Acute Anterior Uveitis with Hypopyon

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Disclosure

- The presenter and Organizer for “Acute Anterior Uveitis with Hypopyon”
- Dr. Ryan Ngo has no financial relationship with any company or products mentioned in this presentation.

Case Study

- 29 year old Caucasian male walk in visit
- CC: Significant left eye pain (8/10) that started 3 days ago
- Associated symptoms: Significant photophobia and blurred vision

Ocular History

- Patient’s Ocular history
  - Anterior uveitis OS x 2018 - initial
  - Anterior uveitis OD x 2019 - work up ordered, but not completed
- Medical history
  - History of uncontrolled Type 1 diabetes, Last A1C: 12.5%
  - Erectile dysfunction
  - Eczema
- Medication
  - Insulin, sildenafil, triamcinolone acetenoid 0.1% exam for eczema

Patient’s 2019 Lab Work Up

<table>
<thead>
<tr>
<th>Test</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>HLA b27</td>
<td>Negative</td>
</tr>
<tr>
<td>Rheumatoid Factor</td>
<td>Negative</td>
</tr>
<tr>
<td>Dilute plasma regain (PPR)</td>
<td>Non-reactive</td>
</tr>
<tr>
<td>Angiotensin converting enzyme (ACE)</td>
<td>55</td>
</tr>
<tr>
<td>QuantIFERON</td>
<td>Negative</td>
</tr>
</tbody>
</table>

Examination

- Entrance Examination:
  - VA 20/15 OD; 20/20 OS
  - Habs, Brunning (O); Brunning (O)
  - Pupils / PS (-) (-)
- MRI large Examination- OIL:
  - Corneal: 8.1 x 6.0 x 10.0 mm, Intact outer zone, and outer ring of intraretinal fluid with surrounding hypodensity
  - Cystoid: 6.1 x 6.1 mm, drusen, macular edema
  - ARI: 1.0 x 0.8 x 0.8 mm, Intact outer zone, and outer ring of intraretinal fluid with surrounding hypodensity
- Orbital Fundus Examination:
  - Chemosis: 6.1 x 6.1 mm, Intact outer zone, and outer ring of intraretinal fluid with surrounding hypodensity
Assessment/Plan

Assessment:
- Recurrent nongranulomatous anterior uveitis OS

Plan:
- Significant improvement in symptoms (pain and photosensitivity) with loading dose of 1gtt PredForte every 5 minutes for 30 minutes in office
- Prescribed PredForte Q1H and atropine BID while awake
- RTC 1 week for follow-up

Anterior Uveitis

- Inflammation of the anterior chamber

Anterior Uveitis: Epidemiology

- 5th leading cause of blindness in the worldwide
- Most common inflammation eye practitioners will see
- 15/100,000 affected, 45,000 new cases every year
- Common between second and fourth decades
- 50% idiopathic, HLA b27 is the most common type

Anterior Uveitis: Pathophysiology

- Breakdown of blood aqueous barrier
- Release of fluids and cells
- Hypopyon
- Keratic precipitates
- iris damage
- Cataract
- Macular edema
- Iris changes (atrophy, heterochromia, koeppen /busacca nodules)
- Circumlimbal injection
- Fibrinous material

Acute Anterior Uveitis: Symptoms

- Symptoms:
  - Pain
  - Redness
  - Photophobia
  - Lacrimation
  - Mild decreased vision

Hypopyon

- Indication of severe inflammation in the anterior chamber
- Consists of tissue debris, inflammatory byproducts, and recruited leukocytes
- Uncommon finding in uveitis
Types of Hypopyons

Classification of Causes of Hypopyon by Ramsay

Differential Diagnosis: HLA-b27 uveitis
- HLA-b27 antigen is a class I major histocompatibility complex
- Second most common nongranulomatous etiology of anterior uveitis up to 50% of acute anterior uveitis with 70% recurrence
- Signs more severe in presentation
- Ages 20-40 years of age
- 2.5x F>M
- Usually recurrent unilateral, bilateral, or alternating nongranulomatous anterior uveitis and may have fine endothelial KPS

Differential Diagnosis: Bechet's Disease
- Chronic, recurrent, and multisystem mucocutaneous inflammatory disorder
- Classic triad: genital and oral ulcers, skin lesions, HLA
- Found along ancient silk-road extending from eastern Asia to the Mediterranean basin
- 10-15% of patients, uveitis is the initial manifestation of the disease
- Affects ages between 20-40s

