

Hydroxychloroquine Ocular Toxicity: Lessons Learned

JEFF C. RABIN, OD, MS, PhD, Professor and Assistant Dean for Graduate Studies, Research and Assessment, and Chief, Visual Neurophysiology Service, University of the Incarnate Word Rosenberg School of Optometry; KIRSTI RAMIREZ, OD, Optometric Resident, University of the Incarnate Word Rosenberg School of Optometry, San Antonio, Texas, USA. Address correspondence to Dr. J.C. Rabin, Chief, Visual Neurophysiology Service, University of the Incarnate Word Rosenberg School of Optometry, 9725 Datapoint Drive, San Antonio, Texas 78229, USA. E-mail: rabin@uiwtx.edu. De-identified case reports with 3 or fewer patients are exempt from formal review by the University of the Incarnate Word Institutional Review Board (UIW IRB) and do not require informed consent although the patient provided verbal consent. This case report satisfied ethical requirements for submission in accord with the UIW IRB. (J Rheumatol 2019;46:1640–1; doi:10.3899/jrheum.181375)

Hydroxychloroquine (HCQ) is efficacious for various diseases^{1,2}, but can produce “bull’s-eye” retinopathy that decreases vision even after discontinuance^{3,4}. Retinopathy can be present in 7.5% of patients after 5 or more years of HCQ treatment, increasing to 20% after 20 years². In 2016, the American Academy of Ophthalmology published new recommendations for the evaluation of HCQ, emphasizing dosage based on body weight (≤ 5 mg/kg)⁵. We describe a case in which dosage significantly exceeded guidelines.

A 50-year-old Hispanic female was referred from a retinal specialist to our Visual Neurophysiology Service. Her weight was 115 pounds. She was receiving HCQ for systemic lupus erythematosus (200 mg twice daily) since 2000 but discontinued in 2017 because of toxicity. Total dosage was 2482 g.

Maximum recommended dosage is $5 \text{ mg/kg} \times 0.454 \text{ kg/pound} = 2.27 \text{ mg/pound}$. The maximum recommended dosage for the patient is $115 \text{ pounds} \times 2.27 \text{ mg/pound} = 261 \text{ mg/day}$. Acuity was 20/25, but central visual fields showed paracentral scotomas, cone-specific color vision was decreased, and multifocal electroretinograms (mfERG), which assess retinal function from multiple sites, showed signal reduction and delay. Optical coherence tomography (OCT) showed full thickness and inner retinal thinning. Figure 1a shows fundus views with a bull’s-eye pattern of photoreceptor and retinal pigment epithelium degeneration more apparent with fundus autofluorescence (FAF; Figure 1b), which shows active degeneration in white and cell death in black.

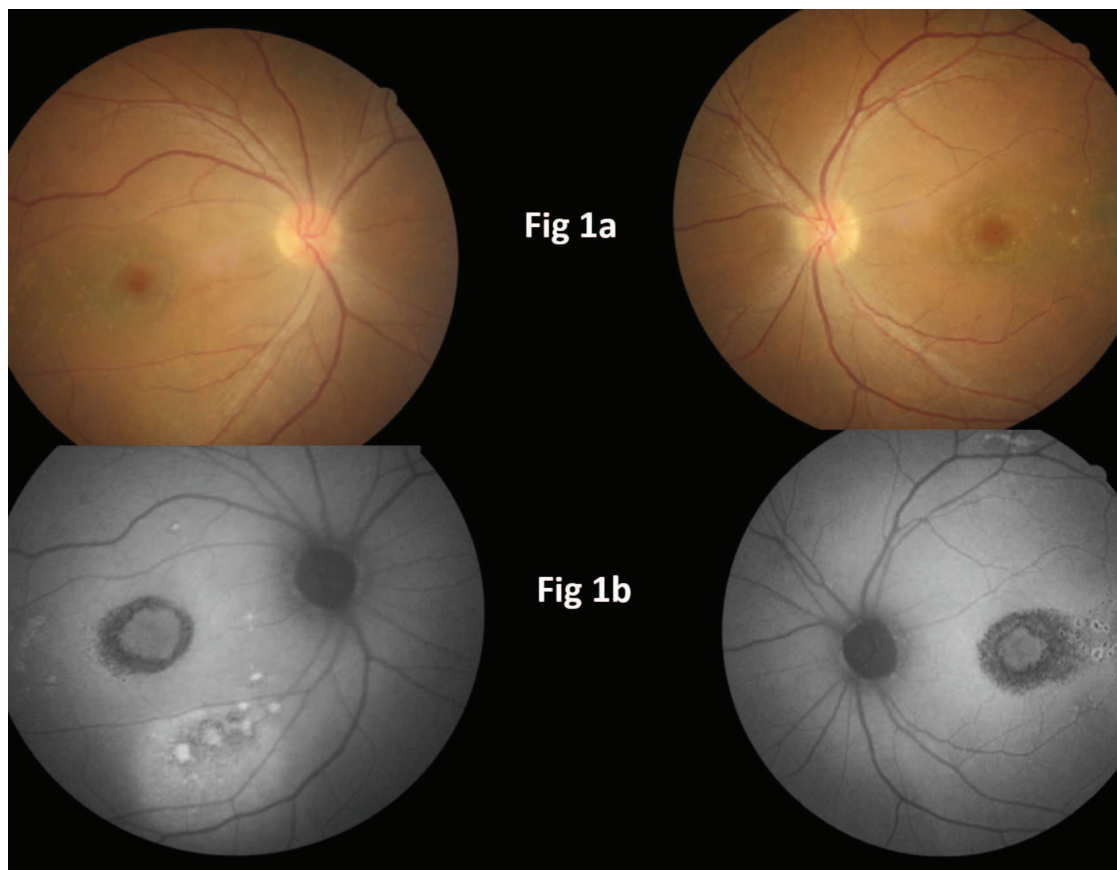


Figure 1. (a) Fundus photos of the posterior poles showing bull’s-eye retinopathy typically only visible with standard fundus evaluation after advanced retinopathy has developed. (b) Fundus autofluorescence is a noninvasive imaging technique that reveals areas of active degeneration in white representing accumulation of the fluorophore lipofuscin. Dark areas of hypofluorescence are associated with degeneration of the outer retina. Central visual fields, spectral domain optical coherence tomography, and multifocal electroretinograms also show hydroxychloroquine toxicity prior to visible fundus changes and are essential for early and periodic screening.

Weight-based dosage and early screening (central fields, OCT, FAF, mfERG) are essential to prevent HCQ toxicity, particularly with risk factors: small stature, high total dosage, diminished renal function, concomitant tamoxifen use, and/or retinal disease⁵. Interprofessional collaboration is essential to optimize patient outcomes.

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