DJO Digital Journal of Ophthalmology
www.djo.harvard.edu

Grand Rounds

A 61-year-old man with cystoid macular edema and chorioretinal folds after cataract surgery

Andrew Lee, MD, and **Sandra R Montezuma, MD**
Author affiliations: Department of Ophthalmology and Visual Neuroscience, University of Minnesota

History

A 61-year-old white man was referred to the University of Minnesota Department of Ophthalmology with a diagnosis of cystoid macular edema (CME) following bilateral cataract surgery 2 months prior. Chorioretinal folds and CME were present on fundus examination and optical coherence tomography (OCT). His past medical history included diabetes mellitus for 18 years, hypertension for 5 years, and heart disease with bypass surgery. His most recent glycosylated hemoglobin test (HbA1C) was 8.3 (normal, <5.7%). The patient reported improvement in visual acuity without correction after surgery with no flashes or floaters.

His preoperative refraction was +7.00 +2.50 ×180 correcting to 20/30 in the right eye and +7.75 +1.50 ×029 correcting to 20/30 in the left eye. A 36 D AcrySof SN60WF (Alcon, Fort Worth, TX) intraocular lens (IOL) was implanted in each eye.

Examination

On examination, uncorrected visual acuity was 20/60 in the right eye and 20/40 in the left eye with no pinhole improvement. Intraocular pressure was 19 mm Hg in the right eye and 18 mm Hg in the left eye. Motility was full, with no ptosis or proptosis. Anterior segment examination was unremarkable. A posterior chamber IOL was present and in good position in both eyes. Fundus examination (Figure 1) in each eye showed macular edema and a pink optic nerve, with slightly blurred disk margins. There was mild tortuosity of the vessels, dilation of the venules, and alternating dark and light yellowish streaks in the posterior pole corresponding to chorioretinal folds.

Ancillary Testing

Optical coherence tomography showed chorioretinal folds and diffuse intraretinal cystoid changes (Figure 2). In addition to chorioretinal folds, evidence of macular edema was apparent on fluorescein angiography (Figure 3). A-scan ultrasonography revealed an axial length of 19.53 mm in the right eye and 19.48 mm in the left eye. B-scan ultrasonography showed increased retinal-choroidal-scleral thickness in each eye (2.09 mm right eye and 2.36 mm left eye; Figure 4).

Treatment

Our patient had CME that could have been secondary to diabetic macular edema (DME), cataract surgery,[1] nanophthalmos,[2,3] or a combination of the above. We decided to treat conservatively with Nepafenac ophthalmic suspension and prednisolone acetate ophthalmic suspension 1%. The patient's symptoms responded partially to the topical medications, with a persistence of CME and chorioretinal folds. Unfortunately, the patient has been lost to follow-up. Additional considerations were anti-VEGF injections for possible DME, or scleral windows if his condition worsened and did not respond adequately to local treatment.

Differential Diagnosis

The differential diagnosis of chorioretinal folds includes hypotony, posterior scleritis, high hyperopia, nanophthalmos, choroidal tumor or detachment, orbital tumor, choroidal neovascular membrane, age-related macular degeneration, autoimmune diseases, optic disk swelling and idiopathic.[4,5]

Published August 29, 2017.

doi:10.5693/djo.03.2017.02.002

Correspondence: Sandra R Montezuma, MD, Ophthalmology Department and CIinic, 9-317C Phillips-Wangensteen Building, 516 Delaware Street S.E., 9th FIoor, Minneapolis MN 55455 (email: smontezu@umn.edu).

digital journal of ophthalmology, vol. 23

digital journal of ophthalmology, vol. 23

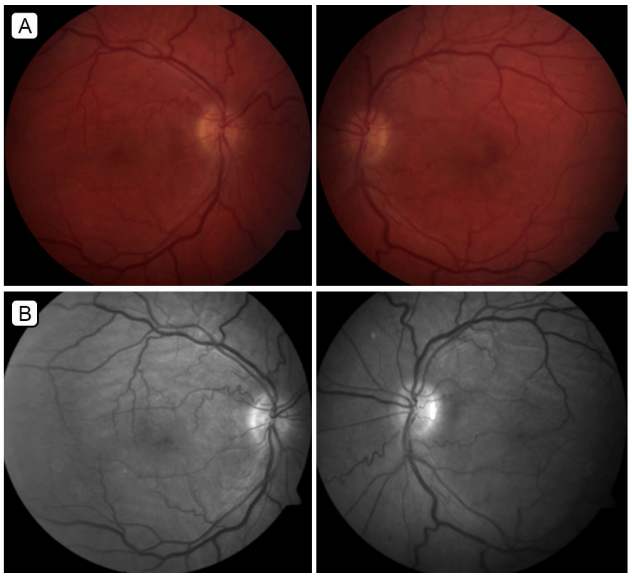

Figure 1. Fundus photographs (A) and red-free photographs (B) showing chorioretinal folds, venular dilation, and vessel tortuosity.

