8.7.2. Immune-mediated diseases (II): Conjunctiva (I)

atopic keratoconjunctivitis

- epidemiology
  - 1/3 of patients with atopic dermatitis
- pathogenesis
  - primarily a type IV reaction
  - mast-cell therapy may not be effective!
  - type I immediate hypersensitivity responses
  - depressed systemic cell-mediated immunity
  - susceptible to
    - herpes simplex virus keratitis
    - colonization of the eyelids with staphylococcus aureus
- clinical presentation
  - similar to VKC with the following differences
    - have disease year-round (minimal seasonal exacerbation)
    - patients are older
    - papillae are small or medium-sized (than giant)
    - papillae occur in the upper and lower palpebral conjunctiva
    - milky conjunctival edema + variable subepithelial fibrosis
    - Figure 7-6 (© 2020 American Academy of Ophthalmology)
    - extensive corneal vascularization and opacification (limbal stem cell dysfunction)
    - Figure 7-7 (© 2020 American Academy of Ophthalmology)
    - eosinophils in conjunctival cytology specimens are less numerous and are less often degranulated
    - conjunctival scarring; occasional symblepharon formation
    - characteristic posterior subcapsular and/or anterior subcapsular lens opacities
    - corneal findings
      - punctate erosions
      - persistent epithelial defects
      - increased incidence of ectatic corneal diseases such as keratoconus and pellucid marginal degeneration
      - increased incidence of staphylococcal and herpes simplex infections
- management
  - allergen avoidance
  - cold compress
  - monitor for complications of infectious diseases
  - pharmacotherapeutic agents similar to those used in the treatment of VKC
  - systemic immune suppression
    - chronic ocular surface inflammation unresponsive to topical treatment
    - severe cases
      - discomfort
      - progressive cicatrization
      - peripheral ulcerative keratopathy
    - oral cyclosporine
    - in coordination with an internist or rheumatologist
  - dermatitis
    - topical tacrolimus

vernal keratoconjunctivitis (VKC)

- epidemiology
  - seasonally recurring
  - year-round in tropical climates
  - male children
  - personal or family history of atopy
- pathogenesis
  - types I and IV hypersensitivity reactions
  - conjunctival inflammatory infiltrate
    - eosinophils, lymphocytes, plasma cells, and monocytes
- clinical presentation
  - symptoms
    - bilateral
    - itching
    - blepharospasm
    - photophobia
    - blurred vision
    - copious mucoid discharge
  - 2 forms
    - palpebral VKC
      - diffuse papillary hypertrophy
      - more prominently on the upper region
      - upper tarsus
      - giant papillae resembling cobblestones
      - Figure 7-3 (© 2020 American Academy of Ophthalmology)
      - bulbar conjunctival hyperemia and chemosis
    - limbal VKC
      - african or asian descent
      - hotter climates
      - alone or in association with palpebral vkc
      - thick gelatinous limbus
      - Figure 7-4 (© 2020 American Academy of Ophthalmology)
      - scattered opalescent mounds
      - vascular injection
      - Horner-Trantas dots
        - whitish macroaggregates of degenerated eosinophils and epithelial cells
    - corneal changes
      - punctate epithelial erosions
        - superior and central cornea
      - pannus
        - superior cornea
        - occasionally 360°
      - noninfectious epithelial ulcers
        - superior or central cornea
        - oval or shieldlike
        - underlying stromal opacification
        - Figure 7-5 (© 2020 American Academy of Ophthalmology)
      - keratoconus
      - stem cell deficiency
- management
  - mild cases
    - topical antihistamines
    - climatotherapy
      - home air-conditioning
      - relocation to a cooler environment
  - mild to moderate disease
    - topical mast-cell stabilizers
      - start at least 2 weeks before symptoms usually begin
  - severe cases
    - topical corticosteroids
      - for exacerbations with moderate to severe discomfort and/or decreased vision
      - intermittent (pulse) therapy
        - every 2 hours for 5–7 days and then rapidly tapered
        - difluprednate ophthalmic emulsion (durezol)
        - soluble corticosteroids
        - dexamethasone phosphate
    - supratarsal injection of corticosteroid
      - evert the upper eyelid
      - dexamethasone phosphate (4 mg/ml)
      - triamcinolone acetonide (40 mg/ml)
      - 0.5–1.0 ml
    - topical immunomodulatory agents
      - cyclosporine
        - 2–4 times daily
        - punctate epithelial keratopathy
        - ocular surface irritation
        - minimal systemic absorption
      - tacrolimus
        - twice daily
  - very severe cases
    - systemic anti-inflammatory therapy

hay fever conjunctivitis and perennial allergic conjunctivitis

- pathogenesis
  - IgE-mediated immediate hypersensitivity reactions
  - vasodilation
  - edema
  - release histamine/inflammatory mediators
  - degranulation
  - conjunctival mast cells
  - airborne allergen
  - eosinophils
  - recruitment of other inflammatory cells
- clinical presentation
  - itching
  - eyelid swelling
  - conjunctival hyperemia
  - symptoms develop rapidly (within minutes) after exposure
  - chemosis
  - mucoid discharge
  - short-lived and episodic
  - allergic rhinitis
  - asthma
  - suffer from other atopic conditions
- diagnosis
  - clinical diagnosis
  - eosinophils
  - conjunctival scrapings
  - challenge testing
- management
  - avoid allergen exposure
  - thorough cleaning (or changing) of
    - unclean or old carpets
    - linens
    - bedding
    - animal dander, house dust mites
  - supportive
    - glasses or goggles
    - cold compresses
    - dilute and flush away allergens
    - artificial tears
  - topical
    - topical antihistamines
      - require continued use over 7 or more days
      - for treating seasonal allergic conjunctivitis
    - topical mast-cell stabilizers
      - ineffective in the acute phase of hay fever conjunctivitis
    - topical nsaids
      - refills should be limited
      - follow-up appointments
      - reports of corneal perforations
    - topical corticosteroids
      - use with caution, except in very severe cases
      - use >5–7 days may predispose to compensatory chronic vascular dilation
    - topical vasoconstrictors
      - provide symptom relief in some patients
    - topical cyclosporine
      - associated dermatitis
    - topical tacrolimus
  - systemic
    - may be associated with increased dry eye
    - systemic antihistamines
  - hyposensitization injections (immunotherapy)

ligneous conjunctivitis

- rare
- pathogenesis
  - composed of
    - fibrin
    - epithelial cells
    - mixed inflammatory cells
    - fibrin-bound tissue plasminogen activator (tPA)
    - matrix metalloproteinase-9
  - deficiency in type I plasminogen
    - severe hypoplasminogenemia (>12% of patients)
      - hypofibrinolysis
    - plasminogen gene (PLG) mutation
    - band 6q26
- clinical presentation
  - all ages
  - bilateral
  - symptoms
    - ocular irritation
    - foreign-body sensation
  - firm ("woody") yellowish, platelike pseudomembranes/masses
    - overlie one or more of the palpebral surfaces
    - Figure 7-8 (© 2020 American Academy of Ophthalmology)
- management
  - cultures
    - to exclude a bacterial pseudomembranous/membranous conjunctivitis
  - surgical excision +- adjunctive cryotherapy
    - recurrences are frequent
  - purified plasminogen
  - fresh frozen plasma
  - heparin
  - corticosteroids
  - azathioprine
  - amniotic membrane
  - +- spontaneous resolution
    - after several months - few years

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