Corneal Pathologies

Dr. Pradeep Bastola
MD, Ophthalmologist
Assistant Professor
15th June, 2011

PERIPHERAL CORNEAL THINNING AND ULCERATION

1. Without systemic disease
   - Dellen
   - Terrien marginal degeneration
   - Mooren ulcer

2. With systemic disease
   - Rheumatoid arthritis
   - Wegener granulomatosis
   - Polyarteritis nodosa

---

Dellen
- Common and unilateral
- Innocuous

**Signs**
- Saucer-like thinning with intact epithelium
- Fluorescein pooling but no staining

**Causes**
- Chemosis, raised limbal lesions
- Abnormal blinking

**Treatment**
- Lubricants and elimination of cause

---

Terrien marginal degeneration
- Uncommon, bilateral but asymmetrical
- Initially asymptomatic

**Signs**
- Fine stromal lipid deposition separated by clear zone
- Mild thinning and vascularization

**Causes**
- Circumferential thinning and increasing astigmatism
- Formation of pseudo-pterygia if longstanding

**Progression**
- Treatment of severe astigmatism - crescent-shaped excision of gutter

---

Mooren ulcer
- Limited form - usually unilateral, affects elderly
- Progressive form - bilateral, affects younger patients

**Signs**
- Chronic and asymptomatic
- Circumferential thinning with intact epithelium ("contact lens cornea")

**Causes**
- Acute and painful
- Circumferential ulceration and infiltration

**Progression**
- Peripheral ulcerative keratitis
- Circumferential and central spread
- End-stage scarring and vascularization

**Treatment**
- Systemic steroids and/or cytotoxic drugs

---

Peripheral corneal involvement in rheumatoid arthritis

**Without inflammation**

**With inflammation**
- Chronic and asymptomatic
- Circumferential thinning with intact epithelium ("contact lens cornea")

**Causes**
- Acute and painful
- Circumferential ulceration and infiltration

**Treatment**
- Systemic steroids and/or cytotoxic drugs
Peripheral corneal involvement in Wegener granulomatosis and polyarteritis nodosa

Circumferential and central ulceration similar to Mooren ulcer

Unlike Mooren ulcer, sclera may also become involved

Treatment - systemic steroids and cyclophosphamide

PERIPHERAL CORNEAL INFLAMMATION

1. Marginal keratitis
2. Rosacea keratitis
3. Phlyctenulosis
4. Acute stromal keratitis

Marginal keratitis

- Hypersensitivity reaction to Staph. exotoxins
- May be associated with Staph. blepharitis
- Unilateral, transient but recurrent

Progression

Subepithelial infiltrate separated by clear zone
Circumferential spread
Bridging vascularization followed by resolution

Treatment - short course of topical steroids

Rosacea keratitis

- Affects 5% of patients with acne rosacea
- Bilateral and chronic

Progression

Peripheral inferior vascularization
Subepithelial infiltration
Thinning and perforation if severe

Treatment - topical steroids and systemic tetracycline or doxycycline

Phlyctenulosis

- Uncommon, unilateral - typically affects children
- Severe photophobia, lacrimation and blepharospasm

Conjunctival phlycten

- Small pinkish-white nodule near limbus
- Usually transient and resolves spontaneously
- Starts astride limbus
- Resolves spontaneously or extends onto cornea

Treatment - topical steroids

CORNEAL INFECTIONS

1. Bacterial keratitis
2. Fungal keratitis
3. Acanthamoeba keratitis
4. Infectious crystalline keratitis
5. Herpes simplex keratitis
   - Epithelial
   - Disciform
6. Herpes zoster keratitis
Bacterial keratitis

**Predisposing factors**
- Contact lens wear
- Chronic ocular surface disease
- Corneal hypoesthesia

**Signs**
- Expanding oval, yellow-white, dense stromal infiltrate
- Stromal suppuration and hypopyon

**Treatment**
- topical ciprofloxacin 0.3% or ofloxacin 0.3%

Fungal keratitis

**Frequently preceded by ocular trauma with organic matter**

**Signs**
- Greyish-white ulcer which may be surrounded by feathery infiltrates
- Slow progression and occasionally hypopyon

**Treatment**
- Topical antifungal agents
- Systemic therapy if severe
- Penetrating keratoplasty if unresponsive

Acanthamoeba keratitis

**- Contact lens wearers at particular risk**
- Symptoms worse than signs

**Signs**
- Small, patchy anterior stromal infiltrates
- Perineural infiltrates (radial keratoneuritis)
- Ulceration, ring abscess/stromal opacification & small, satellite lesions

