The Case of the Peruvian Pinguecula

Asymptomatic arteriovenous malformation caught on routine automated visual field screening. Patient does not follow medical advice and delays neuroimaging. Eventual neuroimaging shows AV malformation with large nidus that is removed in two surgeries.

I. Case History
   a. 32 year old Hispanic male presents for routine care

II. Pertinent findings
   a. Screening visual field shows OD: infero-temporal non-repeatable defect, OS: infero-nasal defect
   b. Threshold visual field confirms OD: infero-temporal defect, OS: larger infero-nasal defect. Both respect the vertical midline. OS defect extends past the horizontal midline
   c. Visual field interpretation is non-congruous (right more than left) homonymous right hemianopsia which can be caused by a lesion in the optic tract.
   d. All other exam findings within normal limits except that the patient has bilateral pinguecula
   e. A neuro consult is scheduled which the patient fails and is lost to follow-up for 18 months. Pt. returns on an urgent basis for evaluation of an inflamed pinguecula. He states that he was non-compliant with the neuro consult because his father (MD in Peru) told him he cannot have any brain lesions in absence of a headache. Father further stated that it had been 2 months since his VF defect and he has not died or had other symptom therefore there must be nothing wrong. VF is discussed with pt. again and pt. is encouraged to get neurological consult due to possible intracranial problem.
   f. Neuro consult reveals no neurologic deficits with in-office testing. MRI ordered based on threshold VF alone.
   g. MRI reveals large arteriovenous malformation (AVM) in the left parietal-occipital region with a nidus of 2.5 x 3 x 3.5 cm (large). Surgical excision is recommended.
   h. Pt. schedules surgery, then cancels. Six months later, he has a seizure and re-schedules surgery. AV malformation is removed successfully in two procedures with no neurological deficits.

III. Differential diagnosis based on VF
   a. Intracranial space occupying lesion
   b. Glaucoma

IV. Diagnosis and discussion
   a. Neurological AVM are developmental anomalies with multiple feeding arteries leading to a nidus (nest) of vessels, and draining into one or more cerebral veins. There are no capillaries separating the arteries from the veins. 300,000 Americans are affected. Brain MRI of our patient will be shown and discussed.
   b. The annual hemorrhage rate of an untreated AVM is 1-4%. The rate of serious morbidity or mortality is 30% or greater in a ruptured AVM.
   c. AVM embolization (surgical treatment) itself has significant morbidity (2-3%) and mortality (1%) with transient deficits of 5-10%. Therefore, the decision for treatment is difficult, especially in an AVM that is causing no symptoms.
   d. Patients can present to the optometrist with headache and transient vision obscurations. For many patients, the first symptom is a seizure or stroke.

V. Treatment and management
   a. Embolization of the AVM is possible under general anesthesia (GA) or IV sedation. GA has the advantage of a still patient resulting in better image quality to guide the surgeon. IV sedation allows the surgeon to question the patient during the procedure to reduce the risk of neurological insult.
b. The surgeon must block the arterial flow to the AVM while ensuring that those arteries do not feed other parts of the brain while assuring that the veins draining the AVM are not obstructed prematurely resulting in hemorrhage.

c. Radiosurgery can be used in combination with embolization or alone if the nidus is small enough. Obliteration of the AVM after treatment can take 1-3 years.

d. Post-surgical considerations include repeat VF and referral for mobility services if functional vision loss. If there is neurological deficit, proper therapies should be instituted.

VI. Conclusion
a. A patient was diagnosed with an intracranial AVM upon routine visual field testing
b. AVM typically have no symptoms until they hemorrhage with a high morbidity and mortality rate.

c. Optometrists can play a key role in diagnosing these patients before AVM hemorrhage, potentially saving their lives.

d. It is possible to have intracranial anomalies without headache.

