Clinical Features in Children with Posterior Polymorphous Corneal Dystrophy

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Introduction

Posterior polymorphous corneal dystrophy (PPCD) is a rare disorder involving metaplasia and overgrowth of the corneal endothelial cells. The area of abnormal endothelium can appear as vesicle-like lesions, band lesions, or diffuse opacities. It is usually bilateral, but the corneal abnormalities may be asymmetrical. Although early-childhood onset of ocular signs and symptoms and the slowly progressive nature of PPCD have been documented, little is known about the clinical manifestations in children with PPCD. Here, we describe the clinical features in children with characteristic vesicle- or band-like endothelial lesions in their first or second decade of life.

Patients & Methods

Seven unrelated Korean pediatric patients who were diagnosed by the presence of characteristic vesicular or band lesions at the level of Descemet’s membrane were included. Specular microscopy was performed on both eyes by using a non-contact specular microscope. The morphologic parameters in the corneal endothelium (endothelial cell density (ECD), average cell area, percentage of hexagonality (HA) and the coefficient of variation (CV) of cell area (standard deviation /mean x 100)) were investigated at the time of initial diagnosis and 3 years later.

Results

Slit-lamp examinations revealed vesicular lesions in one patient and horizontally parallel band-like endothelial lesions in 6 patients. A final visual acuity of more than 20/32 was achieved with appropriate refractive correction in all PPCD-affected eyes. Reduced ECD composed of enlarged endothelial cells was found. At the time of initial diagnosis, the mean ECD value in the PPCD-affected eyes was diminished to 1662.8 ± 601.1 (987–3058) cells/mm², compared with 3382.8 ± 179.0 (3125–3521) cells/mm² in the unaffected eyes. At 3 years after initial diagnosis, the endothelial assessments of the PPCD-affected eyes did not demonstrate any statistically significant differences in ECD, average cell area, CV and HA, compared with those at initial diagnosis.

Discussion

Our study provides additional evidence of the importance of awareness and treatment of high refractive error, especially in children with early-onset PPCD during the reversible period of amblyopia. Children with early-onset PPCD can gain good visual function by early appropriate refractive correction with occlusion therapy. Long-term monitoring of corneal endothelium should be required in pediatric patients with early-onset PPCD.