Differential Diagnosis: Bechet's Uveitis with Hypopyon
- Usually presents with acute nongranulomatous anterior and/or posterior uveitis
- About 28-35% commencement with hypopyon
- Considered as "cold" hypopyon
- Shifts with head positioning
- HLA hypopyon considered as "hot" HLA
- Hypopyon forms and dissolves rapidly
Infectious causes of hypopyon
- Toxocariasis
- Syphilis
- Leprosy
- Herpetic uveitis
- Tubercular uveitis

Diabetic Anterior Uveitis
- Several studies suggest patients with diabetes have a higher incidence of acute uveitis, while those with type 1 and poor glycemic control are at higher risk of developing eye inflammation.
- Studies have shown a positive correlation between hyperglycemia and inflammation in anterior chamber in patients with anterior uveitis.
- Behaviors of uveitis in these patients is more aggressive and occurs more often bilaterally.

When to order lab work up
- Uveitis unresponsive to steroids
- Bilateral
- Alternate, Recurrent
- Positive review of system or systemic examination

Lab work up
- Colonoscopy: Inflammatory bowel disease (Crohn’s & ulcerative)
- Chest X-ray: Sarcoidosis & TB
- Sarcoiliac X-ray: Ankylosing spondylitis
- HLA-B27: Inflammatory Bowel disease (Crohn’s & ulcerative)
- HLA-B51: Behcet’s
- ANA: Juvenile Idiopathic Arthritis
- PPD/Quantiferon: Tuberculosis
- RPR/VDRL & FTA-ABS/TPPA/MHA-TP: Syphilis
- ELISA: Lyme Disease
- CBC: General health status (anemia, infection, leukemia)
- CRP: Determine cause or location of inflammation of the body
- ESR: With CRP, detect inflammation, serves as a monitor for underlying etiology

Goals for managing Acute Anterior Uveitis
- 1.) Aggressively resolve ocular inflammation to prevent potential of visual loss & relieve ocular pain
  - Treat any infectious etiologies
- 2.) Determine underlying systemic etiology and when indicated, make appropriate referral for evaluation and treatment of condition

Conventional Treatment for Acute Anterior Uveitis
- 1. Topical corticosteroids
  - Prednisolone acetate 1%
  - Difluprednate 0.05%
- 2. Cycloplegics
  - Atropine 1% MD & cyclopentolate 1% TD
  - Homatropine and scopolamine
  - *Treat any infectious etiologies
Immunosuppressants

- Indications:
  - Noninfectious Uveitis
  - Mainly for recalcitrant cases that are unresponsive to conventional therapy
  - Autoimmune diseases (HLA-B27 associated uveitis, Bechet’s…)

Adalimumab
(Humira, AbbVie)

- Biologic proteins
- Tumor necrosis factor (TNF) blocker
- Specific source of inflammation that appears to have a role in uveitis
- FDA approved in 2016
- By blocking it, the inflammatory effect of uveitis is reduced
- Requires baseline screening for tuberculosis and hepatitis, should be avoided in patients with demyelinating disorders
- Cost: $4,500 per month

Follow up #1

- CC: worsening vision OS, but improvement in redness & pain
- Current ocular medications: Pred Forte Q1H and Atropine BID only

Assessment/ Plan

- Assessment:
  
  - Alternating recurrent non-granulomatous anterior uveitis, OS
  
  - HLA-B27, seronegative, Ty. Sarcoidosis, Demyelinating

- Plan:
  
  - Switched to Durezol q2h while awake
  
  - Continue Atropine
  
  - Add brimonidine BID to assist with elevated IOPs

Follow up visits #2: day 7

- Assessment:
  
  - Alternating recurrent non-granulomatous anterior uveitis, OS
  
  - Ty. Sarcoidosis, seronegative

- Plan:
  
  - Continue Durezol hourly, brimonidine bid & prescribed Tobradex qhs
  
  - Ordered lab work up: Chest X-ray & blood draws
Follow up visits #3: day 10

Assessment:
- Alternating recurrent non-granulomatous anterior uveitis, OS
  - Significant improvement in symptoms: VA 20/40
  - 1+ Cell with small amount of fibrin membrane over pupil but no hypopyon
  - States he has issues with dry skin, but no actual lesions. Has had one mouth ulcer but that was when he had a wisdom tooth abscess

