

Longitudinal changes in retinal crystals in a patient with Bietti crystalline dystrophy

dystrophy

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Abstract: We present a long-term follow-up of a patient with Bietti crystalline dystrophy. The clinical findings were documented over a period of 4 years. The most striking features were crystalline deposits in the retina that were formed de novo with old ones replaced by chorioretinal atrophy. The visual fields were affected progressively during the course. Bietti crystalline retinopathy is a progressive retinal disease characterized by retinal crystals gradually replaced by atrophy of the retinal pigment epithelium and gradual constriction of visual fields. Our findings may contribute to a better understanding of the natural course of Bietti crystalline dystrophy.

Keywords: Bietti crystalline dystrophy, optical coherence tomography, longitudinal study.

INTRODUCTION

Bietti crystalline dystrophy (BCD) is characterized by deposits of crystals in the marginal cornea and the Para central and peripapillary retina [1-5]. Few reports have documented the longitudinal changes in clinical features of BCD [1-4]. We present long-term fundus photographic documentation in a patient with BCD.

CASE REPORTS

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. Slit-lamp examination was unremarkable. Fundus examination revealed intraretinal crystals in the posterior pole and midperipheral retina associated with chorioretinal atrophy (Figure 1).

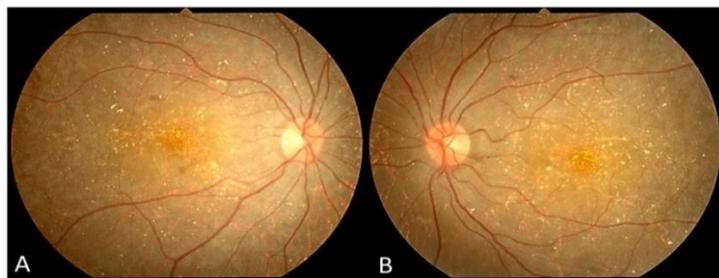


Fig 1: Fundus photographs of the (A) right and (B) left eyes at the initial visit.

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

Fluorescein angiography demonstrated a transmission defect with granular hyper fluorescence and hypo fluorescent areas corresponding to atrophy or loss of choriocapillaris in both eyes (Figure 2).

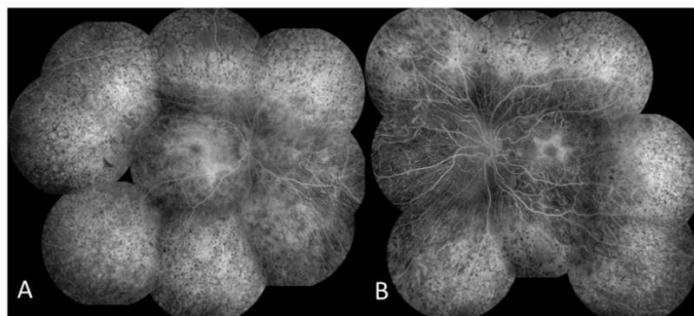


Fig 2: Fluorescein angiography of the (A) right and (B) left eyes at the initial visit.

Note the granular hyper fluorescence and hypo fluorescent areas

The anterior segment optical coherence tomography showed no hyper reflective plaques beneath the corneal epithelium (Figure 3).

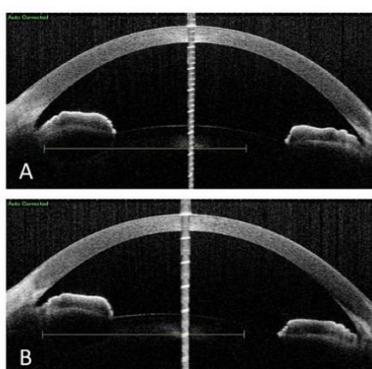


Fig 3: Anterior segment optical coherence tomography of the (A) right and (B) left eyes at the initial visit.

Note no hyper reflective plaques beneath the corneal epithelium

Goldmann perimeter was performed and showed para central scotoma with outer borders intact in both eyes (Figure 4).

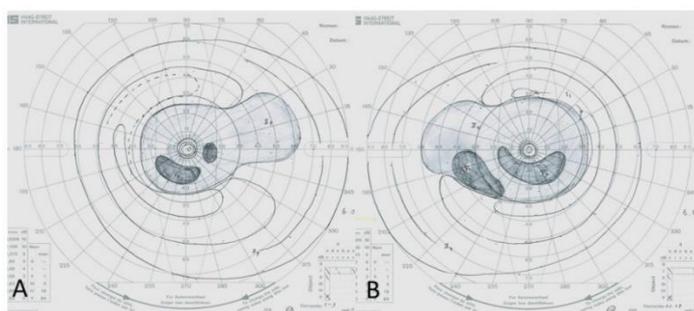


Fig 4: Goldmann perimetry of the (A) right and (B) left eyes at the initial visit.

