1. New England Medical Center Grand Rounds - Uveitis-Glaucoma-Hyphema Syndrome

New England Medical Center Grand Rounds

Uveitis-Glaucoma-Hyphema (UGH) Syndrome

My Hanh T. Nguyen, M.D.
May 19, 2005

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2. Case Presentation

Case Presentation

- 80 year old female
- CC: “recurrent bleeding in right eye”
- HPI:
  - Episodes of blurry vision OD
  - Diagnosed with recurrent hyphemas OD associated with high IOP
    - June 2004  IOP 50
    - October 2004  IOP ?? ‘high’
    - December 2004  IOP 50
    - February 2005  IOP 38
    - March 2005  IOP 30
  - Treated with Diamox, Alphagan, Travatan without sequella
- POHx:
  - Pseudophakic OU 22 yrs ago
  - No trauma or infections OU

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3. Case Presentation

Case Presentation

- **PMHx:**
  - HTN
  - Hypercholesterolemia

- **Medications:**
  - Dorzolamide / timolol maleate BID OU
  - Metoprol Succinate
  - Valsartan
  - Atorvastatin

- **Allergies:** NKDA

- **Fam Hx:** unremarkable

- **Soc Hx:**
  - Retired
  - Denies tobacco
  - 1 glass wine QD

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4. Office Examination

Office Examination

- **BCVA:** 20/20 OU
- **T:** 20 OD, 20 OS
- **Pupils:** 5mm → 3mm OU, no RAPD OU
- **EOM:** full OU
- **VF (conf):** full OU

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5. Office Examination

Office Examination

- **SLE**
  - LLL: wnl OU
  - C/S: W & Q OU
  - K: clear OU
  - A/C: 2+ cells OD (most RBC’s); D & Q OS
  - I:
    - Patent I’s OU
    - No rubeosis OU
    - No transillumination defects OU
  - L:
    - Sulcus PCIOL OD (vertical), ACIOL OS (vertical)
    - Posterior capsule intact OD, open OS
    - Blood staining between optic and capsule OD
    - No pseudophakodystrophy OU
    - No pseudoexfoliation (PXF) OU
  - V: 2+ cells OD (most RBC’s); no cells OS

- **DFE:** M/V/P wnl OU

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6. Differential Diagnosis of Hyphema

Differential Diagnosis of Hyphema?

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7. **Differential Diagnosis**

### Differential Diagnosis

- **Trauma**
- **Vascular abnormalities**
  - Rubeosis iridis
  - Ocular ischemic syndrome
  - Diabetes mellitus
  - Retinal artery occlusion
  - Retinal vein occlusion
  - Swan syndrome (NV of wound)
  - Juvenile xanthogranuloma
  - Iris varices
  - Vascular tufts
  - Hereditary hemorrhagic telangiectasia
- **Inflammation (Iritis)**
  - Fuchs heterochromic iridocyclitis
  - Herpes simplex
  - Herpes zoster
- **Iatrogenic Causes**
  - Intraocular surgery
  - Laser trabeculoplasty
  - Indotomoty
  - Uveitis-glaucoma-hyphema Syndrome (UGH)
- **Neoplasm**
  - Retinoblastoma
  - Melanoma
  - Iris hemangiomas
- **Systemic Disorders**
  - Sickle cell trait or disease
  - Coagulation disorders
  - Anticoagulation medications

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8. **What would you do next?**

### What would you do next?

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9. Gonioscopy

Gonioscopy

- CBB 360° OU
- No PAS OU
- No rubeosis OU
- 1+ pigment inferiorly OD

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10. Differential Diagnosis

Differential Diagnosis

- Trauma
- Vascular abnormalities
  - Rubeosis iridis
    - Ocular ischemic syndrome
    - Diabetes mellitus
    - Retinal artery occlusion
    - Retinal vein occlusion
    - Swan syndrome (NV of wound)
    - Juvenile xanthogranuloma
    - Iris varices
    - Vascular tufts
    - Hereditary hemorrhagic telangiectasia
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  - Herpes simplex
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  - Intraocular surgery
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  - Uveitis-glaucoma-hyphema Syndrome (UGH)
- Neoplasm
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  - Sickle cell trait or disease
  - Coagulation disorders
  - Anticoagulation medications

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11. Ultrasound Biomicroscopy (UBM)

Images courtesy of New England Eye Center 2006
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13. **UGH Syndrome a.k.a. "Ellingson Syndrome"**

- Ellingson, 1978
- Originally described in association with first-generation ACIOL and contact between IOL haptic or optic with uveal tissue
  - Excessive lens movement
    - Small size
    - Decentration or dislocation
  - Poorly manufactured edges
  - Iris-clipped IOL
  - Rigid, closed loop haptics
- Definition expansion
  - Unstable sulcus fixation
  - PCIOL decentration (zonular weakness i.e. PXF, trauma)