Plan:
- Continue Durezol hourly, brimonidine bid & continue tobradexung qhs
- Did not get chest x-ray or labs, recommended lab work up today
- RTC 1 week for follow up

Follow up visits

- Cancelled and no showed 4x

Conclusion

- Hypopyon uveitis is an uncommon occurrence in uveitis
- Result of inflammatory, infective, neoplastic, or therapeutic stimuli
- Requires close communication and co-management with rheumatologist

Special Thanks

- Doctors of Spokane VAMC
  - Dr. Jeffrey Urness
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  - Dr. Len Koh
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  - Dr. Tom Kollodge
  - Dr. Lindsay Kleinschmit
  - Dr. Anna Wells
  - Dr. Allison Makadia

References

- Kopplin, L. When to consider systemic treatments (2020). Retinal Specialist A publication by Review of Ophthalmology, 6(3).
Symblepharon: A Case Study

Lora Cretella, O.D.
Spokane VAMC
June 12th, 2018

Course Objectives

1. Recognize various clinical presentations of symblepharon.
2. Estimate risk of progression and sight loss based on clinical history and presentation.
3. Understand mechanism of symblepharon formation and most common underlying conditions.
4. Know appropriate treatment and referral.
5. Explain different treatment courses which might be employed.

Disclosures

The presenter and organizers for "Symblepharon: A Case Study" by Dr. Lora Cretella has no financial relationship with any company or products mentioned in this presentation.

The Patient

- CC: 74 yo WM presents for DFE F/U
- POH: CE/IOL 7/2019 OU, H/O Drance Heme
- PMH: DM, Coronary Artery Disease, BPH, Hypertension, Dyslipidemia, Colon Polyps
- FMH: Unremarkable
- FOH: Unremarkable
- SH: Unremarkable
- Meds:
  - AT's Refresh + Genteal
  - Aspirin 81 mg
  - Atorvastatin
  - Hydrochlorothiazide
  - Losartan
  - Metformin
  - Metoprolol
  - Triamcinolone cream
- Allergies - PCN, Fosinopril
- A1C: 6.7

The Exam

- BCVA: OD: 20/20-- OS: 20/20--
- IOP: OD: 15 mmHg OS: 15 mmHg
- CVF: FTFC OU
- PUPILS: PERRL
- EOM: FROM

The Exam

- BCVA: OD: 20/20-- OS: 20/20--
- K/R: OD: 0.6 Sc 3
d- OS: 0.6 Sc 3
- PUPILS: PERRL
- EOM: FROM
Anterior Segment Exam

<table>
<thead>
<tr>
<th>FINDING</th>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>ADNEXA</td>
<td>Unremarkable</td>
<td></td>
</tr>
<tr>
<td>LIDS</td>
<td>Mild ectropion, complete voluntary closure</td>
<td></td>
</tr>
<tr>
<td>CONJ</td>
<td>White and quiet</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Fornix shortened temporally</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Symblepharon of palpebral conjunctiva near lid margin and inferior bulbar conjunctiva</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Associated subtemporal subconjunctival bulbar (3-6 o'clock) and palpebral hemorrhage</td>
<td></td>
</tr>
<tr>
<td>CORNEA</td>
<td>Clear</td>
<td></td>
</tr>
<tr>
<td>ANT CHAMBER</td>
<td>D/Q</td>
<td></td>
</tr>
<tr>
<td>EYES</td>
<td>Flat</td>
<td></td>
</tr>
<tr>
<td>LENS</td>
<td>PCOIL Clear + Centered</td>
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</table>

Posterior Segment Exam

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<th>OD</th>
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</thead>
<tbody>
<tr>
<td>C/O</td>
<td>0.40/0.45</td>
<td></td>
</tr>
<tr>
<td>NERVES</td>
<td>Distinct margins, good color</td>
<td></td>
</tr>
<tr>
<td>MACULA</td>
<td>Flat, annular configuration</td>
<td></td>
</tr>
<tr>
<td>POSTERIOR POLE</td>
<td>Unremarkable</td>
<td></td>
</tr>
<tr>
<td>AVASINO</td>
<td>2-3</td>
<td></td>
</tr>
<tr>
<td>PERIPHERY</td>
<td>Flat, 50% m Palmer/teams/attachments</td>
<td></td>
</tr>
<tr>
<td>VITREOUS</td>
<td>PVD, Grade Clear</td>
<td></td>
</tr>
</tbody>
</table>