VII. References
Twelve Years Later, Trouble Returns

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Jesse Brown Veterans Affairs Medical Center

Disclosure Statement:
Nothing to disclose

Case History

- Chief Complaint:
  - c/o HA x 3 days with nausea and vomiting
  - Blurred vision since HA began
  - Denies trauma
- Ocular History
  - Blunt trauma with baseball OS age 10
  - Glaucoma suspect, previously tx by outside doctor
  - Pseudophakia OD, OS-2003 by outside doctor

Case History

- Medical History-Non-contributory
- No Known Medical Allergies
- No current ocular or systemic medications

Initial Presentation

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<td>Open to CBB, flat approach, 180° angle recession</td>
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<tr>
<td>Lens</td>
<td>PCIOL</td>
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Initial Presentation

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#aaoptom14
Differential Diagnosis

- Phacoanaphylactic Glaucoma
- Delayed Onset Pseudophakic Endophthalmitis
- Lens Particle Glaucoma

Phacoanaphylactic Glaucoma

- Granulomatous inflammation directed at previously sequestered lens material
- Can begin within 1 day to couple weeks after inciting event
- Inflammation can range from mild uveitis to hypopyon
  - Keratic precipitates are common, PAS and posterior synechiae can form
- Definitive treatment is surgical removal of lens material

Delayed Onset Pseudophakic Endophthalmitis

- Presents 6 weeks or more after surgery
- Low grade inflammation persists for months
- Can present with white capsular plaque
- Most commonly caused by Propionibacterium Acnes, less commonly Staphlococcus Epidermidis or Candida Albicans
- Treated surgically with vitrectomy, capsulotomy and IOL removal or exchange

Len Particle Glaucoma

- Hypothesized that lens particles as well as phagocytotic macrophages mechanically obstruct flow through the TM
- Initial treatment with steroids & IOP lowering agents
- If initial therapy fails, surgical removal of the lens material is warranted

Back To Our Case

- Low grade inflammation
- Cortical material now presenting in anterior and posterior chambers TWELVE YEARS AFTER SURGERY
- High IOP
- Diagnosis: Lens Particle Glaucoma

Twelve Years After Initial Surgery
Initial Treatment

- 1 gtt Latanoprost, Cosopt & Brimonidine in office
- 2x250mg Diamox PO
- IOP lowered to 28 mmHg in office
- Began PredForte Q1H, Cosopt BID OS, Brimonidine BID OS
- Follow up in 3 days

Follow Up #1

- Headache & nausea resolved, blurred vision persists

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- Decrease Pred Forte to q2H OS, continue Cosopt BID OS, Brimonidine BID OS, follow up in 2 weeks

Follow Up #2

- Feels vision is improved, no headache or nausea

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- Continue present management, follow up in 2 wks

Follow up #3

- Blurred vision has returned, though improves after first 3 doses of PredForte, no headache or nausea

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- Dilated fundus exam stable.
- Add Latanoprost qhs OS, continue all other meds, follow up 2 weeks

Follow Up #4

- Vision still blurred, no headache or nausea

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- Dilated fundus exam stable.
- Refer to ophthalmology for surgical management

Surgical Management

- Evaluated by both cornea and glaucoma specialists
- Given angle recession the patient may have had compromised aqueous outflow prior to cataract extraction
- Recommended to have cortical aspiration and Ahmed valve placed
Post Op Follow Up

- Vision has improved
- Ahmed valve functional at 1 month post op
- IOP hovering in the mid-teens on Cosopt and Brimonidine

60 year old man underwent extracapsular cataract extraction and was left aphakic
- No visible lens fragments on SL exam, though cytology showed lens material and phagocytotic macrophages
- IOP was uncontrolled on medications
- Surgical intervention controlled IOP and inflammation

60 year old male presented with pain and blurred vision 15 years after extracapsular cataract surgery
- Initially treated with topical agents but IOP remained uncontrolled
- Surgically aspirated retained cortical material from anterior and posterior surface of PCIOL
- Aqueous humor evaluated showed macrophages engulfed lens particles

Obstruction of aqueous outflow by lens particles and by heavy-molecular-weight soluble lens proteins

David L. Epstein, Judith A. Jedziniak, and W. Morton Grant

Emulsified human eyes were perfused via the anterior chamber at 25 mm Hg pressure with lens particles (whole lens homogenate) in one series of experiments and with soluble lens proteins from human cataractous lenses in another series. Adding 1% of a homogenate of single cataractous lens to the anterior chamber induced a 66% decrease in outflow. Perfusion with 10&#x200f;10<sup>6</sup> soluble lens proteins (1 mg/mL, MW more than 120 million) caused a 20% decrease in outflow in 1 hr. In neither series was the obstruction to outflow relieved by subsequent irrigation of the anterior chamber with balanced salt solution or alpha-chymotrypsin. The results show that both lens particles and soluble lens proteins can directly obstruct the aqueous outflow pathways of human eyes. Such obstruction may be a significant factor in certain lens-induced glaucomas.

