due to adduction lag, abduction nystagmus or deficient VOR response.

In some instances in which MLF are damaged, adjacent structures, particularly the paramedian pontine reticular formation, abducens nuclei, abducens fascicle and oculomotor nuclear complex, are also involved. Ptosis, partial nuclear oculomotor nerve palsy and trochlear nerve palsy are some of the clinical signs described with INO.[3–5] Isolated pupil-sharing third nerve palsy has also been reported as a presenting sign in a patient with MS.[2]

Our patient presented with horizontal diplopia and unilateral ptosis. The clinical signs suggested bilateral INO with left partial oculomotor nerve palsy. Though the MRI of the midbrain did not show any foci of demyelination, it is likely that both the MLF and a part of the oculomotor fascicles were involved. The presence of ptosis suggests that the fibres controlling the levator palpebrae superioris muscle are located caudally in the oculomotor fascicles.

A clinical diagnosis of MS was made on the basis of the patient's past history and her most recent problems. MRI helped to confirm the diagnosis. In most patients, visual symptoms resolve completely, because of either recovery of function of MLF axons or central adaptive mechanisms. Our patient had complete resolution of symptoms in less than 2 weeks.

We are not aware of any reports of bilateral INO and partial unilateral oculomotor nerve palsy in MS. This is an example of the diverse ocular presentations of this demyelinating disorder.

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Sir,

A family with Rieger's syndrome and aniridia

Rieger's syndrome is an autosomal dominant disorder that affects the anterior segment of the eye bilaterally, causing glaucoma in 50% of those affected. It is associated with dental hypoplasia, craniofacial dysmorphism and umbilical stump abnormalities. The responsible gene, *RIEG/PIX2*, is on chromosome 4q25.[1] There are also other genes associated with Rieger's syndrome, such as *RIEG2*[2] and *FKHL7*.[3]

Aniridia encompasses a group of congenital ocular disorders with iris hypoplasia in common. It may be inherited in an autosomal dominant or recessive pattern. There is also a sporadic form associated with Wilms' tumour. Aniridia is caused by mutations in the transcriptional regulator, the *PAX6* gene on chromosome 11p13.[4]

We report here the rare occurrence of both Rieger's syndrome and aniridia in a family and the molecular study of their *PAX6* status.

Case report

We examined a 39-year-old Chinese woman who had a phthisical right eye with only hand-movement visual acuity, and a left eye with visual acuity of 20/20. She had a history of total retinal detachment of her right eye after complicated combined trabeculectomy and cataract surgery 5 years earlier. Her left eye had open-angle glaucoma, controlled with four topical medications. On examining her left eye, there were peripheral iris strands attached to a posterior embryotoxon, iris stromal hypoplasia, corectopia and ectropion uveae (Fig. 1). The cup: to disc ratio of her left eye was 0.6, which was compatible with her visual field loss. On general examination she was noted to have a broad flat nasal bridge, maxilla hypoplasia, telecanthus, hypertelorism and microdontia, compatible with Rieger's syndrome. Her husband had normal vision and no ocular abnormalities were detected.

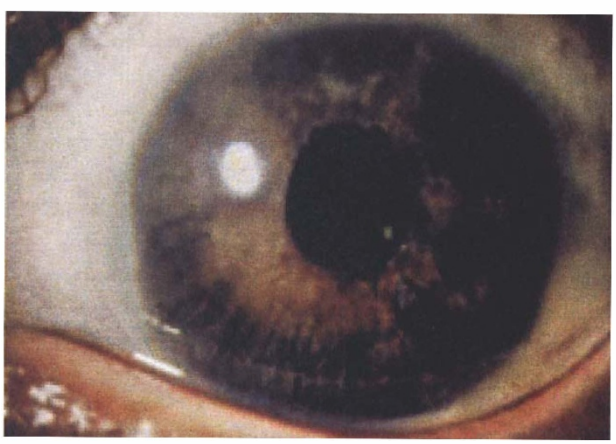

Fig. 1. *Slit-lamp photograph of the patient's left eye, showing typical Rieger's features including posterior embryotoxon, iris stromal hypoplasia, corectopia and ectropion uveae.*

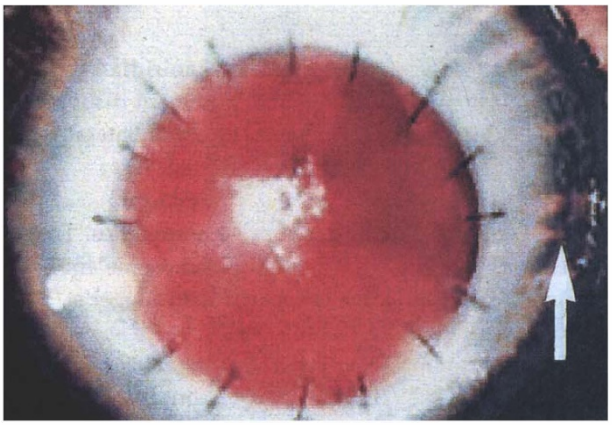

Fig. 2. *Photograph of the right eye of the son, taken through the operating microscope, showing the penetrating keratoplasty and aniridia, with the ciliary processes shown against the red reflex (arrow).*

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.

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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]