BLOOD PRESSURE:
- RAS: 196/88 @ 9:54 am *Patient reports not taking his 9am pill today*
- RAS: 177/82 @ 10:58 am *after instructed to take hypertensive med*

Assessment

1. Symblepharon OS>OD
   - New
   - Asymptomatic
   - Likely HTN related
   - Potentially related to subconjunctival hemorrhage

2. Subconjunctival Hemorrhage
   - New
   - Asymptomatic
   - Likely HTN related
   - Potentially related to symblepharon formation

Plan

1. Symblepharon
   - PredForte QID OU x 10 days then BID until F/U
   - RTC 1 month w/ in house Ophthalmologist
   - Pt ed on condition and need for F/U testing

2. Subconjunctival Hemorrhage
   - Pt ed on blood pressure today, risks of uncontrolled BP, and need for PCP consult.
   - Pt instructed to take hypertensive med.
   - PCP notified.
   - Educated on expected resolution.
   - RTC if not resolving.

3. HVF / Pachymetry / Glaucoma Work up
4. Monitor Annually
5. Continue Simpatine
6. Dry Eye
7. Ectropion OU

Follow up #1 — Findings

- All findings stable
- Symblepharon OS>OD
- Pseudophakia
- Paracentesis OU
- OD usual astigmatism
- OS 2 separate strands temporal and nasal
- Subconjunctival hemorrhage resolved
Follow up #1 --- Assessment

- Symblepharon OS>OD
  - Asymptomatic
  - No intervening trauma other than cataract surgery
  - No other skin changes to suggest SJS
  - Referral to corneal specialist for biopsy to look for ocular cicatricial pemphigoid

Top differentials

- Traumatic scarring secondary to cataract surgery
- Progressive cicatrization due to ocular pemphigoid
- Cicatrization due to medication reaction

Conjunctiva

- Mucous membrane covering the globe and inner eyelids
- Clear tissue
- Contains goblet cells

Fornix

- Caucasians:
  - 15.6 mm upper
  - 10.9 mm lower
  - (~0.3 mm less for females)
  - Progressive decline with age

Symblepharon

- Adhesion of palpebral to bulbar conjunctiva
- Can eventually involve cornea
**Signs of Conjunctival Scarring**
Visible adhesions, conjunctival fibrosis, and fornix shortening.

**Symptoms**
- **EARLY:**
  - Itching
  - Non-specific: redness, stinging, dryness, watering
  - (Goblet cells affected)

- **LATE:**
  - Restricted eye movements
  - Poor cosmesis
  - Reduced vision
  - Discomfort from eyelid malposition

**Complications**
- Dry eye
- Injection
- Inadequate blinking
- Eyelid malposition / entropion / ectropion
- Restricted range of motion
- Corneal involvement w/ reduced vision

**Predicting?**
- The more conjunctival area injured, the more likely
- More common in areas in apposition
- More likely inferior, can be superior

**Important clinical questions**
- Underlying cause?
- Systemic or ocular?
- Self limiting vs. progressive?
- Is this a blinding condition?
- Surgical intervention?

**Categorizing**
- Self limited
  - Chemical/thermal burns
  - Infectious diseases
  - Adenovirus
  - Herpes virus
  - Chlamydial conjunctivitis

- Progressive
  - "Cicatrizing conjunctivitis"
  - Mucous Membrane Pemphigoid (MMP)
  - Stevens-Johnson syndrome (SJS)
  - Toxic Epidermal Necrolysis (TEN)
Squamous papilloma of the conjunctiva

Xeroderma pigmentosum

Porphyria cutanea

Ectodermal dysplasia

Sjogren's

Sarcoid

Host vs. graft disease

Neoplastic growth

Lupus

Lichen planus

Ocular rosacea

Atopic keratoconjunctivitis (AKC)

Diagnosis often delayed due to unfamiliarity

HLA-DR4 gene increases susceptibility

2:1 women: men

Onset age 60-80

No racial predilection

Can be many etiologies, but most commonly linked to OCP, SJS, TEN

Most commonly Mucous Membrane Pemphigoid (60%)

Cicatrizing Conjunctivitis

7.7-18% of all ocular trauma

Larger defect → more extensive scarring

Superior burn can still permeate

Alkali more penetrating and traumatic (ammonia, lye)

Healing takes weeks. Occurs in phases.

