2) Outer Eyeball Coat, Cornea & Sclera - Dr. Ali Taqi

Gross Anatomy:

The cornea is the clear transparent anterior one sixth of the fibrous tunic of the eyeball. The cornea is set into the opaque sclera like a watch glass, this corneo-scleral junction is called the limbus.

- The cornea has the following features:
  1. It has a smooth brilliant surface.
  2. It is very richly supplied with nerve fibers.
  3. It has no blood vessels.
  4. The cornea is the most powerful refractive medium of the eye.

Microscopic Anatomy:

Epithelium: This is a regular layer of uniform thickness of non-keratinized stratified squamous epithelium.

1. **Bowman’s Membrane:** This is a clear uniform structureless membrane that runs underneath the epithelium throughout the cornea and ends abruptly at the limbus.

2. **Substantia Propria:** It forms about 90% of the entire thickness of the cornea. It is made up of regularly arranged lamellae of collagenous fibers associated with some elastic fibers lying in a mucopolysaccharide matrix.

3. **Descemet’s Membrane:** This is a thin fine transparent and highly elastic layer situated posterior to the substantia propria of the cornea.

4. **Endothelium:** A single layer of flat hexagonal cells arranged along the inner surface of Descemet's membrane.

- **Nerve Supply:** Derived from the naso-ciliary branch of the ophthalmic division of the trigeminal nerve through its long ciliary nerves.

Functions of the cornea:

1. **Optical:** The cornea is the most powerful refractive medium of the eye (42 Diopter).

2. **Protective:** The extreme sensitivity of the cornea is an efficient protective mechanism producing a very quick lid reflex.

Inflammations of the cornea:

Inflammations of the cornea may be divided into two types:

1. **Ulcerative Keratitis:** A corneal ulcer is a condition in which there is destruction of some portion of both the epithelium and the underlying Stroma of the cornea.

2. **Non-ulcerative keratitis:** is a condition in which the Stroma of the cornea is only affected by the inflammatory process while the overlying epithelium remains intact.
Ulcerative keratitis:

Clinically it appears as a saucer-shaped yellowish grey pit, which stains green with fluorescein

Causes of Corneal Ulceration:

1. **Exogenous Infections:** The cornea is exposed throughout life to minor trauma, virulent micro-organisms present in the conjunctival sac may gain access to the corneal tissue.
2. **Endogenous Agents:** The inflammation is typically a cell-mediated immune response to a foreign antigen and often occurs in the periphery of the cornea, e.g. phlyctenular ulcer.
3. **Chemical Agents:** Acids, alkali, tincture of iodine or lime produce corneal ulceration by destroying the epithelium and the superficial layers of the Stroma.
4. **Exposure:** Incomplete covering of the cornea leads to constant ulceration and dryness of the cornea.
5. **Loss of corneal sensation:** Corneal anesthesia, caused by trigeminal nerve paralysis, leads to trophic changes in the corneal epithelium resulting in degeneration and ulceration. This condition is called neuropralytic keratitis.
6. **Vitamin A Deficiency:** This leads to degenerative changes in the corneal epithelium leading to ulceration, it also induces lack of resistance of the cornea to micro-organisms of low virulence.

**Bacterial ulcerative keratitis:**

**Hypopyon ulcer**

- Also called pneumococcal ulcer, is a disc-shaped ulcer of destructive nature near the centre of the cornea associated with a diffuse keratitis, a violent iridocyclitis and a hypopyon.

**Aetiology:**

- Any defect in the basic defense mechanism of the cornea, from trauma can predispose to corneal ulceration. Hence, hypopyon ulcer often starts as a minor abrasion to the cornea, e.g. by a scratch with a finger nail or by a foreign body.
- Virulent conjunctival pathogens can then lodge in the exposed corneal Stroma and cause necrosis of the corneal parenchyma.

**Symptoms:**

- Pain: it is of a neuralgic nature and is often referred to the eyebrow region.
- Photophobia.
- Blepharospasm.
- Excessive reflex Lacrimation.
- Defective vision.

**Signs:**

- Circumcorneal vascular injection.
- Haziness and dullness of the corneal surface.
- Central disc-shaped ulcer.
- Vascularization of the cornea.
- Mild to severe iridocyclitis.
- Secondary rise of ocular tension.
Complications of the Hypopyon Ulcer:

- **Complication before perforation:**
  1. Anterior uveitis.
  2. Secondary glaucoma.
  3. Corneal opacity.

- **Complication after perforation:**
  1. Prolapse of the iris.
  2. Anterior Synechiae.
  3. Leucoma Adherent.
  4. Anterior Staphyloma.
  5. Anterior Polar Cataract.
  7. Intra-ocular Hemorrhage.
  8. Endophthalmitis and Panophthalmitis.

