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

Digital Journal of Ophthalmology

Grand Rounds

A 31-year-old man with vitritis, chorioretinitis, and hydrocephalus

Andrew J. Schneier, MD

Author affiliations: Author affiliations: Massachusetts Eye and Ear Infirmary, Boston, Massachusetts

History

A 31-year-old man presented to the emergency department of the Massachusetts Eye and Ear Infirmary with a chief complaint of blurry vision and redness in his right eye of 1 week's duration. He was a native of Brazil, and the history and examination were performed with the aid of a Portuguese interpreter.

The patient was in his usual state of excellent health preceding the episode. He reported mild pressure in the affected eye along with a constant, bilateral, temporal headache that may have predated the blurred vision. The headache was readily relieved by acetaminophen. He denied flashes, photosensitivity, or other visual aberrations. He maintained that his vision was "weaker" on the right at baseline. On further questioning, he felt that his symptom was best characterized as "many floaters" in the right eye. He believed he may have been exposed to a toxic chemical at his workplace 2 months prior to this presentation.

The patient's past medical history was unremarkable. He had no known drug allergies, and he denied a family history of medical or surgical conditions. His social history was significant for the use of occasional alcohol. He denied high-risk sexual activities, intravenous drug use, and blood transfusions.

Examination

On ophthalmic examination, visual acuity was 20/200 in the right eye, with pinhole improvement to 20/100, and 20/40 in the left eye, with pinhole improvement to 20/20. Ishihara color vision testing showed correct identification of 3 of 8 plates in the right eye and 8 of 8 plates in the left eye. There was a relative afferent pupillary defect present in the right eye. Slit-lamp examination revealed trace conjunctival injection in the right eye, along with small keratic precipitates on the inferior corneal endothelium and 3+ cell in the anterior chamber. The anterior segment of the left eye was normal. Normal and symmetric intraocular pressures were obtained.

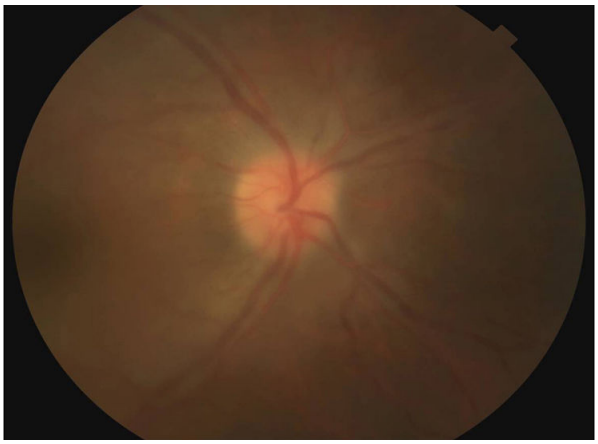

Figure 1. Right fundus, optic nerve head margins appear blurred. Details obscured due to vitritis.

Dilated stereoscopic fundus examination revealed 2+ vitreous cell in the right eye and bilateral optic nerve head edema (Figures 1–2). Indirect ophthalmoscopy of the right eye demonstrated a large, pigmented, inferotemporal chorioretinal scar with a superjacent area of presumed retinitis (Figure 3). The macula and peripheral retina of the left eye were normal.

Treatment

The patient's headache and disc edema warranted a referral to Massachusetts General Hospital for neuroimaging and evaluation. Our recommendations included

Published May 14, 2012.

doi:10.5693/djo.03.2012.03.001

Correspondence: Andrew J. Schneier, MD, 243 Charles Street, Boston, MA 02114 (email: andrew_schneier@meei.harvard.edu)

Digital Journal of Ophthalmology

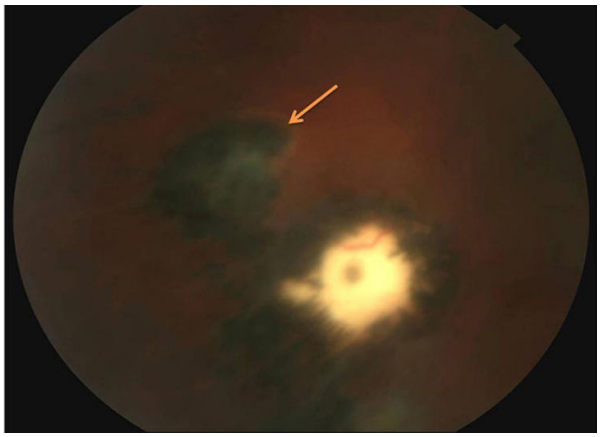

Figure 2. Right fundus, centered on chorioretinal lesion infero-temporal to macula. The lesion is notable for encircling pigment and the suggestion of retinitis at superior border (denoted by arrow).