42% will progress even with no clinical inflammation "White inflammation"

75% untreated will progress

End: pannus / blindness

Late: fornix shortening / symblepharon

Early: recurrent inflammation (mimics dry eye, conjunctivitis)

Usually starts unilaterally, bilateral in 2 years

Symptoms vary widely: burning/dryness to scarring / blindness

Mucous Membrane Pemphigoid (MMP)

Subset of systemic autoimmune pemphigoid diseases

60-80% present with ocular/conjunctival involvement

Same as Ocular Cicatricial Pemphigoid (OCP)

Affects all mucous membranes:

genital, oral, ocular

Mucous Membrane Pemphigoid (MMP) Courses

Symptomology:

- Symblepharon in late phase healing, 3-4 weeks
- Fornix shortens
- Conjunctival sac volume decreases

Superior damage → more necrosis → delayed epithelization

 Inferior damage → more necrosis → delayed epithelization

Ocular Burns

27-28% of all ocular trauma

Alkali more penetrating and traumatic (ammonia, lye)

Superior burn can still permeate → inferior damage

Healing takes weeks. Occurs in phases.

Cicatrational ulcers / burns

Fat necrosis

Symblepharon in late phase healing, 3-4 weeks

Larger defect → more necrosis → delayed epithelization

→ more extensive scarring

"Cicatrizing Conjunctivitis"

Progressive loss of conjunctival scarring associated with complications

Most commonly Mucous Membrane Pemphigoid (100%)

Can be many etiologies, but most commonly linked to OCP, SJS, TEN
**MMP: Mechanism**

- Type 2 hypersensitivity: cytotoxic attack on conjunctival basement membrane
- Unclear exact antigen:
  - protein BP180
  - alpha-6 beta-4 integrin of hemidesmosomes
- Confirmed by biopsy w/ direct immunofluorescence microscopy (DIF)

**MMP: Staging**

- Stages of Ocular Cicatricial Pemphigoid (simplified)
  - Stage I: Subconjunctival fibrosis
  - Stage II: Forniceal shortening
  - Stage III: Symblepharon
  - Stage IV: Keratinization of the ocular surface, Ankyloblepharon

**Mondino's Classification System**

- Stage I: up to 25% inferior fornical depth loss
- Stage II: 25-50% inferior fornical depth loss
- Stage III: 50-75% inferior fornical depth loss
- Stage IV: >75% inferior fornical depth loss

**MMP: Treatment**

- 75% progress without treatment
- 10% progress despite treatment
- Anti-inflammatory and immunomodulatory
  - Dapsone for mild
  - Corticosteroids if moderate/severe
  - Rituximab/infliximab
- 2-3 years of stability, perhaps D/C medications
- 22% of patients relapse
- Surgical repair

**Steven's Johnson Syndrome (SJS)**

- Acute inflammatory of skin + mucous membranes
- Adverse reaction to medications:
  - Usually sulfonamides, also NSAIDS, anticonvulsants, antigout
- Acute, life-threatening blistering and necrosis, followed by chronic scarring
Toxic Epidermal Necrolysis
- More severe, toxic variant of SJS
- SJS: <10% of surface area
- TEN: >30% of surface area
- Rare: SJS + TEN = < 8 cases in 1 million yearly

Early: Mucopurulent conjunctivitis, chemosis, hyperemia
Late: scarring

Topical Treatment?
- Each case is different and based on underlying etiology
- No proven improvement to MMP with topical medications / steroids
- Lubrication to help relieve symptoms

Prevention?
- 1 ounce of prevention = a pound of treatment!
- Consider Symblepharon rings
- Prokera lens similar effect
- Anti-inflammatory and anti-scarring
- Large conjunctival defects, especially inferior, consider Prokera lens
- If very large, can consider tissue transplant

Surgical Treatment
- Historically, glass rod to separate
- Surgical repair
  - Restoring the fornix, separate conjunctiva, cover defects
  - Limbal graft from opposing eye, amniotic membrane, or oral mucosa transplant
- Outcome depends on stability of inflammation.
  - Stability before surgery

Clinical Pearls
- Any signs of conjunctival scarring, closely reassess OCT/Tonometry
- Educate patients on many possible causes, but need to rule out the most visually devastating.