Fig. 1. Effect of cataracts on aqueous outflow. Percent of the homogenate of a single cataract was added to the anterior chamber of an emulsified human eye. Aqueous was aspirated after a preliminary 1-hr aqueous perfusion and measurement of outflow. Results are shown for six homogenate-treated eyes (filled circles) and for six detergent-treated controls (triangles).
Clinical Pearls

- Lens particle glaucoma can occur years after cataract extraction
- Must rule out other differential diagnoses

References


I. Background
   A. Initial documentation of findings similar to acute retinal necrosis (ARN) syndrome was reported in 1971 Japan.
   B. Young and Bird published the first account of “bilateral acute retinal necrosis (BARN)” in 1978.
   C. Often, ARN was misdiagnosed until the American Uveitis Society set forth standard diagnostic criteria for ARN in 1994.

II. Epidemiology
   A. Reported 1 case per 1.6-2 million population per year in the UK.

III. Clinical features
   A. Most common: sudden vision loss ~85%
   B. Photophobia/Ocular pain/Conjunctival injection
   C. Anterior uveitis/Vitritis/Peripheral retinal involvement ~80%
   D. Multifocal retinal lesions that gradually enlarge and fuse
   E. In the acute phase often only retinal arterial involvement is appreciated and fluorescein angiography reveals “knob-like dye leakage” or “string-of-beads appearance”.

IV. Diagnosis
   A. Standard diagnostic criteria for acute retinal necrosis by the Executive Committee of the American Uveitis Society:
      1. at least one foci of retinal necrosis with discrete borders in the peripheral retina
      2. rapid progression of necrosis in the absence of anti-viral therapy
      3. circumferential progression
      4. occlusive vasculitis with arteriolar involvement
5. pronounced anterior chamber and vitreal inflammation
   i. *Causative agents and the status of ones systemic health are not diagnostic factors*

B. Vitreal biopsy for virological evaluation of a causative agent using Polymerase chain reaction is imperative for diagnosis/treatment
   1. Causative agents: Varicella-zoster virus, Herpes simplex virus type 1, Herpes simplex virus type 2, Cytomegalovirus
      a. Herpes simplex virus type 2 more likely to cause ARN in patients <25 years of age
      b. Herpes simplex virus type I and Varicella-zoster are more likely to cause ARN in patients >50 years of age

V. Primary Differentials
   A. Progressive Outer Retinal Necrosis
      1. Distinguishing feature: usually *no* retinal vasculitis, vitreal or anterior segment inflammation.
   B. Cytomegalovirus Retinitis
      1. Distinguishing feature: confluent retinal necrosis with *extensive* hemorrhaging that develops in most cases in the *posterior pole*
   C. Syphilis
      1. Distinguishing feature: a *chorioretinitis* is one of the *most frequent* ocular manifestations with syphilis. Additionally “ground glass” retinitis and associated vasculitis are very characteristic of Syphilis
   D. Sarcoid
      1. Distinguishing feature: a retinal vasculitis is common however limited to the retinal *veins* and “*candle wax drippings*” seen as peri-venous exudates are characteristic
   E. Tuberculosis
      1. Distinguishing feature: usually presenting as a choroiditis with choroidal tubercular granulomas. Vasculitis may occur, and is more commonly seen affecting the *veins* than arteries
VI. Treatment/Prognosis

A. No definitive evidence based treatment
   1. Accepted current standard of treatment is intravenous Acyclovir 7-10 days
      and long term oral Acyclovir. Intra vitreal anti-viral is an additional option
      as well.
   2. Presenting visual acuity and retinal detachment occurrence determine
      visual prognosis
   3. Oral prednisolone usually considered 24-48 hours following the start of
      anti viral therapy

B. High potential for contralateral eye involvement if untreated
   1. Relatively low risk for contralateral eye involvement with standard
      treatment regimen

C. ~50-80% risk of retinal detachment with anti-viral treatment

VII. Pearls

A. Acute retinal necrosis is a rare retinitis and a devastating ocular emergency. If
   it exists as a differential then treat promptly and accordingly as ocular
   morbidity is extremely high

B. Key feature of acute retinal necrosis and imperative to this case especially:
   1. Fluorescein angiography: occlusive arteritis
References
