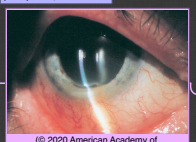
Peripheral Ulcerative Keratitis (PUK)

Case Description
40-year-old woman with unilateral eye pain, photophobia, and redness

Image Description

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peripheral corneal thinning and vascularization

Data acquisition

History
- medical history
  - autoimmune (connective tissue) disease
    - PUK
    - joint pain
    - rheumatoid arthritis
    - lupus
    - Wegener's
    - polyarteritis nodosa
    - inflammatory bowel disease
- ocular history
  - onset & course of symptoms
  - eye pain
    - PUK
    - Mooren's ulcer
  - rules out
    - Terrien marginal degeneration
    - pellucid marginal degeneration
    - furrow degeneration
  - contact lens wear
  - herpetic eye disease
  - history of ocular trauma/ surgery/parasitic diseases
    - Mooren ulcer

Physical Exam
- facial/lid vesicles/scar
- lagophthalmos
- rosacea/blepharitis
- corneal sensation
- herpetic corneal scar
- corneal epithelial defect
- corneal thinning/ perforation
- corneal infiltrate/scar
- other signs of eye inflammation
  - anterior scleritis
  - anterior uveitis
  - hypopyon
  - vitritis
  - retinochoroiditis
  - PUK
- posterior scleritis
  - nerve fiber layer infarcts
  - exudative RD
  - choroidal folds

Additional Testing
- CBC, differential
- ANA
- ANCA
- syphilis serology
- ACE, lysozyme
- PPD
- CXR
- UA
- corneal smear/culture if infection

Assessment
PUK

Treatment

Medical
- q2h
  - antibiotic ointment
- cycloplegia
- topical steroids usually not used in presence of significant corneal thinning ⚠
- systemic steroid
  - exclude TB, syphilis
- steroid-sparing agents
  - to minimize steroid side effects
  - immunosuppressants
    - MTX
    - mycophenolate
    - azathioprine
    - cyclophosphamide
    - infliximab
- metalloproteinase inhibition
  - oral doxycycline
- collagen synthesis promoter
  - ascorbic acid
- if corneal infiltrates
  - culture
  - empiric broad-spectrum topical antibiotics

Surgical
- conjunctival excision/ recession
- for (impending) perforation
  - cyanoacrylate glue
  - corneal transplant

Referrals/ Consultations
- internal medicine consult
  - systemic work-up for autoimmune diseases associated with PUK
  - PUK may be the first sign of the underlying systemic disease!

Patient Education

General
- Dos
  - eye protection
    - glasses
  - eye shield at night
- Complications
  - risk of eye perforation
  - risk of superinfection
- Follow-up
  - examine daily for severe disease
    - continue treatment until epithelium heals
      - taper treatment

Differential Diagnosis

systemic immune-mediated/ rheumatic diseases
- rheumatoid arthritis
- Wegener granulomatosis
- systemic lupus erythematosus
- polyarteritis nodosa
- ulcerative colitis
- relapsing polychondritis
- rosacea

PUK
- usually unilateral
- 1 sector of cornea
- within 2 mm of limbus
- absent epithelium
- pain
- underlying stromal thinning
  - keratolysis
  - +- significant cellular infiltrate in the corneal stroma
- vaso-occlusion of adjacent limbal vessels
- adjacent conjunctiva can be minimally or severely inflamed
- sclera is often involved

Mooren ulcer
- clinical presentation
  - uni- or bilateral
  - chronic progressive
  - symptoms
    - pain
      - can be intense
    - tearing
    - photophobia
  - precipitating factors
    - trauma
    - surgery
    - exposure to parasitic infection
      - incidence of Mooren ulcer is particularly high in areas where parasitic infections are endemic
    - hepatitis C infection
  - ulceration of corneal stroma and epithelium
    - corneal periphery
      - starts in interpalpebral fissure
      - spreads circumferentially
        - similar to Terrien
      - then centripetally
        - leading undermined edge
        - slower ulceration toward sclera

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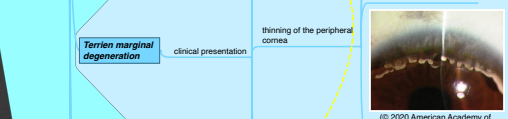
- in contrast to PUK
- eye is inflamed
  - extensive vascularization and fibrosis
- perforation
  - minor trauma
  - secondary infection

Terrien marginal degeneration
- clinical presentation
  - unilateral or asymmetrically bilateral
  - second or third decade of life
  - painless
  - slowly progressive
  - thinning of the peripheral cornea
    - begins superiorly, spreads circumferentially
    - steep central wall
    - gradually sloping peripheral wall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- in contrast to PUK & Mooren ulcer
- in contrast to PUK & Mooren ulcer
  - epithelium remains intact
  - fine vascular pannus
  - line of lipid deposits at the leading edge of the pannus
  - spontaneous perforation is rare
    - can occur with minor trauma
  - against-the-rule astigmatism

staphylococcal marginal keratitis
- erythema/telangiectasia of lid margin
- inferior cornea

bacterial or herpetic keratitis

exposure keratopathy

neurotrophic keratopathy

severe dry eye

dellen

pellucid marginal degeneration
painless, bilateral asymmetric thinning of the inferior peripheral cornea

furrow degeneration
clear zone between senile arcus and limbus

Ophthalmology Mind Maps/Arman Mashayekhi, MD