A rare case of symptomatic hydroxychloroquine maculopathy from short duration of 7 months treatment for rheumatoid arthritis
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Abstract: A 77-year-old female with a seven-month duration of hydroxychloroquine treatment present with hydroxychloroquine maculopathy, confirmed using Humphrey threshold visual field 10-2 testing and spectral domain optical coherence tomography.

I. Case History
1. 77-year-old white female who weighs 94 pounds and is 63 inches in height
2. Chief complaint: darkening of the vision OU, noted for the past 7-8 months
3. Ocular history:
   - nuclear sclerosis cataracts OU, perimacular drusen OS, orbital fracture OD
4. Medical history:
   - rheumatoid arthritis, diagnosed in May 2012, diabetes type 2, coronary artery disease, hyperlipidemia, hypothyroidism
5. Medications
   - hydroxychloroquine sulfate, 400 mg per day initiated January 2014
   - amitriptyline HCl, aspirin, nortriptyline HCl, paroxetine HCl
6. Family ocular history unremarkable

II. Pertinent findings
1. BCVA: 20/40+1 OD, 20/40-1 OS
2. Anterior segment: mild nuclear sclerotic and anterior cortical cataracts OU
3. Dilated posterior segment examination
   - Optic discs: no pallor or elevation OU
   - Retinal blood vessels exhibit good caliber OU
   - Macula: moderate retinal pigment epithelium (RPE) mottling and pigment changes with few scattered drusen and absent foveal reflexes OU
4. Cirrus spectral-domain optical coherence tomography (SD-OCT) – Macular Cube and HD 5-Line Raster
   - Thin parafoveal retinal layers 360 degrees OD, and superiorly and temporally OS
   - Mildly depressed foveal contour, RPE clumping, disruption of the ellipsoid portion of the inner segment (EPIS) and cones outer tips (COST) lines in the parafoveal area, and thinning of the outer retinal layers
5. Humphrey visual field 10-2 SITA Standard
   - Dense parafoveal ring scotoma OD>OS
6. D-15 color vision
   - Tritan defect OD, OS
7. Multifocal electroretinogram (mfERG) is pending

III. Differential diagnosis
1. Primary differential diagnosis: hydroxychloroquine maculopathy, due to the characteristic appearance of the Humphrey visual field 10-2 and the macular SD-OCT
2. Other differential diagnoses: age-related macular degeneration, cone and rod dystrophies, classical cone dystrophies, neuronal ceroid lipofuscinosis, Stargardt's disease, and
fenestrated sheen macular dystrophy

IV. Diagnosis and discussion
1. Hydroxychloroquine is used to treat inflammatory diseases such as systemic lupus erythematosus, rheumatoid arthritis, and other inflammatory and dermatologic conditions.
2. Hydroxychloroquine binds to melanin in the RPE, leading to toxicity that includes a characteristic bilateral bull's eye macular appearance, although functional visual loss may precede RPE changes.
3. Cumulative dose of hydroxychloroquine is the primary risk factor for maculopathy with the risk of toxicity increasing toward 1% after 5-7 years of use, or a cumulative dose of 1000 g.
4. Special caution should be used in individuals taking hydroxychloroquine in excess of 6.5 mg/kg/day, calculated based on lean body weight.
5. Other complicating factors for hydroxychloroquine maculopathy are obesity, renal or hepatic failure, existing retinal pathology, previous chloroquine or hydroxychloroquine usage, and age greater than 60 years.
6. The case being discussed is unusual due to the severity and characteristic nature of hydroxychloroquine maculopathy detected in a patient with a short duration of treatment using standard dosages who is not obese, does not have renal or hepatic failure, and has no previous chloroquine or hydroxychloroquine usage.

V. Treatment and management
1. The only treatment for hydroxychloroquine maculopathy is timely cessation of hydroxychloroquine treatment, which depends on early detection.
2. If hydroxychloroquine is stopped before maculopathy becomes advanced, there is possibility of regression; however, maculopathy may also progress even after the medication is discontinued.
3. Current recommendations include a baseline screening of patients treated with hydroxychloroquine that includes biomicroscopy, Humphrey threshold visual field 10-2 testing, and an objective test such as SD-OCT, mfERG, or fundus autofluorescence, followed by annual screening in all high-risk patients starting 5 years from initiation of treatment.

VI. Conclusion
1. While rare, visual loss from hydroxychloroquine toxicity is a very real risk that should not be ignored even in patients who have taken hydroxychloroquine for less than 5 years at standard dosages in the absence of obesity and renal or hepatic failure.
2. Functional vision loss, including reduced best-corrected visual acuities, color vision deficits, and symptoms of dimming can severely affect a patient's activities of daily living even in new cases of hydroxychloroquine maculopathy.

References