Treatment of Uncomplicated Corneal Ulcer:

- Antibiotic to control infection.
- Atropine to Relieve Uveal Irritation.
- Application of heat to increase blood flow.
- Wash out of conjunctival discharge.
- Protection of the eye.
- Promotion of re-epithelialization.

Viral Ulcerative Keratitis:

Herpes Simplex Keratitis:

- Is primarily a superficial epithelial infection of the cornea, usually develops after common colds or upper respiratory tract infections, in association with herpetic vesicles in the lips and face.
- The disease has a definite tendency to recurrences. Primary ocular involvement may present as an acute follicular conjunctivitis or kerato-conjunctivitis with nonsupurative preauricular Lymphadenopathy and often with notable vesiculating periorcular skin involvement.

Clinical picture:

- Symptoms:
  - Photophobia, reflex lacrimation, pain blurring of vision, foreign body sensation & corneal hypoesthesia.
- Signs of Primary Ocular Infection:
  - Vesicles involving the lids and periorbital area.
  - Acute follicular conjunctivitis.
  - Fine or coarse epithelial punctate keratitis which may progress to dendritic figure.
- Signs of Recurrent herpetic Keratitis:
  - Dendritic Epithelial Ulceration.
  - Geographical Herpetic Ulceration.
  - Stromal Herpetic Keratitis without or with Epithelium Ulceration.
  - Herpetic iridocyclitis.

Treatment of Herpes Simplex Keratitis:

- Medical lines of Treatment:
  - Antiviral agents are usually the first line of treatment, acyclovir eye ointment 5 times/day for up to 14 days.
  - The usual routine treatment of corneal ulcers, namely, atropine drops, antibiotic against secondary bacterial infection.
- Surgical lines of treatment:
  1. Debridement: removal of virus-containing cells is indicated in recurrent dendritic ulcers.
  2. Lamellar Keratoplasty
  3. Penetrating Keratoplasty.
Herpes Zoster ophthalmicus:

1. Is caused by a virus identical with that causing chicken-pox,
2. The chief focus of infection is in the Gasserian ganglion whence the virus travels down one or more of the branches of the ophthalmic division of the fifth nerve.
3. Ocular complications usually arise when the naso-ciliary branch of the trigeminal nerve is involved.
4. The appearance of vesicles on the tip of the nose often precedes corneal affection (Hutchinson’s rule).
   - Herpes Zoster affects elderly and often it is rare under 40 years of age.

Keratitis in rheumatoid arthritis

Presentations:

1. Peripheral corneal thinning.
2. Keratoconjunctivitis sicca.
3. Scleritis.

Rheumatoid arthritis is the most common collagen vascular disorder to affect the peripheral cornea.

Peripheral corneal thinning (contact lens cornea): is characterized by gradual resorption of peripheral corneal tissue leaving the epithelium intact.

Because the central part of the cornea remains normal, the appearance resembles a contact lens placed on the eye.

Treatment

1. Topical steroids may be useful in acute stromal keratitis, but they should be avoided in peripheral corneal guttering and keratolysis for fear of inducing further corneal thinning and possibly perforation.
2. Systemic therapy with steroids and/or cytotoxic drugs is required for active scleritis.
3. Conjunctival excision may halt the progression of corneal furrowing in refractory cases.
4. Keratoplasty may be required either as an emergency measure to prevent perforation or, electively, to restore visual acuity.

Keratoconus (conical cornea)

- Is a fairly common, progressive disorder in which the cornea assumes an irregular conical shape.
- The hallmark of Keratoconus is central or paracentral stromal thinning.
- Both eyes are affected in about 85% of cases, although the severity of involvement may be markedly asymmetrical.
- Keratoconus occurs with increased frequency in the following disorders…….

Systemic disorders include:
- Down's syndrome
- Turner's syndrome
- Ehlers-Danlos syndrome
- Marfan's syndrome
- Osteogenesis imperfecta

Ocular associations include:
- Vernal disease,
- Retinitis pigmentosa,
- Blue sclera,
- Aniridia and ectopia lentis.
- The wearing of hard contact lenses and constant eye rubbing have also been proposed as possible predisposing factors.
CLINICAL FEATURES

- Onset is typically between the ages of 10 and 20 years, with impaired vision in one eye caused by progressive astigmatism and myopia.
- Early signs, which are easy to miss, can be detected by the following methods of examination:
  1. Retinoscope shows an irregular 'scissor' reflex.
  2. Keratometry initially shows irregular astigmatism where the principal meridians are no longer 90 D apart and.
  3. Photokeratoscopy or Placido's disc shows irregularity of the reflected ring contours.
  4. Slit lamp biomicroscopy shows very fine, deep, stromal, oblique striae (Vogt's lines) which disappear with external pressure on the globe.

