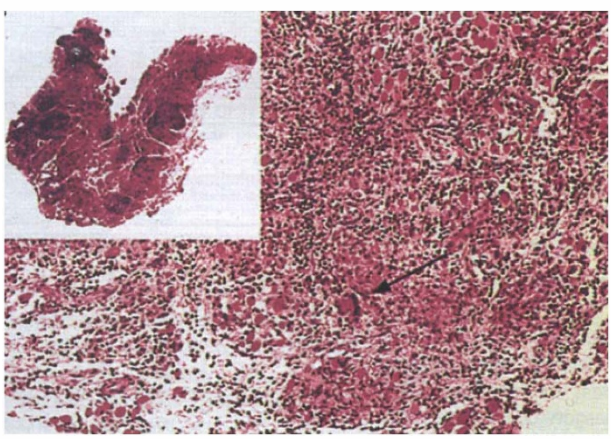

Fig. 2. *The fibres of the inferior rectus muscle are infiltrated by a giant cell granulomatous inflammatory reaction (arrow). The inset is a low-power view of the larger specimen: note the clusters of inflammatory cells scattered throughout the muscle (H&E ×120, inset ×10).*

Comment

A lipogranulomatous reaction due to necrosis of orbital fat is sometimes seen in orbital myositis. However, primary granulomatous inflammation of the extraocular muscles, though recognised, is extremely rare.[10] Such painless myositis may represent a localised manifestation of sarcoidosis.[11] Interestingly, idiopathic orbital inflammatory disease has been referred to in the past as 'non-specific granuloma of the orbit',[12] which is correct in the macroscopic sense, characterising a non-neoplastic space-occupying lesion.

Treatment of the condition with systemic steroids results in rapid resolution of the inflammation. Premature withdrawal may result in recurrence. The underlying cause, however, of this rare sporadic disorder remains an enigma.

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Sir,

Polyhexamethylene biguanide (0.02%) alone is not adequate for treating chronic *Acanthameoba* keratitis

The use of polyhexamethylene biguanide (PHMB), a cysticidal drug with minimal ocular surface toxicity, was reported by Larkins *et al.*[1] in 1992 for the treatment of *Acanthamoeba* keratitis. Its effectiveness in killing trophozoites and cysts of *Acanthamoeba* has been documented by *in vitro* sensitivity testing.[1,2] Currently, most authors recommended multi-drug therapy, often in combination with PHMB, for the treatment of *Acanthamoeba* keratitis.[2] The use of multiple drugs, however, increases the risk of ocular surface toxicity. We therefore prospectively evaluated the safety and efficacy of monotherapy with topical PHMB 0.02% in the treatment of chronic *Acanthamoeba* keratitis. In the three patients reported, monotherapy with topical PHMB 0.02% resulted in persistent infection. The use of combination therapy, with the addition of propamidine and neomycin, eradicated the corneal infection and subsequent surgery resulted in good visual recovery.

Case report

The three patients with culture-proven *Acanthamoeba* keratitis presented 12, 15 and 8 months after onset of symptoms with pre-treatment visual acuities of hand movement, 1/60 and 3/60 respectively. The results of *in vitro* sensitivity testing are summarised in Table 1. All three patients with *Acanthamoeba* keratitis were treated initially with only topical PHMB 0.02%. The dosing schedule was one drop every hour, round the clock in the first week, two hourly between 0800 and 2400 hours for the next week and then gradually reducing, based on the clinical response. Although clinical signs and symptoms initially improved, the condition subsequently worsened with positive culture results from corneal re-scrapings in all patients.

Propamidine and neomycin were added to the therapeutic regimen, resulting in resolution of infection in all patients. The eyes were clinically uninflamed and there were no clinical signs suggestive of continuing infection. The combined treatment was continued for a total of 6 months even though the infection seemed clinically to have settled well long before the cessation of treatment. Subsequent surgery for the corneal opacity (cases 1, 2, 3), glaucoma (cases 1, 2) and cataract (cases 1,

Table 1. In vitro *sensitivity patterns of two isolates of* Acanthamoeba *from cases 1 and 2*

	Minimum cysticidal concentration (μg/ml)	
Drugs	Case 1	Case 2
Propamidine	125	3.9
Neomycin	> 500	–
Paromomycin	> 500	250
Hexamidine	–	0.49
Chlorhexidine	–	3.9
PHMB	7.8	1.9

PHMB, polyhexamethylene biguanide.