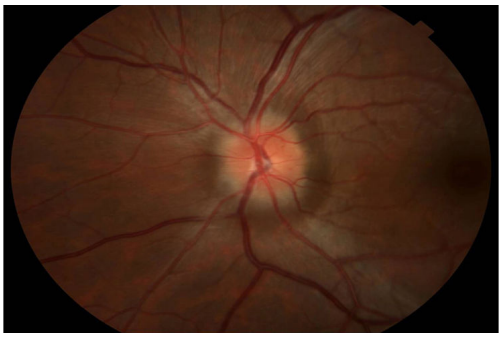

Figure 3. Left fundus showing optic disc edema.

serologic testing for HIV and toxoplasmosis. Computed tomography was performed without contrast and disclosed severe hydrocephalus likely related to cerebral aqueductal stenosis (Figure 4). An MRI scan with and without gadolinium contrast was ordered to further delineate the pathology; this revealed hydrocephalus with dilatation of the lateral and third ventricles with a normal-appearing fourth ventricle (Figures 5–6). The sagittal FIESTA protocol showed webs within the cerebral aqueduct and a lack of CSF flow through to the fourth ventricle.

After neurosurgical consultation, the patient underwent a semi-emergent right endoscopic third ventriculocisternostomy, without complication, to prevent further visual loss and to alleviate the patient's headaches. The patient was discharged the following day.

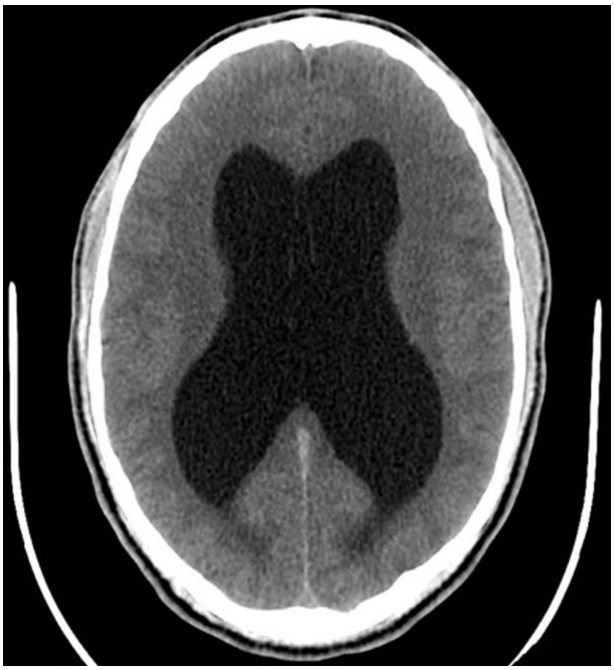

Figure 4. CT brain without contrast, axial view, revealing symmetric enlargement of the lateral ventricles.

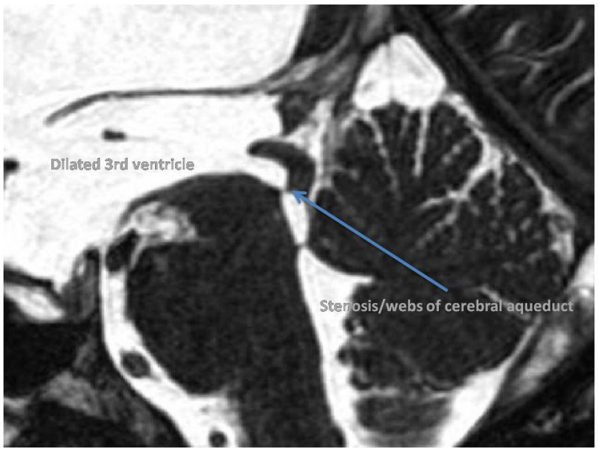

Figure 5. MRI, T2 FIESTA protocol, sagittal view, delineating anatomy of ventricles and CSF flow.

Differential Diagnosis

Our patient presented with headaches and a unilateral subjective visual disturbance. Examination revealed vitritis and characteristic chorioretinal lesions in the right eye as well as bilateral disc edema. Our initial differential diagnoses included cerebral toxoplasmosis/encephalitis, as well as additional etiologies of papilledema, including mass lesion, space-occupying hemorrhage,