**Treatment**
- chlorhexidine or polyhexamethylenebiguanide

Infectious crystalline keratitis

**- Very rare, indolent infection (Strep. viridans)**
- Usually associated with long-term topical steroid use
- Particularly following penetrating keratoplasty

**Signs**
- White, branching, anterior stromal crystalline deposits

**Treatment**
- topical antibiotics

Herpes simplex epithelial keratitis

**Signs**
- Dendritic ulcer with terminal bulbs
- Stains with fluorescein

**Treatment**
- Aciclovir 3% ointment x 5 daily
- Trifluorothymidine 1% drops 2-hourly
- Debridement if non-compliant

Herpes simplex disciform keratitis

**Signs**
- Central epithelial and stromal oedema
- Folds in Descemet membrane
- Small keratic precipitates

**Associations**
- Occasionally surrounded by Wessely ring

**Treatment**
- topical steroids with antiviral cover
**Herpes zoster keratitis**

- Develops in about 50% within 2 days of rash
- Small, fine, dendritic or stellar epithelial lesions
- Tapered ends without bulbs
- Resolves within a few days

**Acute stromal keratitis**

- Uncommon, usually unilateral
- Associated with non-necrotizing scleritis

**Nummular keratitis**

- May become chronic

**Treatment**
- Topical steroids, if appropriate

**Acute stromal keratitis**

- Superficial or mid-stromal infiltration
- Opacification and vascularization

**Treatment**
- Topical steroids and systemic NSAIDs

**CORNEAL DEGENERATIONS**

1. **Age-related**
   - Arcus senilis
   - Vogt white limbal girdle
   - Crocodile shagreen
   - Cornea guttata

2. **Lipid keratopathy**
   - Primary
   - Secondary

3. **Band keratopathy**

4. **Spheroidal degeneration**

5. **Salzmann nodular degeneration**

**Arcus senilis**

- Innocuous and extremely common in elderly
- Occasionally associated with hyperlipoproteinaemia

**Vogt white limbal girdle**

- Innocuous and very common in elderly
- Bilateral

- White, crescentic line along nasal and temporal limbus
- Type 1 - separated from limbus by clear zone
- Type 2 - not separated by clear zone

**Crocodile shagreen**

- Uncommon and usually bilateral
- Peripheral border separated from limbus by clear zone
- Clear zone may be thinned (senile furrow)

- Polygonal stromal opacities separated by clear space
- Most frequently involve anterior stroma (anterior crocodile shagreen)
- Occasionally involve posterior stroma (posterior crocodile shagreen)
Cornea guttata
• Common, bilateral and usually innocuous
• Rarely progression to Fuchs dystrophy

![Tiny dark spots on central endothelium. Similar peripheral lesions are Hassell-Henle bodies.]

Causes of Band Keratopathy
1. Ocular (common)
   • Chronic iridocyclitis, particularly in children
   • Associated with phthisis bulbi
   • Silicone oil in anterior chamber
2. Metabolic (rare)
   • Increased serum calcium and phosphorus
   • Hyperuricaemia
   • Chronic renal failure
3. Hereditary (rare)
   • Familial band keratopathy
   • Hereditary ichthyosis
4. Age-related (uncommon)

Spheroidal degeneration
• Rare, typically affects outdoor workers
• Starts with peripheral, interpalpebral, small amber-coloured granules in superficial stroma

![Central spread, coalescence and opacification. Advanced lesions become nodular and elevated.]

Treatment
- Debridement or superficial keratotomy if mild
- Keratoplasty if severe

Salzmann nodular degeneration
• Uncommon, unilateral or bilateral
• Secondary to chronic keratitis

![Discrete superficial stromal opacities and nodules. Base of nodule may be surrounded by iron deposits. Treatment - similar to spheroid degeneration.]

Progression

Central spread, coalescence and opacification
Advanced lesions become nodular and elevated

Treatment
- Debridement or superficial keratotomy if mild
- Keratoplasty if severe
CORNEAL DYSTROPHIES

1. Anterior
   • Cogan microcystic
   • Reis-Bucklers
   • Meesmann
   • Schnyder

2. Stromal
   • Lattice I, II, III
   • Granular I, II, III (Avellino)
   • Macular

3. Posterior
   • Fuchs endothelial
   • Posterior polymorphous

Cogan microcystic dystrophy
- Most common of all dystrophies
- Neither familial nor progressive
- Recurrent corneal erosions in about 10% of cases

Signs of Cogan dystrophy
Four types of lesions - in isolation or combination

<table>
<thead>
<tr>
<th>Dots</th>
<th>Microcysts</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maps</td>
<td>Fingerprints</td>
</tr>
</tbody>
</table>

