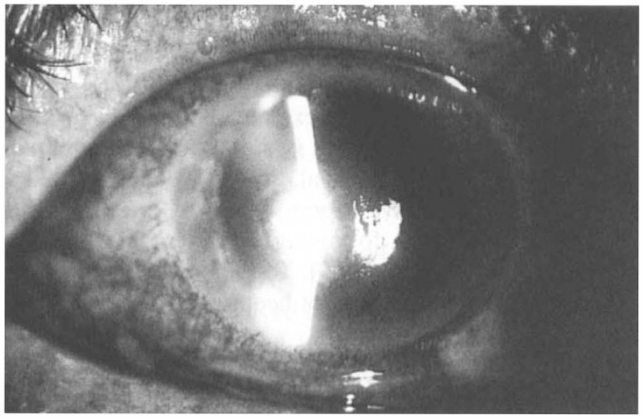

Fig. 2. *Left eye after a week of intensive treatment, showing the abscess resolving.*

has now become a recognised form of treatment for myopia. Its use on fully sighted human eyes was preceded by trials on animal models, blind and partially sighted human eyes.

A number of studies on corneal epithelial changes following PRK have shown no significant long-term deleterious effects.[1–4] The majority of clinical trials have incorporated the use of a topical steroid regimen following the PRK and some studies have suggested that regression of correction and corneal haze can be controlled by titration of the steroid dose.[5–7]

None of the reports to date has attributed any significant complication to the use of steroid following PRK.[8–11] The use of post-operative steroids to modulate wound healing remains a controversial subject. Piebenga *et al.*[10] in their trial found no statistically significant role for steroids. Gartry *et al.*[12] in their trial found better results in the steroid group at an early stage though the advantages were lost at 3 and 6 month follow-up. They have suggested that long-term steroid usage would be unacceptable given the risks and the lack of benefits.

Blepharitis is usually associated with peripheral rather than central corneal lesions.[13] We attribute the bacterial keratitis seen in our patient to a combination of factors including pre-exisiting blepharitis, possible ocular surface abnormality following the excimer laser treatment and long-term steroid usage. This case demonstrates the need for closer monitoring of patients commenced on steroids following PRK and focuses attention on the need, if any, for steroid treatment in these patients.

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Sir,

Blepharophimosis Syndrome: An Atypical Case

Blepharophimosis was first reported in 1841 by von Ammon.[1] Blepharophimosis syndrome, which was subsequently described by Spaeth in 1956,[2] is characterised by a narrowed horizontal palpebral aperture, ptosis, telecanthus and epicanthus inversus. Several other associations of blepharophimosis syndrome have been described involving mainly the lacrimal drainage system, the lids and certain systemic features.[3,4] We describe here a case with features of blepharophimosis syndrome together with several other unique associations not reported previously in the ophthalmic literature.

Case Report

A 20-year-old man presented with bilateral ptosis. He gave no history suggestive of birth trauma or mental retardation. There was no history of consanguinity and academically he was of average intelligence from his records. The ptosis had been present since birth with no previous history of any surgery. There was no family history of a similar illness.

On physical examination the patient was a tall, thin individual with clinical features and anthropometric measurements suggestive of arachnodactyly. He had low-set ears with a normally arched palate. Radiological survey of the long bone confirmed the arachnodactyly. However, neurological and cardiovascular examinations revealed no other abnormality.

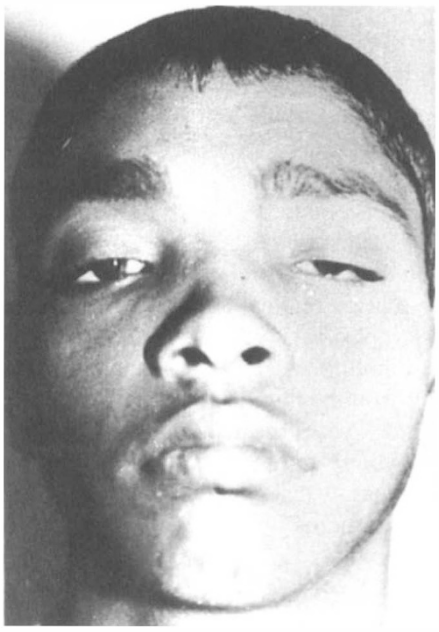

Fig. 1. *Pre-operative photograph of the patient showing bilateral ptosis, left more than the right.*

