Papilledema secondary to ocular hypotony provides helpful insights on discriminating true from pseudo-optic nerve head swelling

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Abstract
Optic nerve head swelling can be a significant challenge in clinical practice when clinicians are required to differentiate true swelling (papilledema) from pseudo-papilledema (benign elevation of the optic nerve head). This case presentation describes a patient who experienced an ocular laceration that led to a vitreal prolapse and ocular hypotony. The ocular hypotony led to papilledema and retinal edema extending throughout the macular region of the right eye. The patient has been followed over a 2 year period with gradual improvement in visual acuity, visual field and various aspects of optical coherence tomography (OCT) measurements. The long-term follow up provides useful insights on the time-course of potential recovery, including the time course of reduction in retinal and optic nerve edema and identifies many useful and some pitfalls to be avoided when interpreting OCT imaging. Finally, an overview will be provided on discriminating true from pseudo-optic nerve head swelling using selected cases from the Centre for Eye Health.

I – Case History
A 28 year old female patient presented for an eye examination within two weeks after hospital discharge to repair a scleral laceration secondary to a knife injury to the right eye. She was complaining of visual disturbance OD and seeking an explanation for the cause of her visual disturbance and the timeline for recovery. She has been followed over 2 years.

II – Pertinent findings
Entering aided acuities were 20/25 OD and 20/20+ OS.
Ocular motility testing showed a full range of eye movements OU.
Evidence of the laceration included scleral sutures below the nasal conjunctiva extending over approximately 6-8mm length. A 10 day period of high systemic steroids did not improve vision in the right eye. No further treatment was recommended by the ophthalmology department.

Funduscopy and retinal photography revealed ONH and macular swelling OD. OCT scans revealed an edematous right ONH which has visibly reduced over the 2 year period (see below). Visual field assessment confirmed the improvement evident in the fundus photographs and reduced OCT thickness measurements. Intra-ocular pressure was within 2-3 mm Hg of the left eye within 6 weeks of surgery and has remained there over the 2 year follow up.
III – Differential diagnosis

"Unilateral optic nerve head swelling" can be caused by inflammatory, ischemic, infiltrative, direct optic nerve compression, trauma or through toxicity. In this case, ocular hypotony is the most likely cause.

IV – Diagnosis and discussion

Severe hypotony can lead to papilledema, choroidal folds resulting in choroid/retinal relationship compromise. The resultant retinal edema leads to visual disturbance experienced by this patient. One of the unique features in this case is the significant swelling of the retinal nerve fiber layer, which continues to improve over the 2 year period. Careful assessment of the OCT information comparing the two eyes provides additional information relating to the increased thickness which is masked if only the normative data are analysed. Although the optic nerve swelling in the acute phase is thought to be due to reduced axoplasmic flow, the optic nerve continues to remain swollen 2 years after the initial injury. It is also possible to obtain corneal edema, choroidal detachment and cataract secondary to ocular hypotony.

Although the more common causes of hypotony are secondary to glaucoma surgery (trabeculectomy–leaking bleb), secondary to inflammatory disease (~8% uveitis) and secondary to intraocular infection, this case displays a wide spectrum of ocular changes associated with ocular hypotony.

V – Treatment, management

Management of ocular hypotony rests in identifying the underlying cause. Surgery was indicated and undertaken on the day of the laceration injury. Subsequent to that, a course of high systemic steroids was undertaken with no beneficial effect. In other causes of ocular hypotony, particularly secondary to bleb surgery or inflammation, managing the underlying cause is a major concern. In this case, providing timely information and likely prognosis, alleviated patient anxiety relating to return of her vision.

VI – Conclusion

Several cases of monocular and binocular ‘swollen’ optic nerves will be contrasted with this patient to highlight some similarities and significant differences to help clinicians differentiate the different types of disc swelling.

Abstract: 44 year-old Hispanic male presents with bilateral blurry vision and associated light flashes. Symptoms are exacerbated by activity and loud sounds. Exam findings reveal exuberant disc edema and hemorrhages with visual field defect on testing.

I. Case History

Patient demographics

- 44 year old male of Hispanic descent presents to the eye clinic on consult from a nearby hospital. He was recently diagnosed with cryptococcal meningitis secondary to undiagnosed Human Immunodeficiency Virus (HIV) and is currently undergoing treatment. The diagnosis is confounded by visual symptoms and discomfort.

