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3

Late-Stage Hydroxychloroquine-Associated Retinopathy

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Section V, case studies including one or two patients are not subject to IRB oversight.

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A 58-year-old woman with rheumatoid arthritis presented with six months of central scotoma and photosensitivity. She had been treated with hydroxychloroquine 400 mg daily for five years, then 600 mg daily for another five years. Visual acuity was 20/25 and 20/30 in the right and left eye, respectively. Fundus examination and fundus autofluorescence revealed a characteristic “bull’s-eye” pattern of parafoveal pigment change in both eyes [representative left eye shown in color fundus photograph (a), and fundus autofluorescence image (b), with a dark hypofluorescent ring indicating atrophy of the retinal pigment epithelium; white arrows marking “bull’s eye”]. Retinal optical coherence tomography was positive for the “flying saucer sign” — loss of the ellipsoid zone (orange arrow in c), representing photoreceptor damage, in a ring pattern around the fovea (bracket in c). Visual fields revealed a paracentral scotoma in both eyes (d; central visual field overlaid to demonstrate field loss corresponding to the area of retinal pigment epithelium atrophy, with darker-shaded areas of visual field corresponding to decreased sensitivity to light stimuli). A diagnosis of hydroxychloroquine-associated retinopathy was made and the medication was stopped. Retinal toxicity is a known and devastating side effect of hydroxychloroquine due to increased cumulative and daily doses [1]. This patient’s 600 mg daily dose exceeded the recommended real-weight-adjusted maximum dosage of 5 mg/kg/day. Her cumulative dosage of 1,825 g also far exceeded the 1,000-g threshold that has been associated with increased risk of retinal toxicity [2]. Although hydroxychloroquine-associated retinopathy is irreversible, late effects, such as the bull’s-eye maculopathy seen here, are preventable. Through appropriate referral for baseline ophthalmologic examination as well as annual screening starting at five years of therapy, toxicity can and must be detected early, avoiding presentations like this one [1].

Author Contributions

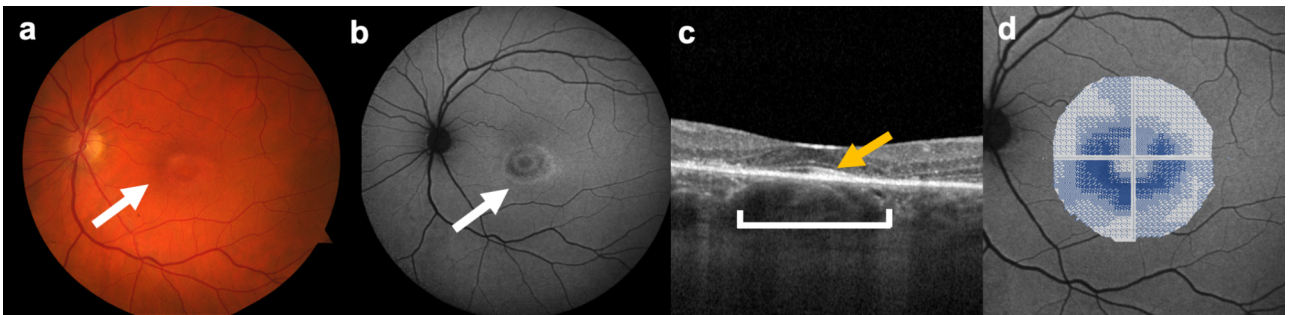
M.K.M. contributed to study conception and design and analysis and interpretation of data, drafted and revised the article, and approved the final version to be published.

D.A.B. contributed to study conception and design, data acquisition, and analysis and interpretation of data, substantially revised the article, and approved the final version to be published.

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