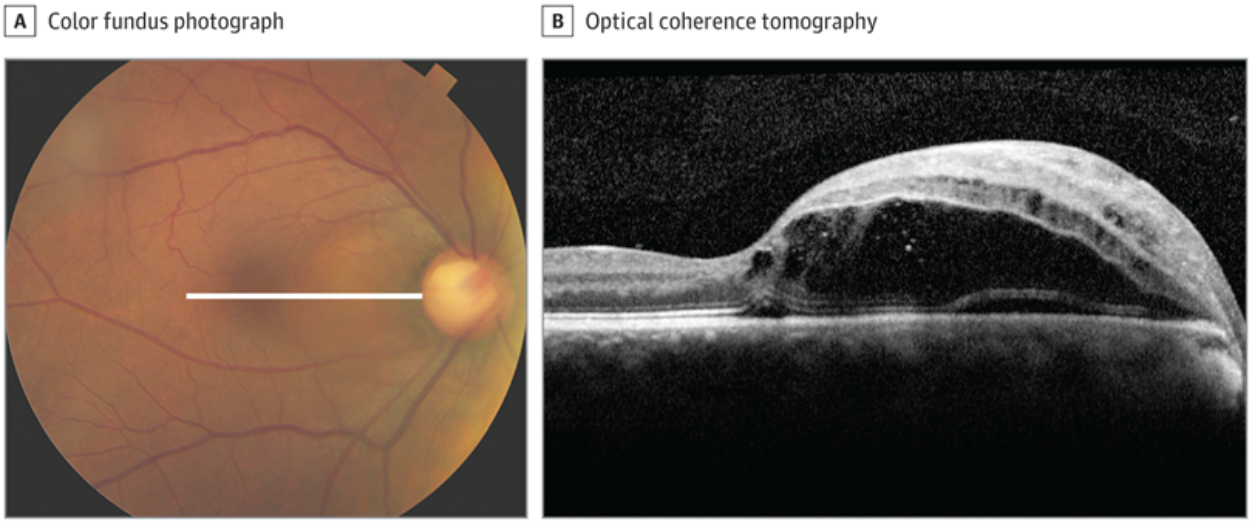
Macular Fluid in a Patient With a Reported History of Normal-Tension Glaucoma

Case

A patient with a history of normal-tension glaucoma presented with a blurred spot in the central vision of the right eye. Examination showed best-corrected visual acuity of 20/50 OD, 20/30 OS, and intraocular pressure of 13 mm Hg OU. In the right eye, dilated fundus examination and optical coherence tomography (OCT) of the macula revealed subretinal fluid and cystoid spaces within the outer nuclear layer and inner nuclear layer in the nasal macula (Figure 1). Results of the left eye fundus examination and macular OCT were normal. In both eyes, the optic cup appeared very deep with a large cup-disc ratio.

Figure 1.



Color fundus photograph (A) and optical coherence tomography of the macula (B) illustrating intraretinal and subretinal fluid at the nasal macula. The white line indicates the position of the optical

coherence tomography scan.

Five years prior to presentation, the patient started treatment with latanoprost nightly in both eyes for an enlarged cup-disc ratio presumed due to normal-tension glaucoma. At that time, intraocular pressure was 12 mm Hg OD and 14 mm Hg OS and central corneal thickness was normal. Visual field and OCT of the retinal nerve fiber layer were without reproducible defects over 5 years of follow-up. The patient denied a family history of eye disease.

What Would You Do Next?

1. Intravitreal anti–vascular endothelial growth factor
2. Fluorescein angiography
3. OCT of the optic nerve head
4. OCT-angiography of the macula