Chief complaint

- Ocular complaints - Patient reports the following symptoms which began about 2 weeks prior to presentation in our clinic:
  - Blurred vision: Present for two weeks and worsening, Right eye > Left eye. Exacerbated by head movement, activity, and loud sounds.
- Systemic associations:
  - Hallucinations: patient experienced hallucinations about three weeks prior but are no longer present.
  - Malaise: patient notes an overall feeling of malaise with sweating and headaches. Notes the headaches are mild and diffuse, worse upon movement. He denies nausea or diplopia.

Ocular, medical history

- Ocular History:
  - Denies the following: history of patching, redness to either eye, ocular trauma or ocular surgery
  - No previous ocular history
  - Last eye exam was 3 months prior and unremarkable
- Medical History: Review of patient’s medical history was significant for the following:
  - Recently diagnosed Human Immunodeficiency Virus (HIV) with recent CD4 count of 44
  - Cryptococcal meningitis

Medications

- Excedrin migraine
- Acetaminophen
- Ondansetron
- Diazepam
- Meclizine
- Flucytosine (administered intravenously at the hospital)

II. Pertinent findings

Clinical

- VA: OD 20/100, OS 20/200
- EOMS: full, unrestricted, denies worsening of pain on eye movement
- Confrontation fields: full to finger counting OD, nasal hemianopsia OS
- Manifest Refraction:
  - OD: +2.00 -3.00 x 010, VA 20/60
  - OS: +2.00 -2.75 x 160, VA 20/70
Slit lamp examination: rare cell in the anterior chamber; otherwise, unremarkable

Tonometry: OD 6, OS 6

Dilated fundus examination: (see photos below for OU)

<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
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<tbody>
<tr>
<td>Vitreous</td>
<td>Trace anterior vitreous cells</td>
<td>Trace anterior vitreous cells</td>
</tr>
<tr>
<td>ONH Rim and Margins</td>
<td>2-3+ disc edema 360 with flame-shaped hemes greater inferiorly</td>
<td>2-3+ disc edema 360 with few flame-shaped hemes</td>
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<tr>
<td>ONH</td>
<td>0.15/0.15 H/V</td>
<td>0.10/0.10 H/V</td>
</tr>
<tr>
<td>Macula</td>
<td>Slight epiretinal membrane</td>
<td>Questionable cystic changes in the fovea</td>
</tr>
<tr>
<td>Vessels</td>
<td>A/V 2/3 with mild tortuosity, no vasculitis</td>
<td>A/V 2/3 with mild tortuosity, no vasculitis</td>
</tr>
<tr>
<td>Posterior Pole</td>
<td>See photo below</td>
<td>See photo below</td>
</tr>
<tr>
<td>Periphery</td>
<td>No inflammatory lesions or hemorrhages</td>
<td>No inflammatory lesions or hemorrhages</td>
</tr>
</tbody>
</table>

HVF 30-2:

Physical

Past hallucinations at onset of symptoms, headaches, general malaise, increased sweating, fever, visual symptoms worse upon head movement

Laboratory studies

The patient had already undergone an intensive workup in the emergency department where it was concluded that he had cryptococcal meningitis and was severely immunocompromised with a positive HIV test on admittance. His CD4 count on admittance to the hospital was 44 cells/mm³
III. Differential diagnosis
The patient’s symptoms and clinical manifestations are consistent with bilateral optic disc edema. Had the patient first presented to the eye clinic instead of the emergency department, a full work-up would have been initiated to rule out the differentials below that can cause bilateral disc edema. A workup may have involved MRI with contrast of the head and brain and lumbar puncture with CSF analysis and opening pressure if the MRI was normal.

- **Primary or metastatic intracranial tumors**
- **Infectious or inflammatory differentials**
  - Brain abscess
  - Meningitis (e.g., syphilis, tuberculosis, Lyme disease, bacterial, fungal, inflammatory or neoplastic)
  - Encephalitis
- **Intracranial Hemorrhaging**
  - Subarachnoid hemorrhage
  - Subdural or epidural hematoma
- **Idiopathic intracranial hypertension** (diagnosis of exclusion)

IV. Diagnosis and discussion

Given the patient was recently diagnosed with cryptococcal meningitis on his admittance to the hospital just days earlier, a workup was not conducted. The bilateral disc edema was presumed to be papilledema due to raised intracranial pressure in the setting of meningitis.