Reis-Bucklers dystrophy
Inheritance - autosomal dominant
Onset - early childhood with recurrent corneal erosions

Superficial polygonal opacities
Honeycomb appearance

Treatment - keratoplasty if severe

Meesmann dystrophy
Inheritance - autosomal dominant
Onset - first decade with mild visual symptoms

Tiny, epithelial cysts, maximal centrally
Clear in retroillumination
Grey in direct illumination

Treatment - not required

Schnyder dystrophy
Inheritance - autosomal dominant
Onset - second decade with visual impairment

Subepithelial ‘crystalline’ opacities

Treatment - excimer laser keratectomy
Inheritance - autosomal dominant
Onset - late first decade with recurrent corneal erosions

**Lattice dystrophy type I**

**Progression**

- Fine, spidery, branching lines within stroma
- Later general haze may submerge lesions

Treatment - penetrating keratoplasty if severe

**Lattice dystrophy type II**

*(Familial amyloidosis with lattice dystrophy, Meretoja syndrome)*

**Inheritance**

- Autosomal dominant

**Onset**

- Middle age with progressive facial palsy and lattice dystrophy identical to type I

**Systemic features**

- Cranial and peripheral neuropathy
- Skin laxity
- Renal and cardiac failure

**Treatment**

- Penetrating keratoplasty if severe

**Lattice dystrophy type III**

**Inheritance - autosomal dominant**

**Onset - fourth decade**

- Thick, ropey lines and minimal intervening haze
- May be asymmetrical and initially unilateral

Treatment - penetrating keratoplasty if severe

**Granular dystrophy type I**

**Inheritance - autosomal dominant**

**Onset - first decade with recurrent corneal erosions**

**Progression**

- Initial superficial and central crumb-like opacities
- Later deeper and peripheral spread but limbus spared
- Eventual confluence

Treatment - penetrating keratoplasty if severe

**Granular dystrophy type II**

**Inheritance - autosomal dominant**

**Onset - fourth or fifth decade with mild recurrent erosions**

- Superficial, discrete crumb-like opacities

Treatment - penetrating keratoplasty if severe

**Granular dystrophy type III (Avellino)**

**Inheritance - autosomal dominant**

**Onset - late in life; frequently asymptomatic**

- Few, superficial, discrete, ring-shaped lesions
- Increase in density and size with time

Treatment - not required
**Macular dystrophy**
Inheritance - autosomal recessive
Onset - second decade with painless visual loss

<table>
<thead>
<tr>
<th>Progressive</th>
<th>Inheritance</th>
<th>Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial dense, poorly delineated opacities</td>
<td>autosomal recessive</td>
<td>second decade with painless visual loss</td>
</tr>
<tr>
<td>Later generalized opacification</td>
<td>autosomal recessive</td>
<td>second decade with painless visual loss</td>
</tr>
<tr>
<td>Thinning</td>
<td>autosomal recessive</td>
<td>second decade with painless visual loss</td>
</tr>
</tbody>
</table>

Treatment - penetrating keratoplasty

**Fuchs endothelial dystrophy**
Inheritance - occasionally autosomal dominant
Onset - old age

<table>
<thead>
<tr>
<th>Progressive</th>
<th>Inheritance</th>
<th>Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gradual increase in cornea guttata with peripheral spread</td>
<td>autosomal dominant</td>
<td>old age</td>
</tr>
<tr>
<td>Later central stromal oedema</td>
<td>autosomal dominant</td>
<td>old age</td>
</tr>
<tr>
<td>Eventually bullous keratopathy</td>
<td>autosomal dominant</td>
<td>old age</td>
</tr>
</tbody>
</table>

Treatment - penetrating keratoplasty if advanced

**Posterior polymorphous dystrophy**
Inheritance - usually autosomal dominant
Onset - difficult to determine because asymptomatic

- Subtle, vesicular, geographic, or band-like lesions
- Frequently asymmetrical

Treatment - not required

**CORNEAL ECTASIAS**

1. Keratoconus
2. Keratoglobus
3. Pellucid marginal degeneration

**Morphological classification of keratoconus**

<table>
<thead>
<tr>
<th>Nipple cone</th>
<th>Oval cone</th>
<th>Globus cone</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small and steep curvature</td>
<td>Larger and ellipsoidal</td>
<td>Largest</td>
</tr>
</tbody>
</table>

**Signs of keratoconus**
Bilateral in 85% but asymmetrical

- Oil droplet reflex
- Vogt striae
- Prominent corneal nerves
- Bulging of lower lids on downgaze
- Fleischer ring & scarring
- Munson sign
- Acute hyperopic shift
Systemic associations of keratoconus
- Atopic dermatitis
- Down syndrome
- Ehlers-Danlos syndrome
- Marfan syndrome
- Crouzon syndrome
- Osteogenesis imperfecta