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14. **UGH Syndrome - Etiology of Uveitis**

- **UGH Syndrome**
- **Etiology of Uveitis**

- Activation of innate immunity

- Theories
  - Cytokine and eicosanoid synthesis triggered by mechanical exoriation of the angle or iris by the haptics or optic
    - Iris chafing
    - Warpage of foot plates
    - Erosion of angle structures / ciliary sulcus
  - Plasma-derived enzymes (especially complement or fibrin) activated by the surface of the IOLs (especially PMMA)
  - Adherence of bacteria and leukocytes to the IOL surface
  - Toxicity caused by contaminants on the IOL surface during manufacturing or implantation

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15. UGH Syndrome

UGH Syndrome

- Clinical spectrum
  - Iris pigment epithelial defects
  - Pigment dispersion
  - Microhyphema
  - Macrohyphema with elevated IOP
- Timing postoperatively
  - Usually weeks to months
  - May be several years
    - Literature: 1-8 years
    - Our patient: 22 years

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16. UGH Syndrome - Presenting Symptoms

UGH Syndrome
Presenting Symptoms

- Intermittant blurring
- Intermittant “white out”
- Reddening of vision (erythropsia)
- Eye pain
- Photophobia
- Red eyes

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17. UGH Syndrome - Complications

UGH Syndrome
Complications

- Pseudophakic bullous keratopathy (PBK)
- Corneal staining
- Chronic inflammation
- Vitreous hemorrhage
- Glaucomatous nerve damage
- Cystoid macular edema (CME)

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18. UGH Variations

UGH Variations

- **UGH Plus**
  - Uveitis
  - Glaucoma
  - Hyphema
  - PLUS vitreous hemorrhage

- **IPUGH**: incomplete posterior UGH
  - Bleeding into the posterior chamber
  - +/- Glaucoma
  - No uveitis

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19. **UGH Syndrome - Treatment Options**

UGH Syndrome

**Treatment Options**

- Observe and treat each episode
  - Topical steroids
  - IOP lowering drops
- Miotics and mydriatics
- IOL rotation
- Laser therapy
- Explantation of IOL +/- replacement

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20. **IOL Explantation - Indications**

**IOL Explantation Indications**

- ACIOL & iris-fixated lenses
  - PKB with corneal decompensation
  - UGH syndrome with CME
- Older PCiol & PMMA PCiol
  - Decentration / dislocation
  - Corneal edema and inflammation
- Foldable 3-pc monofocal silicone PCIOL
  - 40% incorrect lens power
  - 32% decentration / dislocation
  - 9% damaged IOL during insertion

- Foldable 3-pc acrylic PCIOL
  - 39% incorrect lens power
  - 24% glare / optical aberrations
  - 15% decentration / dislocation
- Foldable plate-haptic silicone PCIOL
  - ≥ 50% decentration / dislocation
  - 22% incorrect lens power
  - 13% damaged IOL during insertion
- Foldable 3-pc multifocal silicone PCIOL
  - 89% glare / optical aberrations
  - 11% others

Intermountain Ocular Research Center at the University of Utah
ASERS & ESCRS, 2000 (259 surveys)

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21. IOL Explantation - Retrospective Reviews

IOL Explantation
Retrospective Reviews

- University of Florida, Gainesville, Florida
- 97 patients
- 1983-1987
- Type of IOL
  - ACIOL: 54%
  - Iris-fixated lenses 34%
- Indications for surgery
  - PBK 69%
  - UGH 9%
  - IOL instability 7%
- Sinskey Ophthalmic Center, Santa Monica, California
- 79 patients
- 3 month to 12 year follow up
- Type of IOL
  - ACIOL 35%
  - PCIOL 61%
- Indications for surgery
  - Decentered / displaced 42%
  - Corneal decompensation 28%
  - Incorrect IOL power 13%
  - UGH 10%

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22. Case Summary

Case Summary

- 80 year old female 22 years s/p CE/IOL OU

- Presentation
  - Recurrent hyphemas OD
  - Associated high IOP OD

- Findings
  - Sulcus PCIOL OD
  - UBM with inferior haptic pressing on ciliary body / iris junction

- Diagnosis: UGH Syndrome

- Treatment options
  - Observation
  - IOL rotation, placing haptics horizontally
  - Endolaser

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 Sources


Crouch ER and Crouch ER. Emedicine. Management of Traumatic Hyphema: Therapeutic Options.


Mamalis N and Spencer TS. Hyperguide. Complications of foldable IOLs.


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