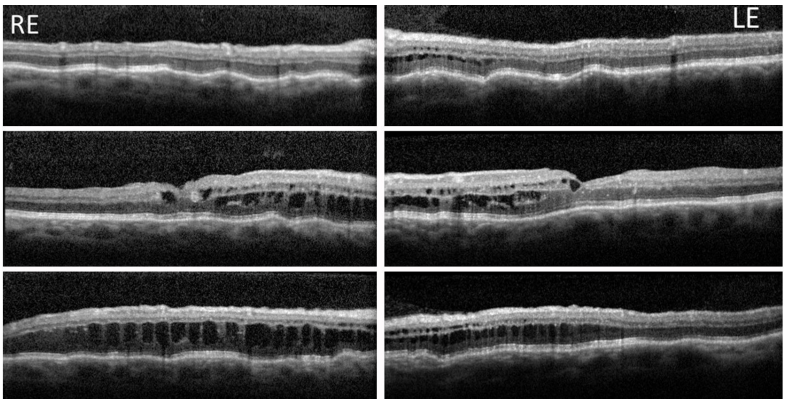

Figure 2. Optical coherence tomography showing choroidal folds and diffuse intraretinal cystoid changes in both eyes, with a central macular thickness of 441 µm in the right eye and 387 µm in the left eye.

Diagnosis and Discussion

In our patient, the finding of chorioretinal folds prompted further evaluation of the differential diagnosis. His cataract surgeries were performed outside our institution; hence in our initial evaluation we had no preoperative records. The patient had CME but did not report a history of diabetic retinopathy or DME. The patient's high hyperopia was discovered after operative and preoperative records became available. Ancillary testing revealed a short axial length and an increased retinal-choroidal-scleral thickness (Figure 4), resulting in a diagnosis of nanophthalmos.

Nanophthalmos is a rare condition; a preliminary diagnosis can be made with a measurement of axial length <21 mm.[6] Presentation is normally bilateral, and patients can have slight enophthalmos and narrow palpebral fissures.[7] Other characteristics include severe hyperopia, increased retinal-choroidal-scleral thickness, a crowded anterior chamber, and chorioretinal folds.[5,6,8,9] The crowded anterior chamber of nanophthalmos is of particular concern, because it predisposes the patient to acute angle-closure glaucoma.[10] Additional complications arise from a thick scleral collagen, which can result in choroidal effusion, retinal detachment, and cystoid macular edema.[11]

digital journal of ophthalmology, vol. 23

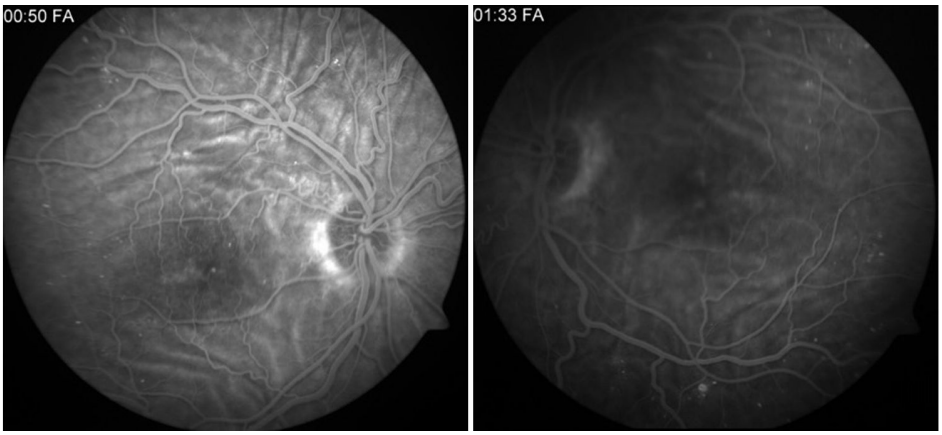

Figure 3. Fluorescein angiography showing alternating hypo- and hyperfluorescent lines in both eyes, corresponding to chorioretinal folds. There was mild early hyperfluorescence in the macula that increased during the course of the study, corresponding to macular edema. Pinpoint hyperfluorescent spots correspond to microaneurysms.

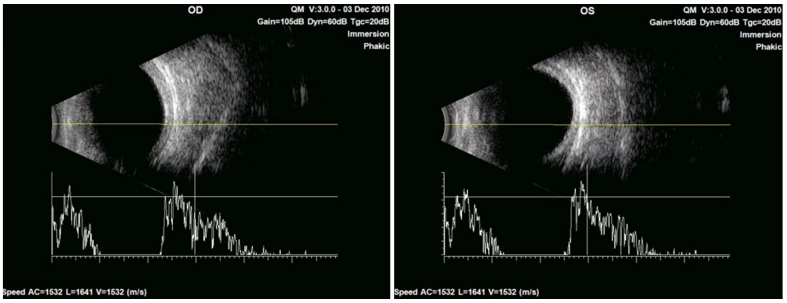

Figure 4. Ultrasound imaging showing retinal-choroidal-scleral thickness of 2.09 mm in the right eye and 2.36 mm in the left eye. A-scan overlay showing the short axial lengths of each eye, the right eye measuring 19.53 mm and the left eye measuring 19.48 mm.