Keratoglobus
- Bilateral protrusion and thinning of entire cornea
- Associations - Leber congenital amaurosis and blue sclera

Pellucid marginal degeneration
- Bilateral crescent-shaped inferior corneal thinning
- Onset between 20 and 40 years

CONGENITAL CORNEAL ANOMALIES
1. Microcornea
- Very rare, hereditary, unilateral or bilateral
- Shallow anterior chamber but other dimensions are normal
- Ocular associations - Glaucoma, cataract, cornea plana, leukoma and iris abnormalities
- Associated systemic syndromes - Turner, Ehlers-Danlos, Weill-Marchesani and Waardenburg

2. Megalocornea
- Very rare, hereditary, bilateral
- Corneal diameter 13 mm or more
- Very deep anterior chamber
- High myopia and astigmatism
- Occasionally lens subluxation
- Systemic associations - Marfan, Apert, Ehlers-Danlos and Down syndromes
- Osteogenesis imperfecta
- Renal carcinoma and mental handicap

3. Sclerocornea

4. Cornea plana

5. Keratectasia

Microcornea

Megalocornea
**Sclerocornea**
- Very rare, usually bilateral
- Peripheral opacification and vascularization of cornea
- ‘Scleralization’ makes cornea appear smaller

**Cornea plana**
- Very rare, bilateral severe decrease in corneal curvature
- Hypermetropia and shallow anterior chamber

**Keratectasia**
- Very rare, usually unilateral
- Severe corneal opacification and protruberance
- Probably caused by intrauterine keratitis

**CORNEAL SURGERY**
1. Penetrating keratoplasty
2. Keratoprosthesis
3. Refractive surgery
   - Radial keratotomy
   - Photorefractive keratectomy (PRK)
   - Laser in-situ keratomileusis (LASIK)
   - Non-contact laser thermal keratoplasty

**Penetrating Keratoplasty**
1. Indications
   - Optical (e.g. bullous keratopathy, dystrophies)
   - Tectonic (e.g. severe stromal thinning, descemetocele)
   - Therapeutic (e.g. severe keratitis)
   - Cosmetic
2. Adverse prognostic factors
   - Severe stromal vascularization
   - Absence of corneal sensation
   - Progressive conjunctival inflammation (e.g. pemphigoid)
   - Tear film dysfunction
   - Glaucoma

**Technique of penetrating keratoplasty**
- Excision of donor tissue
- a, b - Excision of host tissue
- c - Fixation of donor tissue
**Signs of late graft rejection**

- Epithelial
  - Intensive topical and periocular steroids
  - Occasionally systemic steroids

- Endothelial
  - Iritis and inflammation at graft-host junction
  - Endothelial precipitates (Khodadoust line)
  - Linear epithelial opacity
  - Subepithelial opacities

**Radial keratotomy**

- Decreases myopia by flattening cornea
- Deep incisions from edge of optical zone to limbus

**Treatment**

- Stable myopia of up to 8D
- Otherwise normal cornea (Accidental perforation)
- Intrastromal epithelial cysts

**Laser in-situ keratomileusis (LASIK)**

<table>
<thead>
<tr>
<th>Technique</th>
<th>Main complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reshaping of cornea by excimer laser ablation of Bowman layer and anterior stroma</td>
<td>Subepithelial haze which usually resolves after 1-4 months</td>
</tr>
</tbody>
</table>

**Indications**

- similar to PRK but corrects higher degrees of myopia

<table>
<thead>
<tr>
<th>Main complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Thin flap of cornea fashioned</td>
</tr>
<tr>
<td>- Bed treated with excimer laser</td>
</tr>
<tr>
<td>- Flap repositioned</td>
</tr>
<tr>
<td>- Wrinkles in flap</td>
</tr>
<tr>
<td>- Cellular interface proliferation</td>
</tr>
</tbody>
</table>

**Non-contact laser thermal kerato-plasty**

- Patients over 40 years with hypermetropia up to 2D
- Following overcorrection of myopia

- Corneal curvature is steepened by application of laser heat to stroma
- Holmium laser spots applied to mid-cornea

**Red Eye**
Introduction

- Relevance
  - Red Eye
    - Frequent presentation to GP
    - Must be able to differentiate between serious vision threatening conditions and simple benign conditions

Basics

- Red Eye
  - Refers to hyperemia of the superficially visible vessels of the conjunctiva, episclera, or the sclera
  - Caused by disorders of these structures themselves, or of adjacent structures like the eyelids, cornea, iris, and ciliary body

