Peripheral Corneal Melts

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Peripheral Corneal Melt
- Destructive inflammatory corneal disease
- Progressive juxtalimbal corneal stroma thinning
- Multiple pathologies
- Mechanism poorly understood

Complications
- Systemic implications
- Potentially serious eye complications include
  - corneal perforation
  - severe corneal scarring with thinning
  - vascularisation

Perilimbal Arcades
- Central cornea derives oxygen from tear film and aqueous
- Peripheral cornea derives additional oxygen/nutrients from the perilimbal capillary arcades
- Perilimbal vascular and lymphatic arcades primarily act as a reservoir for immunocompetent cells

Perilimbal Arcades
Deposition of immune complexes in the terminal ends of limbal vessels
- Increases immunologic activity
- Vascular occlusion
- Subsequent leakage of inflammatory cells, along with collagenases and proteases
**Corneal Stromal Melt**

- **Infectious**
  - Bacterial, Viral, Amoeba, Fungal
- **Traumatic**
  - Chemical, Thermal, Radiation
- **Immunological**
  - Cytokine, Immunological, Exposure, Inflammation
- **Local**
  - Mooren’s ulcer, Allograft rejection, Geographic
- **Vascular**
  - Neurological, Neurotrophic keratitis
- **Systemic**
  - Autoimmune, RA, IBD, Collagen vascular disease, ANCA
  - Dermatological, Rosacea, OCP, SJS
  - Malignancy, Infection

**Ocular predisposition to melts**

- **Systemic associations**
  - Autoimmune, RA, PAN, Collagen vascular disease, ANCA
  - IBD
  - Dermatological, Rosacea, OCP, SJS
  - Malignancy

**Corneal Scrape - Usual Suspects**

- **Bacteria**
  - Staphylococcus, Streptococcus, Gonococcus, Moraxella, Haemophilus, Pseudomonas
- **Viral**
  - HSV, HZO
- **Fungal**
  - Acanthamoeba

**Always Exclude Infection**

- **Traumatic**
  - Chemical, thermal, radiation
  - Remove ongoing source of inflammation
  - Support epithelialization

**Abnormalities of Eyelids or Eyelashes**
Neurotrophic Keratitis
- HSV/HZO
- Topical anesthetic abuse
- Chemical and thermal burns
- Contact lens abuse
- Topical drug toxicity
- Irradiation to eye or adnexa
- Corneal surgery
- Non-ocular causes include:
  - Trauma
  - Stroke
  - Aneurysm
  - Diabetes
  - Intracranial masses

Terrien’s Marginal Degeneration
- Slowly progressive non-inflammatory, peripheral corneal thinning
- Unilateral or asymmetrically bilateral
- Associated with corneal neovascularization, opacification and lipid deposition

Terrien’s Marginal Degeneration
- Inflammation typically absent
- Epithelium intact with fine vascularisation
- Characteristic demarcation of peripheral thinning from central cornea - grey lipid line
- Most patients can be managed conservatively

Systemic Peripheral Ulcerative Keratitis
- Rare destructive inflammatory corneal disease
- 3 per million per year
- May be associated with numerous ocular and systemic infectious and noninfectious conditions
- Final common pathway is peripheral corneal thinning

Peripheral Ulcerative Keratitis
- Ulceration of the peripheral cornea in the presence of an associated epithelial defect, with evidence of inflammatory infiltrates in the corneal stroma
- Absence of infection
- Crescent-shaped ulcer, with progression centrally and circumferentially.
Presentation

- Patients typically present with pain though may be absent
- Bilateral or unilateral
- Other associated symptoms include
  - Excessive lacrimation
  - Photophobia
  - Change in vision due to astigmatism or corneal opacity

Examination

- Perilimbal corneal opacity due to stromal cellular infiltrates
- Crescent-shaped corneal ulcers develop with breakdown of the overlying epithelium.
- Varying degrees of vascularization and corneal thinning
- May progress to perforation
- Adjacent conjunctival, episcleral, and scleral inflammation
- 36% have an associated scleritis

Complications

- 21% to 25% worse than 6/60
- 50% require emergency corneal surgery
- 50% recurrence
- Up to 10% may require enucleation
- Overall mortality may approach 30%

