9.7.16. Infectious Ocular Inflammatory Diseases (XVI): Bacterial Uveitis (I): Ocular Syphilis (I)

Epidemiology

- Treponema pallidum — spirochete
- transmission
  - sexual contact — most common
  - transplacental — after the tenth week of pregnancy
- <2% of all uveitis cases
- incidence in US
  - all-time low in 2000
  - incidence rate at any disease stage is rising — especially among — homosexual men — African Americans — have 5x greater risk than non-Hispanic whites
  - sharp declines in both primary and secondary syphilis among women over the past decade — rate of congenital syphilis — 2000: 14.3/100,000 live births; 2010: 8.7/100,000 live births
- one of the great masqueraders of medicine
- can be cured with appropriate antimicrobial therapy, even in patients with HIV/AIDS
- references

Acquired syphilis

- stages
  - primary syphilis
    - incubation period = 3 weeks
    - painless, solitary lesion at site of inoculation — chancre
    - spontaneous resolution within 12 weeks
    - no neurologic findings ✖ ← CNS seeding
  - secondary syphilis
    - 6–8 weeks later
    - lymphadenopathy
    - prominent on the palms and soles — generalized maculopapular rash
    - 10% — uveitis
  - latent period
    - from 1 year (early latency) to decades (late latency)
  - tertiary syphilis
    - 1/3 of untreated patients
    - types
      - benign tertiary syphilis — skin and mucous membranes; gumma; choroid and iris
      - cardiovascular syphilis
      - neurosyphilis — ocular syphilis is a variant of neurosyphilis
    - ≤5% of patients whose disease has progressed to tertiary syphilis
    - uveitis
    - can occur at any stage of infection, including primary disease

Congenital syphilis

- risk of transmission to fetus — primary or secondary syphilis > latent syphilis — the longer the mother had syphilis, the less likely is the transmission
- systemic findings
  - early congenital syphilis (≤2 years)
    - hepatosplenomegaly
    - changes of the long bones on radiographic examination
    - abdominal distention
    - desquamative skin rash
    - low birth weight
    - pneumonia
    - severe anemia
  - late congenital syphilis (>3 years)
    - Hutchinson teeth
    - mulberry molars
    - abnormal facies
    - cranial nerve VIII deafness
    - saber shins
    - perforations of the hard palate
    - cutaneous lesions such as rhagades
    - neurosyphilis
    - cardiovascular complications — unusual
- ocular findings
  - present at birth or decades later
  - congenital cataract
  - the most common inflammatory sign of untreated late congenital syphilis
    - 50%
    - most commonly in girls

Figure 7-40 (© 2020 American Academy of Ophthalmology)

  - stromal interstitial keratitis
    - an allergic response to T pallidum in the cornea
    - symptoms — intense pain; photophobia
    - blood vessels invade the cornea
    - late stages — deep "ghost" (nonperfused) stromal vessels; corneal opacities — left untreated corneal inflammation may regress but leave the cornea diffusely opaque
    - anterior uveitis
    - ← glaucoma
    - Hutchinson triad — interstitial keratitis; cranial nerve VIII deafness; Hutchinson teeth
  - uveitis
    - anterior uveitis — accompanies interstitial keratitis; difficult to observe
    - posterior uveitis
      - multifocal chorioretinitis
        - bilateral salt-and-pepper fundus — affects the peripheral retina, posterior pole, or a single quadrant; not progressive; may have normal vision
        - bilateral secondary degeneration of the RPE — may mimic retinitis pigmentosa — narrowing of the retinal and choroidal vessels; optic disc pallor with sharp margins; variable deposits of pigment; less common
      - retinal vasculitis — less common
  - optic neuritis

Clinical presentation

- affects all structures
- unilateral or bilateral
- symptoms: pain; redness; photophobia; blurred vision; floaters
- anterior segment findings: iris roseola; vascularized papules (iris papulosa); larger red nodules (iris nodosa); gummata; interstitial keratitis; posterior synechiae; lens dislocation; iris atrophy
- posterior segment findings
  - most common manifestation
  - chorioretinitis
    - focal or multifocal
    - usually with vitritis
    - may become confluent — small
    - grayish yellow
    - located in the postequatorial fundus

Figure 7-41

    - ← retinal vasculitis
    - ← disc edema
    - ← serous retinal detachment
    - ← exudates
  - pathognomonic of secondary syphilis

Figure 7-42

  - posterior placoid chorioretinitis
    - solitary or multifocal
    - macular or papillary
    - yellowish gray
    - at the level of the rpe
    - vitritis
  - FA
    - early hypofluorescence
    - late staining
    - retinal perivenous staining
  - ICG — hypofluorescent spots corresponding to lesions
  - papillitis/neuroretinitis — ← macular star formation
- less common posterior segment presentations
  - retinitis
    - types
      - focal retinitis

Figure 7-43

      - peripheral necrotizing retinitis — may resemble ARN or PORN
      - punctate inner retinal infiltrates

Figure 7-44

    - may become confluent
    - usually with retinal vasculitis
    - slowly progressive
    - good visual outcome — responds dramatically to intravenous penicillin
  - isolated retinal vasculitis — retinal arterioles, capillaries, and larger arteries or veins (or both); best appreciated on FA
  - may masquerade as a branch retinal vein occlusion
  - exudative retinal detachment
- immunocompromised or HIV/AIDS patients
  - may have atypical/more fulminant ocular disease
  - optic neuritis/neuroretinitis are more common in the initial presentation of these patients
  - disease recurrences are noted even after appropriate antibacterial therapy
- neuro-ophthalmic manifestations of syphilis
  - Argyll Robertson pupil
  - ocular motor nerve palsies
  - retrobulbar optic neuritis — optic neuropathy
  - most often in tertiary syphilis or neurosyphilis

Ophthalmology Mind Maps/Arman Mashayekhi, MD