We understand cultural differences in contact lens wear may influence the incidence of keratoconus between the ethnic groups, however, the evidence for this to cause such a significant difference is limited. Weed *et al*[2] reported only 28% of patients presenting with a new diagnosis of keratoconus had a history of contact lens use. In our study, the most common treatments for refractive error, prior to referral, were glasses or soft contact lens rather than rigid lens, which are postulated to be the most likely cause for keratoconus. Interestingly, in our study, there was no significant difference in initial hospital treatment between the white and Asian patients. A total of 41% of white patients and 44% of Asian patients were treated with contact lens.

Cozma *et al*'s findings further support the theory of a genetic basis to keratoconus and emphasizes the need for genetic research.

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T Georgiou, CL Funnell, A Cassels-Brown and R O'Conor

Department of Ophthalmology
Leeds General Infirmary
St. George Street
Leeds LS2 INS, UK

Correspondence: CL Funnell
Tel: +44 113 2432799
Fax: +44 113 2432799
E-mail: c.l.funnell@doctors.org.uk

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Sir,

Mucous plaque keratitis associated with aniridia keratopathy

Aniridia is a rare congenital disorder with autosomal dominant inheritance in the majority of cases, but it occurs sporadically in 15% of cases.[1,2] Affected individuals show a spectrum of clinical features. Aniridia is the defining feature but developmental defects are not restricted to the iris. Corneal opacification, cataract, glaucoma, and macular hypoplasia all contribute to visual loss. Aniridia patients with sporadic mutation have a higher incidence of Wilms' tumour.[3]

We present a case where marked mucous plaque keratitis developed in a patient with aniridia keratopathy following a corneal graft. Mucous plaques are not a known feature of aniridia keratopathy. It is unusual for mucous plaques to develop unless very severe atopic disease is present. Our patient had marked corneal disease with large mucous plaques in the presence of only mild atopic conjunctivitis. We propose that the nonhealing corneal epithelial defect of aniridia keratopathy provided the environment for rapid formation of mucous plaques.

Case report

A 32-year-old woman with familial aniridia had a long history of progressive corneal opacities in both eyes. Her father, grandfather, and sister were also affected. At the age of 12 years, her visual acuity was 6/36 in both eyes; by the age of 19 years, it dropped to 6/60 and she had developed chronic nonhealing corneal ulcers that were worse in her left eye. Exacerbations of the corneal ulceration were treated with topical lubricants and antibiotics. In 1993, the patient underwent left lamellar keratoplasty with an initially satisfactory result with visual acuity improving from 1/60 preoperatively to 6/36 three months postoperatively. After 3 years, she developed glaucoma in the left eye and her intraocular pressures became refractory to medical treatment. Trabeculectomy augmented with mitomycin C was performed on the left eye in March 1998 and the intraocular pressure has since been maintained below 20 mmHg with timolol drops.

The right eye later developed nonhealing corneal epithelial defects similar to those in her left eye and a lamellar keratopathy was performed in the right eye in October 2000. The corneal graft was initially clear but later developed epithelial disturbance with deposition of mucus which soon turned into persistent plaques on the surface. This was associated with deterioration of vision and pain.

At that time we were not aware of any coexisting problem that might have contributed to the formation of mucous plaques. She used topical steroids and mucolytic agents but the mucous plaques did not resolve. She therefore had a repeat lamellar keratoplasty in October 2002. Further questioning revealed a history of hay fever associated with watery eyes. In addition, she was prone to bronchospasm and was allergic to house dust and cats.