Late signs consist of the following:

1. Progressive central or paracentral corneal thinning, of as much as one-third of the corneal thickness. This is associated with poor visual acuity resulting from marked irregular astigmatism with steep Keratometry (K) readings.
2. Bulging of the lower lid when the patient looks down (Munson's sign ;).
3. Epithelial iron deposits (Fleischer's ring) may surround the base of the cone.
4. Central and paracentral corneal scarring in severe cases.
5. Acute hydrops results from ruptures in Descemet's membrane and acute leakage of fluid into the corneal stroma and epithelium although the break usually heals within 6-10 weeks and the corneal oedema clears, a variable amount of stromal scarring may develop.

MANAGEMENT

1. Spectacle correction in very early cases can correct regular astigmatism and very low amounts of irregular astigmatism.
2. Contact lenses provide a regular refracting surface over the cone.
3. Epikeratoplasty is an effective procedure in patients intolerant to contact lenses without significant central corneal scarring.
4. Penetrating Keratoplasty is indicated in patients with advanced progressive disease, especially with significant corneal scarring. The visual results are excellent with a low risk of allograft rejection.

Contact lenses

The following are the main indications for contact lens wear:

1. Irregular astigmatism associated with Keratoconus can be corrected with a hard contact lens long after spectacles have failed and long before corneal grafting becomes necessary.
2. Superficial corneal irregularities: a contact lens can replace a superficial irregular corneal surface by a smoother and optically more perfect surface.
3. Persistent epithelial defects can be healed more quickly by protecting the regenerating corneal epithelium from the constant rubbing action of the lids.
4. Recurrent corneal erosions, particularly if associated with a corneal dystrophy, may require long-term lens wear.
5. Descemetoceles can be covered with lens a contact lens as a temporary measure to allow time for natural healing to occur.
6. Protection of normal corneal epithelium in eyes with trichiasis or threatened exposure Keratopathy is another indication.
Complications of contact lens wear

**CONJUNCTIVAL COMPLICATION.**

- Giant papillary conjunctivitis (GPC): is fairly common. Although any contact lens can cause the condition, soft lenses are most frequently implicated.
- Presentation:
  - May be months or years after beginning lens wear with ocular itching after lens removal, increased mucus production in the morning, photophobia and decreased lens tolerance.
  - Blurred vision may also occur either from deposits on the lens or when the lens is pulled towards the upper fornix by the upper lid.

**CORNEAL COMPLICATIONS**

The following are the main corneal complications of contact lens wear:

1. Epithelial edema secondary to hypoxia which is usually reversible.
2. Corneal Vascularization develops in some eyes as a response to lens-induced hypoxia.
3. Sterile corneal infiltrates are usually small peripheral opacities.
4. Microbial keratitis with bacteria, acanthamoebae and fungi is the most serious, but fortunately a fairly uncommon, complication.

**Treatment:**

1. Lens hygiene should be optimized and the patient may need to be re-instructed in the use of care solutions.
2. Wearing time of the lenses should be minimized.
3. Attention should be paid to the fit of the lenses and the material from which they are made.
4. Topical treatment with sodium cromoglycate is often effective but long-term steroids should be avoided because the hazards of steroid therapy may be greater than the condition being treated.

**Corneal Degenerations and Dystrophies:**

**Arcus Senilis:**

- It is an age-related hazy grey line of fatty degeneration which encircles the cornea just inside the limbus, no treatment is necessary, however, its early or excessive appearance may indicate hypercholesterolemia.

**Episcleritis**

- Clinical features
  - Episcleritis is a common, benign, self-limiting and frequently recurrent disorder which typically affects young adults. It is seldom associated with a systemic disorder and never progresses to a true scleritis.
  - The two clinical types of Episcleritis are simple and nodular.
- Treatment
  - Simple Episcleritis usually resolves spontaneously within 1-2 weeks although the nodular type may take longer. Mild cases may need no specific therapy but if discomfort is annoying, topical steroids and/or topical non-steroidal anti-inflammatory drugs (NSAIDs) may be helpful.
Scleritis

Scleritis is a granulomatous inflammation of the sclera coat of the eye. It is much less common than Episcleritis. The condition covers a spectrum of ocular disease which

*Classification*

The classification is based on the primary anatomical site of the inflammation and the following associated changes in the sclera vasculature.

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<tr>
<th><strong>Anterior scleritis</strong></th>
<th><strong>Posterior scleritis</strong></th>
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<td>1. Non-necrotizing</td>
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<td>2. Necrotizing</td>
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<td>a. Rheumatoid arthritis</td>
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<td>• without inflammation</td>
<td>b. Connective tissue vascular disorders</td>
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<td>c. Miscellaneous conditions include: relapsing polychondritis, herpes zoster &amp; surgically induced scleritis.</td>
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