Special Thanks
- Doctors of Spokane VAMC
  - Dr. Jeffrey Urness
  - Dr. Megan McChesney
  - Dr. Ryan Ngo
  - Dr. Tom Kollodge
  - Dr. Lindsay Kleinschmidt
  - Dr. Anna Wells
  - Dr. Len Koh
  - Dr. Allison Coit
  - Dr. Chad Gosnell
References


**Limbal Dermoid**

**A CASE REPORT & CLINICAL REVIEW**

THOMAS KOLLODGE, O.D.
OCULAR DISEASE RESIDENT
MANU-GRANDSTAFF VA MEDICAL CENTER
SPOKANE, WASHINGTON

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**Course Objectives**

- Recognize and identify common clinical features of limbal dermoids
- Describe various associated conditions and abnormalities
- Differentiate between limbal dermoids and other anterior segment pathology
- Explain when to refer for surgical management

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**Case LJ**

73 year old Caucasian male

Comprehensive eye exam, no complaints

Medical history:
- Bilateral sensorineural hearing loss
- Subjective tinnitus

Not taking systemic medications

Family history: Non-contributory

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**Case LJ – Entrance Testing**

Refraction/BCVA
- OD: +0.25 -0.75 x 097 20/20
- OS: -0.25 sphere 20/20

Pupils: ERFL, no APD OD/OS

EOMs: FROM OU

Confrontations: FTFC DD/OS

No gross abnormalities of head, neck, or ears

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**Overview**

- Case LJ
- Histology
- Clinical presentation
- Grading
- Epidemiology
- Ocular associations
- Systemic associations
- Differential Diagnoses
- Management
- Summary slide
- Conclusion

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**Disclosures**

The Presenter and Organizers for:

“Limbal Dermoid: A Case Report and Clinical Review”

By Dr. Thomas Kollodge has no financial relationship with any company or products mentioned in this presentation.
Case LJ – Anterior Segment

Ocular adnexa: Unremarkable OD/OS

Lids: Unremarkable, no coloboma OD/OS

Conjunctiva: Clear and quiet OD/OS

Iris: Flat without coloboma OD/OS

Anterior chamber: Deep and quiet OD/OS

IOP:
  - OD: 13 mmHg
  - OS: 12 mmHg

Sclera:
  - OD: White and quiet
  - OS:
    - Non-moveable, superficial, elevated mass within palpebral fissure, partially on temporal cornea and sclera
    - Located at ~4 o'clock relative to the cornea
    - Round, pale yellow to white in color, and approximately 4.5 mm in diameter
    - Extended approximately 2 mm onto the cornea
    - No abnormal vasculature

Cornea:
  - OD: Clear
  - OS: Band of haze parallel to the lesion’s encroachment

Pt reported lesion present and stable his entire life

Case LJ – Anterior Segment

Lens: Trace nuclear sclerosis and trace cortical spoking OD/OS

Optic nerve: Pink and perfuse, no coloboma OD/OS

C/D ratio:
  - OD: 0.20/0.20
  - OS: 0.15/0.15

Macula: Flat with even pigmentation OD/OS

Posterior pole:
  - OD: Unremarkable
  - OS: Flat, round, darkly pigmented, 1/8th disc-diameter CHPE inferior/nasal to optic nerve

Blood vessels: Healthy, 2:3 artery/vein ratio OD/OS

Periphery: Flat and attached 360 OD/OS

Vitreous: Clear OD/OS
Case LJ

Assessment:
1. Limbal dermoid OS
2. Cataract OU
3. CHRPE OS
4. Refractive error OU

Plan:
1. Monitor
2. Defer surgery until functional vision worse
3. Monitor
4. Spectacle Rx released

Limbal Dermoid Histology

Choristoma: Tumor composed of normal tissues in abnormal location
Congenital benign tumor derived from mesoderm and ectoderm
Form during embryogenesis when cells are accidentally captured inside developing tissues
Made up of dense connective/collagenous tissues with pilosebaceous units
Covered with stratified squamous epithelium
Can contain sweat glands, fat, lacrimal gland, and neurologic tissues
No known malignancy potential