Differential diagnosis of red eye

- Conjunctival
  - Blepharoconjunctivitis
  - Bacterial conjunctivitis
  - Viral conjunctivitis
  - Conjunctivitis: allergic, atopic, Tolosa conjunctivitis
  - Toxic conjunctivitis
  - Dry eye
  - Pineal conjunctivitis

- Lid disease
  - Chalazion
  - Stye
  - Abnormal lid function

- Corneal disease
  - Abrasion
  - Ulcer

- Foreign body

- Dacyroadenitis
- Dacryocystitis
- Masquerade syndrome
- Corneal and neural fistula
- Acute angle glaucoma
- Anterior uveitis
- Episcleritis/scleritis
- Subconjunctival hemorrhage
- Factitious

Blepharitis

- Adults > children
- Inflammation of the lid margin
- Frequently associated with styes
- Meibomian gland dysfunction
- Lid hygiene, topical antibiotics, and lubricants are the mainstays of treatment
### Bacterial Conjunctivitis
- Both adults and children
- Tearing, foreign body sensation, burning, stinging and photophobia
- Mucopurulent or purulent discharge
- Lid and conjunctiva maybe edematous
- Streptococcus pneumoniae, Haemophilus influenzae, and staphylococcus aureus and epidermidis
- Conjunctival swab for culture
- Topical broad spectrum antibiotics

### Viral Conjunctivitis
- Acute, watery red eye with soreness, foreign body sensation and photophobia
- Conjunctiva is often intensely hyperemic and there maybe follicles, haemorrhages, inflammatory membranes and a pre-auricular node
- The most common cause is an adenoviral infection
- No specific therapy but cold compresses are helpful

### Allergic Conjunctivitis
- Encompasses a spectrum of clinical condition
- All associated with the hallmark symptom of itching
- There is often a history of rhinitis, asthma and family history of atopy
- Signs may include mildly red eyes, watery discharge, chemosis, papillary hypertrophy and giant papillae
- Treatment consist of cold compresses, antihistamines, nonsteroidals, mast cells stabilizers, topical corticosteroids and cyclosporine

### Chlamydial Conjunctivitis
- Usually occur in sexually active individuals with or without an associated genital infection
- Conjunctivitis usually unilateral with tearing, foreign body sensation, lid crusting, conjunctival discharge and follicles
- There is often non-tender preauricular node
- Treatments requires oral tetracycline or azithromycin

### Dry Eye
- **Symptoms**
  - Burning or foreign body sensation
  - Tearing
  - Usually bilateral
- **Etiology**
  - Idiopathic
  - Collagen vascular diseases
  - Conjunctival scarring
  - Infiltration of the lacrimal gland
  - Vitamin A deficiency
- **Treatment**
  - Artificial tears
Foreign Body

Nasolacrimal Obstruction
- Can lead to Dacryocystitis
  - Pain, redness, and swelling over the innermost aspect of the lower eyelid, tearing, discharge
  - Organisms
    - Staphylococci, streptococci, and diphtheoids
  - Treatment
    - Systemic antibiotics
    - Surgical drainage

Conjunctival tumor

Iritis

Episcleritis

- Episcleritis
  - Can be localized (sectorial) or diffuse redness
  - Often asymptomatic
  - Usually self limited
  - Treatment is topical or systemic NSAIDs

Scleritis

- Pain which maybe severe with tenderness, tearing and photophobia
- Maybe localized, diffuse or associated with nodules
- Can result in scleral necrosis (scleromalacia perforance)
- 30 to 60 % may have an associated systemic diseases
  - RA ...
- May need systemic steroid
Subconjunctival Hemorrhage

- Usually asymptomatic
- Blood underneath the conjunctiva, often in a sector of the eye
- Etiology
  - Valsalva (coughing or straining)
  - Traumatic
  - Hypertension
  - Blinding disorder
  - Idiopathic

Red Eye Treatment Algorithm

- History
  - Trauma
  - Contact lens wearer
  - Severe pain/photophobia
  - Significant vision changes
  - History of prior ocular diseases
- Exam
  - Abnormal pupils
  - Ocular tenderness
  - White corneal opacity
  - Increased intraocular pressure

Is it conjunctivitis?

- History
  - Itching
  - Exposure to person with red eye
  - URTI
  - Past history of conjunctivitis
  - Discharge with morning crusting
  - Exposure to drugs

- Signs
  - Discharge
  - Lid and conjunctival edema
  - Conjunctival redness
  - Preauricular lymph node
  - Facial or eye lid vesicles

Thank you