Note absolute and large relative scotomas.

The patient was followed without any treatment. Over a 4-year follow up period , the most striking features were deposits in the retina that were formed de novo with old ones replaced by choroidal atrophy

(Figure 5). In addition, funduscopy revealed increased visibility of the choroidal vessels. The visual fields were affected progressively during the course (Figure 6). However, her visual acuity maintained 1.2 in the right eye and 0.3 in the left eye.

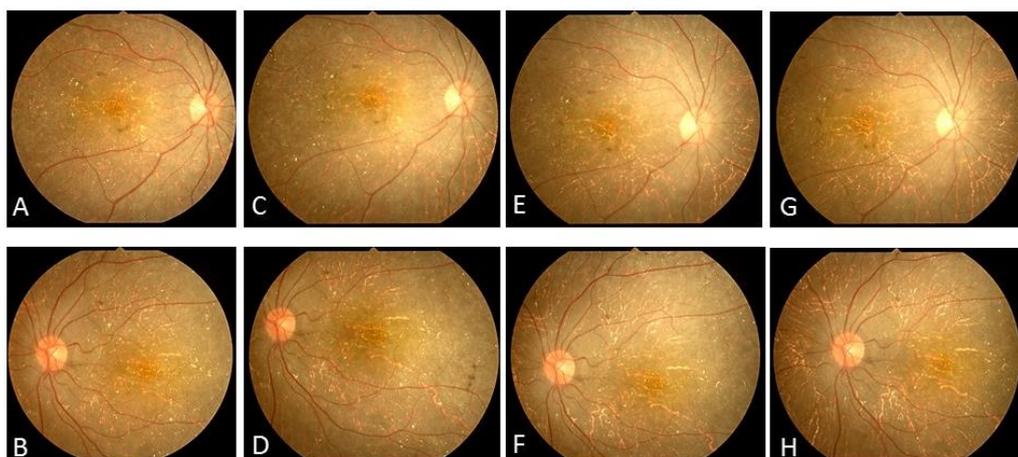


Fig 5: Fundus photographs of the right (top) and left (bottom) fundus over a 4-year period.

Note crystalline deposits gradually replaced by choroidal atrophy in advanced stage.

A, B: 1 year after initial visit; C, D: 2 years after initial visit; E, F: 3 years after initial visit; G, H: 4 years after initial visit.

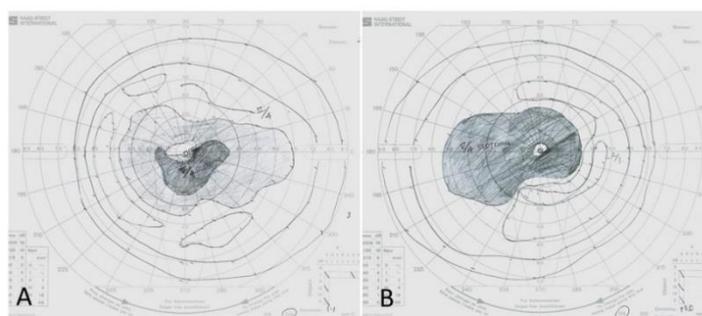


Fig 6: Goldmann perimetry of the (A) right and (B) left eyes at the final visit.

The visual fields revealed an extension of the central scotomas.

DISCUSSION

From the point of view of individual cases, there are few reports having long-term photographic documentation of 30 years [2], 25 years [3], 20 years [4], and 16 years [1]. Mansour *et al.*; described that crystals were apparent in the second decade with mild functional deficit. In the fourth decade, functional impairment becomes severe with marked chorioretinal degeneration. They also reported that OCT demonstrated preferential crystal accumulation in the inner retina. These crystals commonly disappear and are replaced by areas of chorioretinal atrophy. On the other hands, Li *et al.*; [5] evaluated multiple imaging modalities in different stages of BCD. According to their report, OCT changes were observed predominantly adjacent to the retinal pigment epithelium and the photoreceptors in the early stage. In the advanced stage, more extensive atrophy was seen. BCD is a progressive retinal disease characterized by retinal crystals gradually replaced by atrophy of the retinal pigment epithelium and gradual constriction of visual fields. Although our report is based solely on a

patient and only a 4-year follow-up period, our observation may contribute to a better understanding of the natural course of BCD.

Disclosure

The authors have no conflicts of interest to disclose.

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