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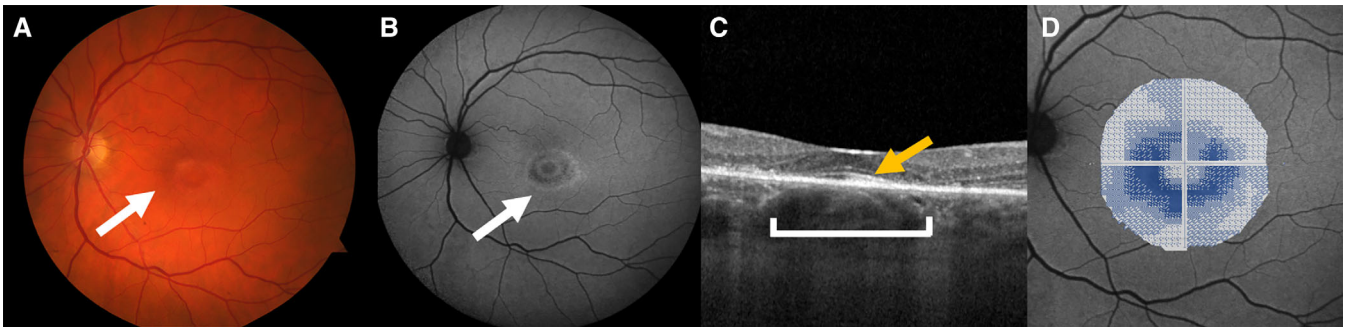
Clinical Images: Late-stage hydroxychloroquine-associated retinopathy



The patient, a 58-year-old woman with rheumatoid arthritis, presented with 6 months of central scotoma and photosensitivity. She had been treated with hydroxychloroquine at a dosage of 400 mg daily for 5 years, then 600 mg daily for another 5 years. Visual acuity was 20/25 and 20/30 in the right eye and left eye, respectively. A 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"). The retinal optical coherence tomography result 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 1825 g also far exceeded the 1000-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 5 years of therapy, toxicity can and must be detected early, avoiding presentations like this one (1).

Under the University of Michigan Human Research Protection Program Operations Manual Part 4 Section V, case studies including one or two patients are not subject to institutional review board oversight.

Author disclosures are available at https://onlinelibrary.wiley.com/action/downloadSupplement?doi=10.1002%2Facr2.11502&file=acr211502-sup-0001-Disclosureform.pdf.

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