Atypical Granular Corneal Dystrophy: A Case Report

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ABSTRACT

Three cases of atypical granular corneal dystrophy (GCD) in the same family were reported. The patients had a history of progressively decreased visual acuity and foreign body sensation. The pathological findings showed hyaline deposits beneath the corneal epithelium and stroma. The patients had an unusual and atypical clinical presentation of GCD characterized by a rapid progression of clinical manifestation and early deterioration of visual acuity.

Key words: atypical granular corneal dystrophy (GCD), pedigree, *BIG-H 3* gene, penetrating keratoplasty.

INTRODUCTION

Granular corneal dystrophy (GCD) is predominantly an inherited condition manifested by opacities located centrally in the cornea in the first decade of life or in puberty. The clinical picture is bilateral discrete, grayish-white opacities that involve the superficial stromal layers of the cornea (Haddad et al.,1977). The patients present with a different appearance and slit lamp examination. Histopathologic studies with a light microscope show that the opacities are hyaline deposits in the stroma, within and beneath the epithelium staining with Masson trichrome, periodic acid-Schiff (PAS) and sometimes with Congo red (Jones and Zimmerman,1961; Folbrg et al.,1988). The rod-shaped bodies (electron-dense deposits) may be seen by electron microscope (Akiya and Brown,1970).

The lesions tend to aggregate, enlarge, and increase in number, spreading both peripherally and more deeply. A clear-zone around the corneoscleral limbus remains characteristically present. Central disk-shaped opacities are formed after the third or forth decade of life. Patients usually experience only gradual decrease in visual acuity and may maintain useful vision for a long time (Haddad et al.,1977). The objective of this study was to examine three cases of atypical GCD who presented with a rapid progression of clinical manifestation and early visual deterioration.

MATERIALS AND METHODS

A retrospective chart review of 3 patients seen between 1997 and 2001 at the cornea clinic, Maharaj Nakorn Chiang Mai Hospital, Chiang Mai University and given a diagnosis of atypical granular corneal dystrophy was performed. Informed consent was obtained from all patients.

Each case, the following preoperative data were collected: age, sex, complete ocular and family history, uncorrected visual acuity, slit-lamp biomicroscopy, and pathological results (case 1).