Systemic Associations

- Rheumatoid arthritis (32 – 42%)
- Polyarteritis nodosa
- Inflammatory bowel disease
- Collagen vascular diseases
  - Systemic Lupus erythematosus (SLE)
  - Relapsing polychondritis
  - Progressive systemic fibrosis,
- ANCA vasculitides
  - Granulomatosis with polyangiitis (GPA; formerly Wegener’s)
  - Churg–Strauss syndrome
  - Microscopic polyangiitis
- Systemic infectious conditions
  - Syphillis
  - Hepatitis C

Work up

- Detailed personal and family history
- Specific attention given to collagen ascular and other autoimmune diseases
- Complete ocular and systemic examination
Investigations

As directed by the systemic review

- Corneal scrape
- FBC
- ESR
- CRP
- U&Es
- Vasculitis screen
  - RF, ANA, ANCA, dsDNA, anti-CCP
- Cryoglobulins
- VDRL
- Hepatitis C
- Urinalysis with microscopic analysis

Management

- Based on the severity of findings within the cornea and the extent of extracocular disease
- Treatments initiated for systemic autoimmune disease have beneficial effects on ocular manifestations
- Treatment of the systemic disease must be taken into consideration
- Outcome influenced by the accompanying disease, and timely diagnosis and treatment
- Requires a multidisciplinary approach

Local Treatment

- Ensure adequate tear film
  - Preservative free lubricants
- Topical antibiotics to prevent secondary infection
  - Chloramphenicol minims qid
- Cycloplegic for pain and anterior chamber activity

Topical Corticosteroids

Topical corticosteroids inhibit collagen synthesis and thereby increase the risk of perforation.

Systemic Treatment

- Systemic corticosteroids are the traditional first-line therapy
- In isolation, are often unable to inhibit disease progression or overcome the autoimmune disease.
- The usual starting dose is 1 mg/kg/day (maximum 60 mg/day), followed by a tapering schedule based on clinical response.
- Pulsed methylprednisolone 1 g/day for 3 consecutive days, followed by oral therapy, might be initiated in patients with imminent danger of vision loss
Systemic Treatment

- No universal agreement about which immunosuppressant or modulator should be used for specific cases.
- Immunosuppressives available for use in these cases include:
  - Anti-metabolites, e.g., Methotrexate, azathioprine, mycophenolate mofetil, and leflunomide
  - Alkylating agents, e.g., cyclophosphamide and chlorambucil,
  - T cell inhibitors, e.g., Ciclosporin, Tacrolimus
  - Biologic agents, e.g., Infliximab, etanercept, rituximab

Systemic Associations

- Patients with collagen vascular disease related PUK often require aggressive systemic treatment.
- Link between systemic immunosuppression and significantly reduced mortality PUK patients.

Surgical Treatment

- Tectonic procedures may be required to maintain the integrity of the globe.
- Options include:
  - Tissue adhesive
  - Bandage contact lens
  - Lamellar graft
  - Tectonic corneal grafting
  - Amniotic membrane transplantation.

Tissue Adhesive

- Consideration in patients with impending perforation and perforations <2.0 mm.
- Followed by application of a bandage contact lens.

Amniotic Membrane Graft

- Amniotic membrane can be used as a patch or graft to reduce inflammation and to promote re-epithelization.

Corneal Transplantation

- Corneal perforations may require a patch graft.
- High risk of graft failure despite systemic immunosuppression.
Resection of Perilimbal Conjunctiva

- Resection of the perilimbal conjunctiva associated with PUK
  - Removes immune complexes
  - Decreases production of collagenases and proteinases
  - Consequently promotes resolution of inflammation
- Controversial, as it is thought that PUK may recur once the conjunctiva grows back to the limbus

Mooren’s Ulcer

- Moores ulcer is a form of PUK develops in the absence of any associated/causative systemic disease
- It is a diagnosis of exclusion
- It is not associated with scleritis
- Occurs at any age, vast majority of patients are ~ 40 years

Mooren’s Ulcer

- Begins in the peripheral cornea, spreads circumferentially and centrally.
- The main difference from PUK is the severity of pain, which is more intolerable in Mooren’s ulcer.
- It may involve one or both eyes.
- The central border exhibits an overhanging edge
- The sclera is rarely involved
- No perilimbal clear zone

Conclusions

- Peripheral corneal melt includes a group of corneal infectious and inflammatory diseases
- Inflammatory causes are usually associated with life-threatening autoimmune collagen vascular diseases
- PUK might be the initial sign of a systemic disease
- Treatment involves controlling both the ocular inflammation and the underlying systemic vasculitic disease
- Initiation of appropriate immunosuppressive therapy can be life-saving

Thank you