Digital Journal of Ophthalmology

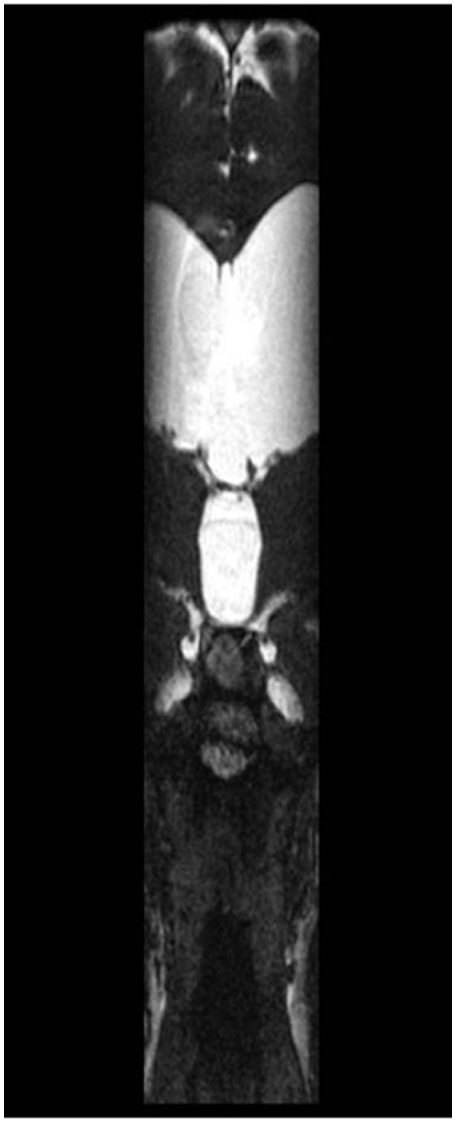

Figure 6. MRI FIESTA, coronal view, revealing engorgement of the lateral and 3rd ventricles.

disturbance of cerebrospinal fluid flow (obstructive, absorptive, overproduction), obstruction of venous outflow, and idiopathic intracranial hypertension.

Diagnosis and Discussion

We felt that the clinical picture was consistent with reactivation toxoplasmosis in the right eye. However, the appearance of disc edema in the left eye merited emergent neuroimaging and further workup. Marked hydrocephalus was discovered and intervention was successful.

The coexistence of obstructive hydrocephalus with attendant papilledema and characteristic chorioretinal lesions in our patient from Brazil raises the question of a hitherto undiagnosed congenital or acquired cerebral toxoplasmosis infection.

Classically, congenital toxoplasmosis is characterized by the triad of hydrocephalus, chorioretinal lesions, and cerebral calcifications.[1] Chorioretinal involvement is present up to 85% of infected subjects before adulthood.[2] In two large, retrospective, population-based studies, 28% to 50% of children diagnosed with congenital toxoplasmosis were found to have hydrocephalus.[3]

Cerebral toxoplasmosis is considered to be the most common opportunistic infection affecting the central nervous system in patients with HIV/AIDS.[4] However, hydrocephalus in the setting of cerebral toxoplasmosis without an associated mass lesion is an extremely rare; it has been described in fewer than 5 case reports.[5]

Though we cannot directly determine the chronicity of our patient's conditions, it is possible that his hydrocephalus was longstanding based on the relatively normal intracranial pressure (16 cm fluid height) obtained upon fenestration of the 3rd ventricle by the neurosurgical team—a so-called "compensated" hydrocephalus. His discs, however, lacked the hallmarks of swelling secondary to chronic ICP elevation, including gliosis of the nerve fiber layer and optociliary shunt vessels. The symptoms of visual decline and headaches were also newly realized by the patient, suggesting at least an acute-on-chronic course.

Our patient initially had a good outcome with a shunting procedure, including resolution of his headache and subjective improvement in his vision. Unfortunately, serologic testing for toxoplasmosis was not obtained prior to his discharge, and he did not return to our service for follow-up. His particular presentation was suggestive of a unifying diagnosis: the cerebral and retinal stigmata of congenital toxoplasmosis. However, it is impossible to confirm this diagnosis without serologic evidence and further surveillance of the course of the disease.

References

1. Jones J, Lopez A, Wilson M. Congenital toxoplasmosis. Am Fam Physician 2003;67:2131-8.15
2. Vasconcelos-Santos DV, Machado Azevedo DO, Campos WR, et al. Congenital Toxoplasmosis Brazilian Group. Congenital toxoplasmosis in southeastern Brazil: results of early ophthalmologic examination of a large cohort of neonates. Ophthalmology 2009;116:2199-205.e1.
3. McAuley J, Boyer KM, Patel D, et al. Early and longitudinal evalu-

ations of treated infants and children and untreated historical patients with congenital toxoplasmosis: the Chicago Collaborative Treatment Trial. Clin Infect Dis 1994;18:38-72.

4. Cota GF, Assad EC, Christo PP, Giannetti AV, Santos Filho JA, Xavier MA. Ventriculitis: a rare case of primary cerebral toxoplasmosis in AIDS patient and literature review. Braz J Infect Dis 2008;12:101-4.
5. Nolla-Salas J, Ricart C, D'Olhaberriague L, et al. Hydrocephalus: an unusual CT presentation of cerebral toxoplasmosis in a patient with acquired immunodeficiency syndrome. Eur Neurol 1987;27:130-2.

Digital Journal of Ophthalmology