Discussion

Diagnosis

What to Do Next

C. OCT of the optic nerve head

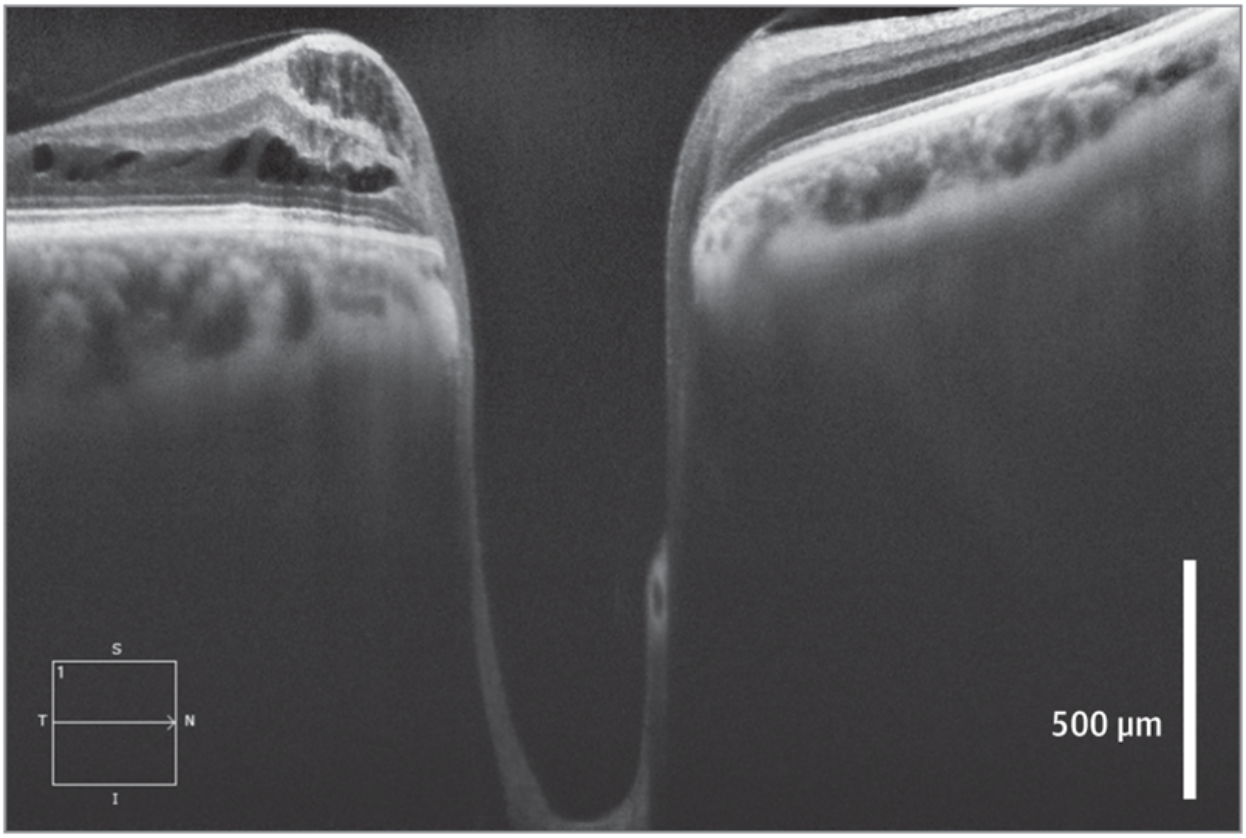
Discussion

The differential diagnosis of macular fluid includes choroidal neovascularization, central serous chorioretinopathy, myopic foveoschisis, optic pit maculopathy, and cystoid macular edema. In this case, the subretinal and intraretinal fluid in multiple retinal layers contiguous with the optic nerve point to the optic nerve,

rather than vascular leakage, as the source of the fluid. Therefore, fluorescein angiography (choice B) and OCT-angiography of the macula (choice D), tests that detect vascular sources of fluid, are not indicated, and anti–vascular endothelial growth factor (choice A), a treatment for vascular leakage, is not appropriate.

Although optic pit maculopathy without inner retinal schisis cavity has been reported, the pattern and distribution of fluid is most consistent with optic pit maculopathy.[1,2] However, clinical examination and OCT of the optic nerve revealed a very deep cup (1388 µm from the Bruch membrane opening) without a focal depression seen in optic pit (Figure 2).

Figure 2.



Optical coherence tomography of the optic nerve head illustrating a very deep cup without a focal optic pit.

Glaucoma can lead to increased cup depth, although this patient's cup depth is far outside the range seen in glaucoma.[3] Excessively deep optic cups have been reported in optic pit but are common in cavitary optic disc anomaly (CODA).[4] Rarely, macular schisis occurs secondary to glaucoma,[5] but the normal intraocular pressure, normal retinal nerve fiber layer on OCT, very deep optic cup, and normal visual fields (data not shown) make CODA, rather than glaucoma, the likely diagnosis in this patient.

Congenital excavated optic nerve abnormalities (optic nerve coloboma, optic pit, morning glory anomaly, and CODA) have a risk of vision loss around age 20 to 40 years due to macular fluid.[4] These abnormalities are usually unilateral and sporadic, but are rarely autosomal dominant because of a triplication upstream of the *MMP19* gene,[6,7] which encodes an extracellular matrix protease in the optic nerve.[6] In CODA, optic nerve abnormalities are highly penetrant with variable phenotype (optic pit, morning glory, atypical coloboma, glaucomatous cupping, and excessively deep optic cups).[4] The central retinal artery is often not visible and retinal vessels appear to emanate directly from the cup edge. The optic cup may gradually enlarge, and excavation may develop from a previously normal optic nerve.[7] These features of CODA mimic glaucoma, but should not be confused to avoid unnecessary treatments.

In excavated optic nerve abnormalities, histologic studies and swept-source OCT imaging reveal that dysplastic retina herniates through the lamina cribrosa or juxtapapillary sclera and protrudes into the subarachnoid space.[8] Fluid is thought to follow a pressure gradient through the defect into the retinal stroma and subretinal space. The source of the fluid may be liquified vitreous and/or cerebrospinal fluid.[8,9]

A period of observation is recommended since the macular fluid may spontaneously resolve, but persistent macular fluid may lead to permanently decreased vision to the 20/200 level or worse. The optimal treatment is debated, but vitrectomy alone may be sufficient in some.[10]

Patient Outcome

A visit 2 months later showed improved visual acuity to 20/20 and spontaneous regression of intraretinal and subretinal fluid. Latanoprost was stopped. Over the next 4 years, the patient remained stable without recurrent fluid or visual field changes.

Back to top

Article Information

Corresponding Author: Eileen S. Hwang, MD, PhD, Department of Ophthalmology and Visual Sciences, University of Utah, 65 Mario Capecchi Dr, Salt Lake City, UT 84132 (eileenhwang@yahoo.com).

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