Most cases of nanophthalmos are spontaneous, although familial types exist. Cases of autosomal recessive (AR) and dominant (AD) nanophthalmos have been attributed to mutations on chromosome 11.(7) AR nanophthalmos has been reported to involve the *MFRP* (membrane frizzled-related protein) gene. The MFRP protein is expressed in the retinal pigment epithelium and ciliary body and is important to later stages of ocular development.[6,7] It is thought that mutations in *MFRP* result in decreased ocular growth, leading to crowding and thickening of the retina and choroid, abnormal foveal development, and chorioretinal folds.[6–8,12] A gene *TMEM98* (transmembrane protein 98) on chromosome 17 has also recently been implicated in a form of AD nanophthalmos.[13,14] Nanophthalmos can additionally be present in less common conditions such as oculo-dento-digital and Kenney-Caffey syndromes.[7]

Nanophthalmos patients are at increased risk for cataract surgery complications.[2,9,15] Two recent studies have reported the complication rate of cataract surgery in nanophthalmos patients: Steijns et al reported a 27.9% complication rate (n = 43), whereas Day et al reported 15.5% complication rate (n = 103).[2,15] Surgical complications in nanophthalmos include choroidal effusion, zonular dehiscence, uveitis, malignant glaucoma, and cystoid macular edema.[2,9,15,16] It has been suggested that retinal-choroidal-scleral thickness be measured preoperatively in hyperopic eyes at risk of glaucoma to properly diagnose nanophthalmos and to allow more careful planning of surgery.[9] Preventative measures to be considered prior to cataract surgery include laser iridotomy, iridoplasty, trabeculectomy, and creation of scleral windows.[16] The use of intravenous mannitol to maintain pressure during surgery might also be considered.

References

1. Rotsos T, Moschos M. Cystoid macular edema. Clin Ophthalmol 2008;2:919-30.

2. Steijns D, Bijlsma W, Van der Lelij A. Cataract surgery in patients with nanophthalmos. Ophthalmology 2013;120:266-70.
3. MacKay C, Shek M, Carr R, Yanuzzi LA, Gouras P. Retinal degeneration with nanophthalmos, cystic macular degeneration, and angle closure glaucoma. a new recessive syndrome. Arch Ophthalmol 1987;105:366-71.
4. Olsen T, Palejwala N, Lee L, Bergstrom C, Yeh S. Chorioretinal folds: associated disorders and a related maculopathy. Am J Ophthalmol 2014;157:1038-47.
5. Yalçindag N, Atilla H, Batıoğlu F. Optical coherence tomography findings of retinal folds in nanophthalmos. Case Rep Ophthalmol Med 2011;2011:491894.
6. Sundin O, Dharmaraj S, Bhutto I, et al. Developmental basis of nanophthalmos: MFRP is required for both prenatal ocular growth and postnatal emmetropization. Ophthalmic Genetics 2008;29:1-9.
7. Traboulsi, E. Nanophthalmos. In: Silva, E.; Sundin, O., editors. Genetic Diseases of the Eye Oxford University Press; 2012. chap 4
8. Serrano JC, Hodgkins PR, Taylor DS, Gole GA, Kriss A. The nanophthalmic macula. Br J Ophthalmol 1998;82:276-9.
9. Wu W, Dawson D, Sugar A, et al. Cataract surgery in patients with nanophthalmos: results and complications. J Cataract Refract Surg 2004;30:584-90.
10. Kimbrough RL, Trempe CS, Brockhurst RJ, Simmons RJ. Angle-closure glaucoma in nanophthalmos. Am J Ophthalmol 1979;88:572-9.
11. Rao A, Padhi T, Jena A, Mandal S, Das T. Atypical features of nanophthalmic macula—a spectral domain Oct study. BMC Ophthalmol 2012;12:12.
12. Walsh M, Goldberg M. Abnormal avascular zone in nanophthalmos. Am J Ophthalmol 2007;143:1067-8.
13. Khorram D, Choi M, Roos B, et al. Novel *Tmem98* mutations in pedigrees with autosomal dominant nanophthalmos. Mol Vis 2015;21:1017-23.
14. Awadalla MS, Burdon KP, Souzeau E, et al. Mutation in *Tmem98* in a large white kindred with autosomal dominant nanophthalmos linked to 17p12-q12. JAMA Ophthalmol 2014;132:970.
15. Day A, MacLaren R, Bunce C, Stevens J, Foster PJ. Outcomes of phacoemulsification and intraocular lens implantation in microphthalmos and nanophthalmos. J Cataract Refract Surg 2013;39:87-96.
16. Sharon S, Grigg J, Higgins R. Nanophthalmos: ultrasound biomicroscopy and Pentacam assessment of angle structures before and after cataract surgery. J Cataract Refract Surg 2006;32:1052-5.

digital journal of ophthalmology, vol. 23