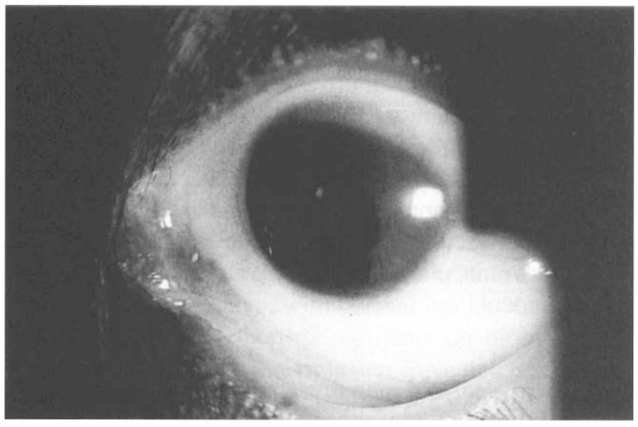

Fig. 2. *Photograph of the left eye showing a typical iris coloboma and ectropion uveae.*

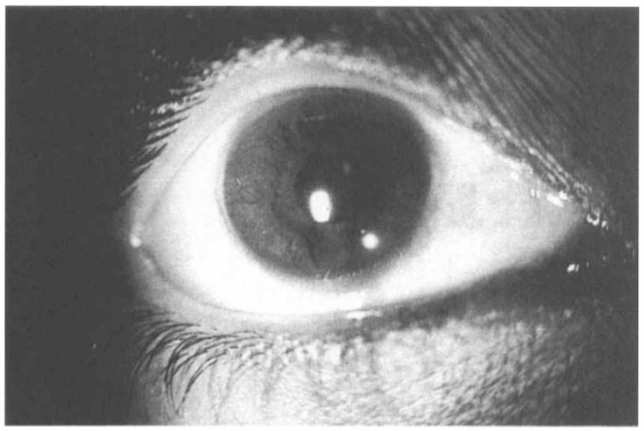

Fig. 3. *Photograph of the right eye showing lateral ectropion and medial entropion. Extension of the colarette inferiorly as a ridge on either side of the forme fruste coloboma is seen with the ectropion uveae.*

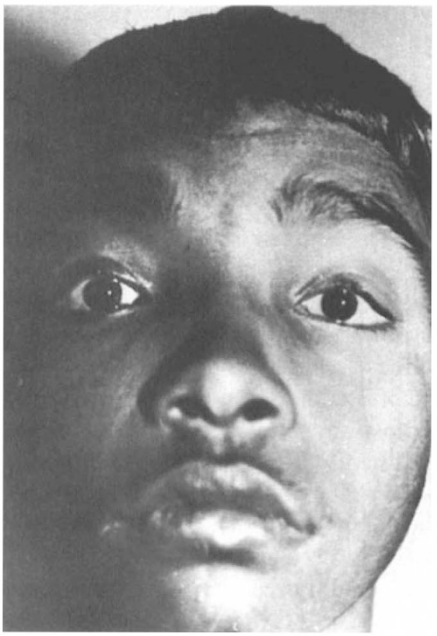

Fig. 4. *Post-operative photograph of the patient with symmetrical correction following the bilateral, modified frontalis sling procedure.*

On ocular examination his best corrected visual acuity was 6/6 (unaided) in the right eye and 6/18 in the left eye with 4 dioptres hypermetropic refraction, revealing anisometropic amblyopia in the left eye. He had a head posture of chin elevation with 3 mm ptosis in the right eye and 4 mm ptosis in the left eye. Levator action was poor (2 mm) in both eyes. The horizontal width of the palpebral aperture was 26 mm in both eyes. Both lower lids showed medial entropion and lateral ectropion with lateral displacement of the inferior puncta. The upper puncta were normal in position and the lacrimal drainage system was patent. There was telecanthus with a broadened and flat nasal bridge. There were no epicanthal folds nor any hypertelorism (Fig. 1).

On slit lamp biomicroscopy, the left eye had an inferior incomplete coloboma (Fig. 2) while the right eye showed hypoplasia of the iris stroma in the corresponding area with a thickened ridge-like tissue on either side of the hypoplastic iris (Fig. 3). Both eyes had marked ectropion uveae with the lenses being clear and normal in position. There were no signs of active or healed iridocyclitis or rubeosis iridis. Fundus and vitreous were essentially normal.

