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In Vivo Laser Confocal Microscopic Findings in a Patient with Bietti Crystalline Dystrophy

Shinji Makino

Department of Ophthalmology, Jichi Medical University, Shimotsuke, Tochigi 329-0498, Japan

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Abstract Case Report

We investigated a patient with Bietti crystalline dystrophy (BCD) examined by confocal microscopy. Although multiple tiny refractile yellowish crystals were detected throughout the posterior pole, corneal crystals were not detected. Our findings may contribute to a better understanding of the BCD, regardless of the presence of corneal deposits.

Keywords: Bietti crystalline dystrophy, confocal microscopy.

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Introduction

Bietti crystalline dystrophy (BCD) is characterized by deposits of crystals in the marginal cornea and the paracentral and peripapillary retina [1-3]. One third to one half of BCD cases display corneal crystals recognizable during slit-lamp biomicroscopy [3]. Previously, specular microscopy was shown to detect crystalline deposits at the limbus of patients with BCD [4, 5]. However, there is one report documented the confocal microscopic features of corneal crystals in

BCD [6]. Herein, we investigated a patient with BCR examined by confocal microscopy.

CASE REPORT

A 37-year-old woman was referred to our hospital. She complained of visual impairment, night blindness and central scotomas. Her best-corrected visual acuity was 1.2 in the right eye and 0.3 in the left eye. Fundus examination revealed intraretinal crystals in the posterior pole and midperipheral retina associated with chorioretinal atrophy (Figure 1).

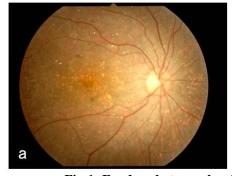




Fig-1: Fundus photographs of the (a) right and (b) left eyes

Note the multiple tiny refractile yellowish crystals throughout the posterior pole.

Fluorescein angiography demonstrated a transmission defect with granular hyperfluorescence

and hypofluorescent areas corresponding to atrophy or loss of choriocapillaris in both eyes (Figure 2).

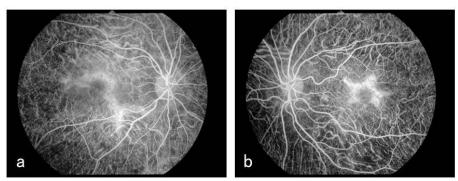


Fig-2: Fluorescein angiography of the (a) right and (b) left eyesNote the granular hyperfluorescence and hypofluorescent areas

The slit-lamp biomicroscopic image showed no hyperreflective plaques beneath the corneal epithelium (Figure 3).

The confocal microscopy was performed using Heidelberg Retina Tomograph II Rostock Cornea Module. Although several confocal images from central and peripheral cornea were taken, hyperreflective corneal crystals were not detected (Figure 4).

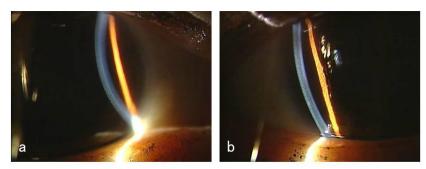


Fig-3: Anterior segment photographs of the (a) right and (b) left eyes

Note no crystalline deposits beneath the corneal epithelium

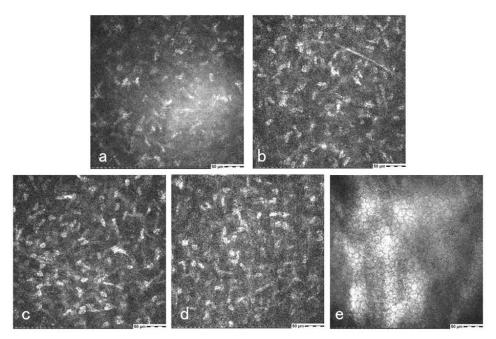


Fig-4: In vivo laser confocal microscopic findings

a: corneal epithelium; b: corneal superficial stroma; c: corneal stroma; d: corneal deep stroma; e: corneal endothelium

DISCUSSION

The corneal crystals in BCD are very fine and located mainly in subepithelial and superficial anterior stroma near the limbus. It has been reported that cornea crystalline deposits are not present or mentioned in half of the reported cases of BCD [3]. Therefore, the idiopathic retinal disorders characterized by retinal crystalline deposits are referred to as BCD, regardless of the presence of corneal deposits.

Confocal enables microscopy high magnification, in vivo examination of the corneal layers at the cellular level. It is a noninvasive technique that allows the early diagnosis of pathologic changes in the cornea. There is one report documented the confocal microscopic features of corneal crystals in BCD [6]. According to their report, needle-shaped or rod-shaped materials up to 40 mm in length and 4-8 mm in width were detected within the epithelium and anterior stroma close to the limbus. Although several confocal images from central and peripheral cornea were taken, hyperreflective corneal crystals were not detected in this patient.

CONCLUSIONS

Although our report is based solely on a patient with BCD, confocal microscopic findings may contribute to a better understanding of the BCD without the corneal deposits.

Disclosure

The author declares no conflict of interest.

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