Limbal Dermoid Histology

Subepithelial fibrous tissue resembling reticular dermis with skin appendages

Typical Clinical Presentation
Elevated, smooth, white/yellow, solid, soft, well-circumscribed subconjunctival mass
Found at birth or during early childhood
Unilateral or bilateral
Inferotemporal or central limbus
Can partially or completely involve the cornea
Can have hair protruding from their surface
Size: Hardly observable to 5+ mm
Corneal lipid deposition can occur near the dermoid edge
Growth is uncommon, could occur during puberty
Grading

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Superficial and smaller than 5 mm in diameter</td>
</tr>
<tr>
<td>II</td>
<td>Extend into corneal stroma and Descemet’s membrane, typically larger than grade I</td>
</tr>
<tr>
<td>III</td>
<td>Affect the entire cornea, extend into anterior chamber, and disrupt all structures between the pigmented epithelium of the iris to the anterior surface of the eye</td>
</tr>
</tbody>
</table>

Epidemiology

Rare

Choristomas are the most common category of epibulbar tumors of young children and infants

Limbal dermoids are the most frequent epibulbar choristoma, occurring in approximately 1 out of 5,000-10,000 people

Ocular Associations

- Iris coloboma
- Eyelid coloboma
- Lacrimal abnormalities
- Corneal staphyloma
- Scleral staphyloma
- Aniridia
- Microphthalmos

Systemic Associations

- Goldenhar syndrome – craniofacial disorder
- Treacher Collins syndrome – craniofacial disorder
- Linear nevus sebaceous of Jadassohn – genetic condition
- SCALP syndrome – nevus sebaceous, ONL malformations, galea cutis congenita, limbal dermoid, and pigmented nevus

Differential Diagnoses

Conjunctival dermolipoma
- Choristoma
- Similar cellular composition to a dermoid
- Typically more yellow in color due to increased fat
- Most often superotemporal bulbar conjunctiva
Differential Diagnoses

Orbital fat prolapse
- Similar appearance to dermolipoma
- Movable
- Yellow-colored mass
- Subconjunctival
- Supertemporal location
- From weakening of Tenon’s capsule due to surgical trauma or aging

Pinguecula
- Area of thickening of the bulbar conjunctiva
- Fatty appearance
- Usually occur nasally within the palpebral fissure
- Associated with increased age and ultraviolet light exposure

Pterygium
- Fibrovascular growth extending from conjunctiva onto cornea
- More often within the nasal palpebral fissure
- Associated with ultraviolet light exposure

Conjunctival/corneal squamous cell carcinoma
- Leukoplakic: Discrete thickening of conjunctiva with overlying white hyperkeratotic plaque (top picture)
- Papillomatous: Extremely vascularized mass (bottom-right)
- Gelatinous: Translucent thickening of conjunctiva (bottom-left)
- All typically present with prominent afferent and efferent blood vessels
- Associated with older age, UV light exposure, and squamous cell carcinoma of the skin

Management

Almost always benign
- Can monitor in clinic
- Reasons to refer for removal
  - Induced astigmatism → refractive amblyopia
  - Blocking visual axis → deprivation amblyopia
  - Chronic irritation from hairs or dellen formation
  - Cosmesis (especially for children)

Management

Various surgical excision methods
- Simple excision
- Lamellar dissection/excision with penetrating sclerokeratoplasty
- Repair after excision
- Lamellar corneoscleral graft
- Intrastromal lenticule graft from SMILE
- Amnion membrane transplantation
- Reconstruction with anterior corneal button from donor DSAEK tissue
- Method/procedure depends on
  - Grade of limbal dermoid
  - Surgeon preference
  - Resource availability (donor corneas)
Removal

Limbal dermoids are typically a benign finding of the anterior segment. Need to be monitored carefully in infants and young children to prevent amblyopia. Presence of a limbal dermoid may also warrant a systemic evaluation to help screen for conditions such as Goldenhar syndrome, along with potential genetic testing. Important to understand various differentials to ensure correct diagnosis. In adults, it is important to monitor limbal dermoids to ensure no significant changes or signs of malignancy occur.

Conclusion

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Questions

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References