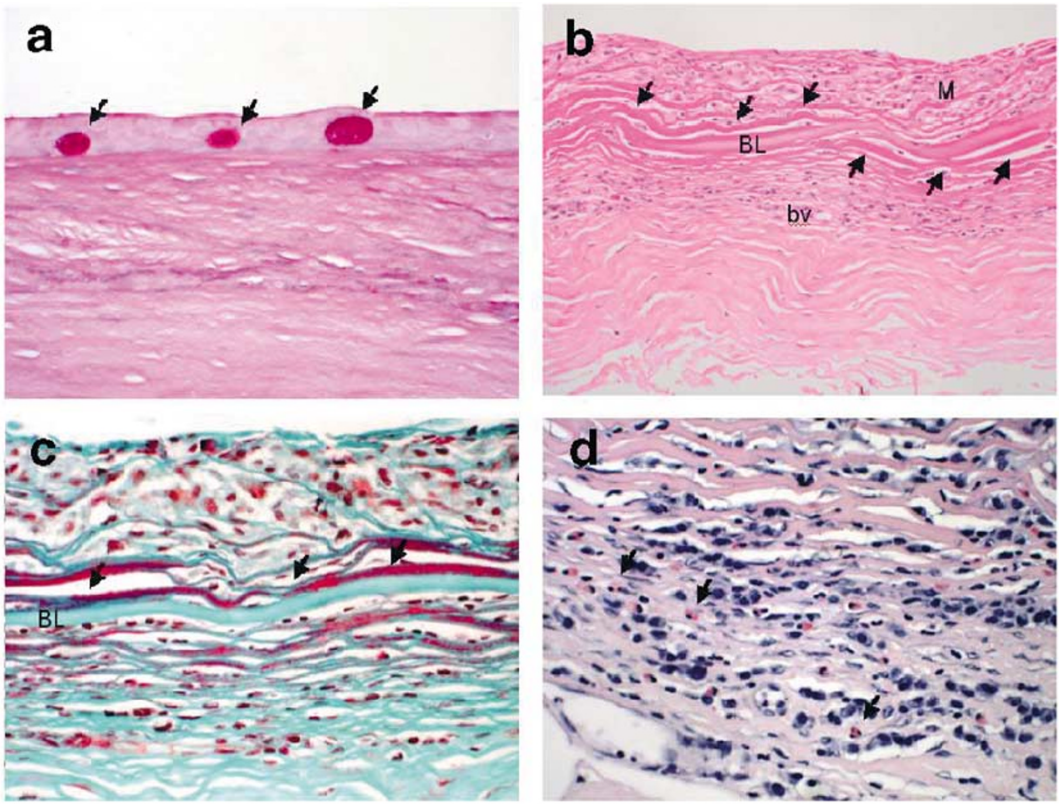

Figure 1 (a) Left lamellar corneal button from 1993 showing goblet cells (arrows) in the surface epithelium characteristic of aniridia keratopathy (PAS, ×400). (b) Right lamellar corneal button from 2002 showing eosinophilic mucous plaques (arrows) either side of Bowman's layer (BL). There is a surface layer of macrophages (M). Blood vessels (bv) are present in the midstroma. (H&E, ×250). (c) Right lamellar corneal button from 2002 showing granular red staining of haemoglobin fragments entrapped in mucous plaques (arrows) (Masson's trichrome, ×400). (d) Right lamellar corneal button from 2002 showing inflammation including numerous eosinophils (arrows) in the midstroma (Carbol chromotrope, ×400).

Examination of the upper tarsal conjunctiva showed a mild papillary response.

The left lamellar keratoplasty specimen from 1993 consisted of a 7 mm lamellar corneal disc with an off-centre subepithelial nodule and also peripheral vascular pannus. Histological examination showed the epithelium of variable thickness with occasional PAS-positive goblet cells in the mid periphery (Figure 1a). Corresponding to the subepithelial nodule seen grossly, there was a subepithelial band of fibrous tissue containing inflammatory cells and small blood vessels. These features were similar to those seen in the right lamellar keratoplasty specimen from 2000.

The right lamellar keratoplasty specimen from 2002 consisted of a 7 mm lamellar corneal disc with patchy white areas and vascular peripheral pannus. On histological examination, the surface was devoid of epithelium and covered by an unusual layer composed predominantly of macrophages. A short surviving segment of Bowman's layer was identified with PAS-positive mucous plaques deposited on either side (Figure 1b). Blood staining of mucous plaque was visible with special staining (Figure 1c). Within the superficial stroma, there was an inflammatory infiltrate composed of lymphocytes, plasma cells, and prominent eosinophils (Figure 1d). The deeper stroma was unremarkable.

Discussion

Aniridia is a rare bilateral congenital anomaly associated with a variety of clinical features. The underlying aetiology is a defect in the PAX-6 gene (short arm of chromosome 11). The corneal changes of aniridia keratopathy are progressive and include thinning of epithelium, peripheral corneal pannus, and scarring of Bowman's membrane leading to opacification of the cornea. The epithelial defects fail to heal properly.

In our case, the pathological features of the first corneal button from the right eye were characteristic of aniridia keratopathy with goblet cells in the

epithelium and secondary inflammation. However, the second graft showed an unusual pannus associated with mucous plaques, corneal vascularisation, and an inflammatory infiltrate rich in eosinophils and macrophages. While some of these features can be interpreted as the consequence of prolonged epithelial ulceration, the possibility of an atopic reaction was considered and confirmed on further questioning and examination. The second graft therefore shows dual pathology of modified atopic keratitis in a patient with recurrent epithelial ulceration due to aniridia keratopathy.

Corneal abnormalities seen in aniridia have been attributed to deficiency of limbal stem cells and incursion of conjunctival epithelial cells onto the cornea. It has also been suggested that low levels of PAX-6 expression result in epithelial fragility related to cytokeratin (K3 and K-12) deficiency and in addition, an abnormal wound-healing response due to deficiency of matrix metalloproteinase-9.[4,5]

Mucous plaques are an uncommon complication of severe atopic and vernal keratoconjunctivitis, but can also be seen with herpes zoster ophthalmicus, keratoconjunctivitis sicca, and superior limbic keratitis.[6,7]

It is unusual for mucous plaques to develop unless very severe atopic disease is present. Our patient had marked corneal disease with large mucous plaques in the presence of minimal conjunctival involvement. We believe that a genetically determined abnormality of corneal epithelial healing in an aniridia patient with a very mild atopic conjunctivitis led to pronounced mucous plaque formation.

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S Saidkasimova, F Roberts and JL Jay

Tennent Institute of Ophthalmology
1054 Gt Western Rd
Glasgow G12 0YN, UK

Correspondence: S Saidkasimova
Tel: +44 141 211 1041
Fax: +44 141 211 2054
E-mail: shohistas@yahoo.co.uk

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Sir,

Chronic myeloid leukaemia presenting as venous stasis retinopathy

I read with interest the comprehensive article by Sharma *et al*[1] reviewing the ophthalmologist's role in the ophthalmic manifestations of acute leukaemia. I would like to report a case of chronic myeloid leukaemia that presented to a DGH eye casualty.

Case report

A 29-year-old man presented with sudden onset diminution of vision in his left eye. He had glandular fever infection 10 years ago. Visual acuity was normal in right eye but restricted to 6/18, N10 in the left eye. Fundus examination revealed bilateral venous stasis retinopathy with scattered roth spots, the left eye being more severely affected than the right. The left eye also revealed macular haemorrhages (Figure 1).

On further history taking, he agreed to easy bruising, decreased appetite, and night sweats. General examination revealed gross hepatospleenomegaly in addition to multiple bruises of different ages on his body. Urgent blood test revealed marked leukocytosis with blood film consistent of Philadelphia-positive chronic myeloid leukaemia.

He was urgently referred to haematology where he received leukophoresis and cell and sperm storage. Oral hydroxyurea, allopurinol, and later imatinib-achieved cytoreduction and remission. His general and