A Precious Fundus- Crystalline Retinopathy

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INTRODUCTION

Bietti's dystrophy (AR) is characterized by deposition of crystals in the retina and superficial peripheral cornea. Crystalline corneoretinal dystrophy is an uncommon hereditary condition first reported by Bietti in 1937 as a corneoretinal degeneration.¹ The hallmark of the disease was the crystalline characteristics of the retinal spots as well as those at the corneal limbus.² The characteristic retinal crystals are observed in all cases, but the associated corneal dystrophy is not a consistent feature of the disease.³ Other reported features include progressive night blindness, constriction of visual field, subnormal ERG and EOG and defective colour vision in advanced cases.¹⁻³

This condition is much more common in East Asians, particularly Chinese, than other ethnicities.⁴ We would like to present a case of Bietti's dystrophy discovered incidentally on routine fundus examination of a 36-year-old female who presented to our outpatient department with blurred vision and mild pain in both eyes.

PRESENTATION OF CASE

A 36-year-old female patient came with decreased vision for distance, mild eye pain in both eyes along with watering for three months. She was not diabetic. She was diagnosed with hypertension six months back, on treatment with tab Envas. There was history of occasional respiratory distress for past two years, on T. Deriphylline. There was no history of eye trauma or surgery. There was no history of any relevant metabolic disorder.

On examination, anterior segment findings were within normal limits in both eyes. Intraocular pressure was 17 mmHg in right eye and 18 mmHg in left eye. She underwent cycloplegic refraction followed by post-mydratic test. Distance vision was correctable to 6/6p in right eye (with -0.5 DS/-0.5 DC x 20) and 6/9 (plano) in left eye; near vision was N6 in both eyes. Her colour vision was normal in right eye (16/16) but abnormal in left eye (3/16).

On dilated fundus exam, both eyes showed normal disc and vessels. Entire retina showed greyish discolouration with multiple small glistening whitish deposits in both eyes. In addition, left eye showed a pigmented scar close to macula. This remarkable fundus picture suggested possibilities for crystalline retinopathy.

Patient had difficulty in night vision. There was no history of similar complaints in any family member. Patient's corneas revealed no crystals on slit lamp evaluation. A provisional clinical diagnosis of 'Both eyes - Bietti's crystalline retinopathy with maculopathy in left eye' was made.

OCT macula revealed normal retinal thickness in both eyes. Perimetry attempted but test had to be aborted as patient was unable to visualise the light stimuli appearing on the visual field even on repeated attempts. Her electroretinogram report showed minimally reduced scotopic and photopic responses in both eyes.

Patient was counselled about eye problem, advised to continue spectacles and kept on follow up. On last follow-up, her vision had dropped marginally- unaided vision was 6/24 in right eye and 6/36 in left eye, improving to 6/18 and 6/24 with pinhole in right and left eyes respectively.

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Image 1. Fundus Photograph- Both Eyes

Image 2.- OCT Pictures- Both Eyes

Image 3. Slit Lamp Photograph Both Eyes

DIFFERENTIAL DIAGNOSIS

Bietti's retinopathy, calcified macular drusen, idiopathic parafoveal telangiectasias and long-standing retinal detachment.

PATHOLOGICAL DISCUSSION

This case is being highlighted as it is relatively rare. The mechanism of Bietti's crystalline corneoretinal dystrophy may be linked to an error in systemic lipid metabolism. Biopsy specimens from patients have demonstrated crystals resembling cholesterol or cholesterol ester and complex lipid inclusions in corneal and conjunctival fibroblasts.⁵ the rate of progression is variable- specific treatment not currently available. Young adults with slowly progressive visual impairment constitute the typical case.

Signs According to Kanski's Clinical Ophthalmology

- Superficial peripheral corneal crystals.
- Numerous fine yellow-white crystals scattered throughout posterior fundus, followed by localised atrophy of RPE and choriocapillaris at macula.
- Diffuse atrophy of choriocapillaris subsequently develops, with decrease in size and number of crystals.
- Gradual confluence and expansion of atrophic areas into periphery, leading to diffuse chorioretinal atrophy in end-stage disease.

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