This case presentation was unusual in that with current antiretroviral therapy (ART) and a decrease in the number of severely immunocompromised individuals, cryptococcal infection is relatively rare. *Cryptococcus neoformans* is the most-common life-threatening fungal pathogen that infects patients with AIDS. It typically manifests as meningitis with about 40% demonstrating abnormal ocular findings. The diagnosis of cryptococcal meningitis is based upon clinical symptoms, radiological findings, and a lumbar puncture including analysis of the cerebrospinal fluid (CSF).

V. Treatment, management

Visual loss in cryptococcal meningitis may occur from elevated intracranial pressure or direct invasion of the optic nerves or visual pathway by the organism. Due to the severity and possible (life-threatening) consequences of a cryptococcal infection, medication treatment in the form of anti-fungals should be aggressively pursued. Optic nerve sheath fenestration has also been shown to offer benefit in some individuals by reducing the effects of chronic disc edema.

The patient in our case underwent medical treatment immediately once a diagnosis was reached. He was followed by our clinic through six weeks of extensive anti-fungal treatment but unfortunately was lost to follow-up thereafter. On his last visit, his disc edema was almost resolved and his vision was correctable to 20/40 in both eyes. The patient’s visual field also improved on testing. Due to improvement in disc edema and visual functioning, optic nerve sheath fenestration was not pursued; however, would have been considered had findings continued to show progression.

VI. Conclusion

Bilateral disc edema could result from a multitude of different conditions. Immediate work-up and appropriate treatment should be coordinated in a timely manner in order to decrease overall fatality and reduce damage associated with chronic optic disc edema.
Bibliography


Water Gradient Silicone Hydrogel Lens as an Alternative Treatment Option for Lid Wiper Epitheliopathy
Anne Tasaki, O.D.

Disclosure Statement:
Nothing to disclose

26-year-old Asian male

• Chief complaint:
  – “Heavy” eyelids with contact lens wear
  – Removes after 5 minutes of wear time
• Current contact lenses
  – Air Optix Aqua® 8.6/14.2/-4.75 DS OU
• Review of systems: eczema

Examination Findings

• Entering acuity: 20/20 OD, OS
• Pupils: equal, round, reactive to light, no APD
• EOMs: unrestricted, no ptosis in all gazes
• Anterior segment:
  – Normal superior tarsal conjunctiva with eversion
  – Absence of corneal staining with NaFl
Differential Diagnosis

• Dry eye
• Contact-lens induced papillary conjunctivitis
• Superior epithelial arcuate lesion
• Lid wiper epitheliopathy

Lid Wiper Epitheliopathy

• Dry eye symptoms without dry eye signs
• Typically found in contact lens wearers – 80% of symptomatic wearers (Korb 2002)
• Subset of dry eye?

Pathophysiology

• Inadequate lubrication between the lid wiper and corneal/bulbar conjunctival surfaces
• Mechanical damage to and inflammation of the lid wiper

Grading System

<table>
<thead>
<tr>
<th>Grade</th>
<th>Horizontal Length of Staining</th>
<th>Sagittal Height of Staining</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>&lt;2 mm</td>
<td>&lt;25%</td>
</tr>
<tr>
<td>1</td>
<td>2-4 mm</td>
<td>25% to &lt;50%</td>
</tr>
<tr>
<td>2</td>
<td>5-9 mm</td>
<td>50% to &lt;75%</td>
</tr>
<tr>
<td>3</td>
<td>&gt;10 mm</td>
<td>&gt;75%</td>
</tr>
</tbody>
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Current Treatment Options

• Artificial tears
• Topical steroids
• Changing contact lens type
• Cessation of contact lens wear

Case Example: Treatment Plan

• Discontinue contact lens wear
• Initiate use of non-preserved artificial tears 3 times daily OU
• 1 week follow-up for contact lens refit
Dailies Total 1®
- Water gradient SiHy lens by Alcon
- “Lubricious” surface
- Significantly impacts pre-lens tear film less than predecessor lens

Contact Lens Refit
- Dailies Total 1® 8.5/14.1/-4.75 DS OU
- Visual acuity: 20/20 OD, OS
- Plano over-refraction OU
- Appropriate lens fit
- 1 week follow-up for contact lens check

Contact Lens Follow-up
- Increased wear time of up to 6 hours
- 1 month follow-up

1 month Follow-up
- Increased wear time of 8-10 hours per day
- Mild dryness after 8-10 hours of wear

Right Eye

Left Eye
Summary

- Lid wiper epitheliopathy is a newly recognized condition
- Current treatment options are limited
- Consider Dailies Total 1® for symptomatic contact lens wearers

References

5. 2008;85:E924-­‐E929.