The patient underwent a bilateral frontalis sling procedure, as described by Betharia,[5] under local anaesthesia. Post-operatively the correction was adequate and symmetrical in the primary position with no other complications (Fig. 4).

Comment

Associations and management of blepharophimosis syndrome have been well described in the literature.[2–4] In this case, amongst the systemic features observed, arachnodactyly was an unique finding and has not been described

as an association of blepharophimosis syndrome in the literature. The patient's marfanoid habitus was not associated with any other systemic or ocular features of Marfan's syndrome. The other physical findings such as low-set ears and a broad nasal bridge are well-known associations of blepharophimosis syndrome.

In our case, incomplete coloboma was evident in the left eye only. The features described in the right eye could possibly be explained as a forme fruste of an iris coloboma. However, we could not find any such description in the literature. Baraitser and Winter[6] reported a new syndrome of three cases, two of them siblings, with a combination of short stature, mental retardation, bilateral iris coloboma, ptosis, hypertelorism and a broad nasal bridge. Our case differed in having normal mental status, arachnodactyly and unilateral iris coloboma and bilateral congenital ectropion uveae. This report bore the closest resemblance to our patient.

The amblyopia observed in the left eye could possibly be explained by the hypermetropic refractive error and the asymmetrical ptosis with the compromise of the pupillary axis. That these factors predispose to the development of amblyopia in blepharophimosis syndrome was highlighted by Beaconsfield *et al.*[7] in their large series of 101 cases of the syndrome.

This atypical case of blepharophimosis syndrome with arachnodactyly and uveal abnormalities highlights one more dimension to the many associated ocular and systemic developmental anomalies which have been described in conjunction with blepharophimosis syndrome.

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Sir,

Vitelliform Macular Dystrophy: A Cause of Macular Holes with Retinal Detachments

Vitelliform macular dystrophy, also known as Best's disease, is one of the heredo-familial macular disorders.[1] The mode of inheritance is probably autosomal dominant with irregular penetrance.[2] One of the remarkable clinical features of this disorder is the surprisingly good visual acuity, which does not correlate with the ophthalmoscopic findings. The classic ophthalmoscopic picture in vitelliform macular dystrophy is a yellow, round, slightly elevated structure resembling an egg yolk, usually found accidentally during routine fundus examination of children between 3 and 5 years old without any visual symptoms. This typical feature may not be encountered but instead other evolutionary stages may be found in which the characteristic structure either undergoes spontaneous resorption, forming the 'scrambled egg' stage with occasional cyst formation and a fluid level resembling a hypopyon giving rise to a so-called pseudohypopyon stage. Finally, the atrophic stage ensues with widespread retinal pigment epithelial (RPE) atrophy. The clinical diagnostic test in this disorder is a subnormal electrooculogram (EOG) with a light/dark ratio rarely exceeding 1.5, whereas the electroretinogram (ERG) is invariably completely normal.[2] Vision is commonly affected late in the disease unless a complication such as haemorrhage in the vitelliform structure or subretinal neovascularisation occurs.[2,3] Recent histopathological studies report flattened RPE cells with displacement of the nuclei towards the apex and diffuse deposition of abnormal lipofuscin granules.[4]

This report documents an unusual clinical presentation of vitelliform macular dystrophy: macular holes with retinal detachment.

Case Reports

Case 1. A 35-year-old man presented with a complaint of recent onset of metamorphopsia in the right eye. He had had defective vision in his left eye 7 years earlier with no specific medical history and no history of trauma to that eye. Visual acuity was 6/12 in the right eye and light perception in the left with normal intraocular pressures and normal anterior segments on biomicroscopy. Examination of the right eye disclosed a macular lesion typical of the pseudohypopyon stage of vitelliform macular dystrophy with normal retinal periphery (Fig. 1A). The left eye showed a large atrophic macular hole with widespread RPE atrophy and long-standing total retinal detachment with retinal thinning and cysts (Fig. 1B). No other peripheral retinal breaks were present.

Electrophysiological studies confirmed the diagnosis of vitelliform macular dystrophy, the EOG showing a subnormal response in both eyes (1.39 in the right eye and 1.15 in the left eye) with a normal ERG tracing. Fluorescein angiography revealed bilateral widespread RPE atrophy as evidenced by the window defects throughout the phases of the angiogram (Fig. 2), in addition to a small leaking hyperfluorescent spot in the right eye denoting subretinal neovascularisation.