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Sir,

Blepharospasm and Fuchs' Endothelial Dystrophy

We would like to report what appears to be an association between bilateral corneal decompensation (secondary to Fuchs' endothelial dystrophy) and severe blepharospasm. We have found this in 4 patients. This association has not previously been reported to our knowledge.

The 4 female patients were aged 52, 50, 49 and 54 years at the time of diagnosis of their blepharospasm; 2 were non-identical twins. In all patients the dystonia progressed to involve the lower facial muscles and efforts at treatment medically with tetrabenzine and clonazepam were unhelpful.

These patients were referred to an ophthalmologist when they developed symptoms of reduced vision. A diagnosis of Fuchs' endothelial dystrophy was made in all patients associated with early cataract and normal intra-ocular pressure. The time to diagnosis from the onset of the blepharospasm was a 22, 20, 16 and 12 years (mean 17.5 years) respectively. All patients developed bilateral corneal decompensation within 6 years of seeing the ophthalmologist and underwent corneal grafting. Histological examination confirmed the diagnosis of Fuchs' endothelial dystrophy.

To test the hypothesis that continued severe blepharospasm might be directly related to the endothelial changes, 10 patients who had suffered blepharospasm for at least 10 years (mean 12 years) and who were attending the botulinum toxin clinic underwent endothelial photographs. Endothelial photographs were also taken of uncomplicated age/sex-matched pre-operative cataract cases as a control. However, comparison of the endothelial morphology and cell count using a grid and counting technique revealed no significant difference between the groups.

Comment

Essential blepharospasm is an involuntary repetitive closure of the eyelids. It is usually bilateral and progressive, with half the patients having other involuntary movements.¹ Approximately 10% of affected patients have been reported to have a family history of some type of dystonia.² There have been reports of dystonias which appeared to be dominantly inherited.³

Fuchs' corneal endothelial dystrophy is a bilateral condition; women are affected more severely and more frequently than men. It may be inherited as an autosomal dominant trait with incomplete penetrance⁴ but there is no other evidence that the dystrophy is primary. The corneal endothelium transports water from the corneal stroma into

the anterior chamber. This produces a state of relative corneal dehydration and maintains its clarity. The fluid transport depends upon aerobic mechanisms and the required oxygen is thought to diffuse from the anterior chamber rather than from the tear film.⁵ However, corneal hypoxia induced by exposing the anterior corneal surface to nitrogen or placing a thick contact lens on the eye has been shown to cause the stroma to swell.⁶ Upon restoration of air to its anterior surface the cornea thins to a degree consistent with the restoration of endothelial pump action. Evaporation from the tear film with a resultant increase in its osmolality also draws water from the cornea.⁷

It has been reported that chronic corneal hypoxia may lead to permanent changes in corneal structure and function.⁶ It was possible that in the cases presented, the prolonged lid closure associated with blepharospasm may have reduced the oxygen available to the anterior surface of the cornea as well as reducing tear evaporation, leading to chronic hypoxia and endothelial damage. However, this hypothesis was not substantiated in the small series studied, which revealed no significant difference in the number or morphology of endothelial cells in those with severe blepharospasm compared with normals. That the endothelial cell counts were the same in the two groups also mitigates against the theory that there might be direct structural damage to the endothelium due to corneal flexing as a consequence of the blepharospasm.

It has been reported that where there is low endothelial cell density there is marked corneal swelling in response to hypoxia and on restoration of oxygen a slow recovery occurs.⁸ These patients did not show an earlier age of onset or faster rate of progression of the corneal decompensation than is usual in Fuchs' endothelial dystrophy.

The observation of 4 cases with both conditions, and particularly the 2 non-identical twins, raises the possibility of genetic linkage. Both conditions have been reported to be inherited as autosomal dominant pedigrees in some cases,^{3,4} although there was no previous family history that indicated prior cases and examination of the children was normal.

A toxic drug effect from treatment of the dystonia was also considered, but this complication has not been reported before with these medications despite wide usage.

We thought it important to report the co-incidence of Fuchs' endothelial dystrophy and blepharospasm, but from the evidence of a small series it seems likely to have been simply fortuitous. Moreover, it could not be shown that even severe blepharospasm had any compromising effect on the endothelial cells, because the corneal endothelial failure seemed to progress rapidly to bilateral corneal decompensation at the same rate as in other patients with Fuchs' endothelial dystrophy.

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