4.14.1. Orbit (I)

Nonspecific orbital inflammation (idiopathic orbital inflammation)

- demographics
  - 5% of orbital lesions
  - adults and children
- clinical
  - abrupt onset
  - pain
  - diffuse
  - compartmentalized
    - orbital myositis — extraocular muscles
    - dacryoadenitis — lacrimal gland
  - "orbital pseudotumor" — may simulate a neoplasm
- histology
  - polymorphous inflammatory response
    - polyclonal
    - perivascular
    - infiltrates muscle
      - involves muscle tendons!
      - Figure 14-2 (© 2020 American Academy of Ophthalmology)
    - early
      - fat necrosis — infiltrates fat
      - CD20 and CD25 receptors
      - sclerosing variant of fibroinflammatory disease — IgG4-positive plasmacytic infiltrate
    - late
      - fibrosing/sclerosing orbititis — may encase extraocular muscles and optic nerve
      - fibrosis
      - interspersed lymphoid follicles with germinal centers
  - Figure 14-3 (© 2020 American Academy of Ophthalmology)

Topography

- 7 bones
  - frontal
  - zygomatic
  - palatine
  - lacrimal
  - sphenoid
  - ethmoid
  - maxillary
  - roof composed of
    - 1 orbital plate of the frontal bone
    - 2 lesser wing of the sphenoid bone
  - lateral wall composed of
    - 1 zygomatic bone
    - 2 greater wing of the sphenoid bone — separated from the lesser wing by the superior orbital fissure
  - floor composed of
    - 1 maxillary bone
    - 2 palatine bone
    - 3 zygomatic bone
  - medial wall composed of
    - 1 orbital plate of the ethmoid bone
    - 2 lacrimal bone — only the lacrimal bone exists wholly within the orbital confines
    - 3 frontal process of the maxillary bone
    - 4 lesser wing of the sphenoid bone — also part of orbital roof
- 30 ml
- proptosis vs. displacement
- lacrimal gland
  - divided into 2 lobes by aponeurosis of levator palpebrae superioris
    - orbital
    - palpebral
  - acini — low cuboidal epithelium
  - ducts
    - low cuboidal epithelium
    - low flat myoepithelial cells

Congenital anomalies

- Congenital cysts of surface epithelium
  - Dermoid cyst
    - most common
    - from embryonic epithelial nests
    - manifests in childhood
    - orbital mass
      - superotemporal orbit
      - cyst rupture --> granulomatous inflammation
      - protrusion through frontozygomatic or frontomaxillary sutures — dumbbell shape
    - histology
      - capsule
      - wall
        - sebaceous glands
        - hair follicles
        - sweat glands
      - lined by keratinized stratified squamous epithelium
      - contains
        - keratin
        - hair
    - Figure 14-1 (© 2020 American Academy of Ophthalmology)
  - Simple epithelial (epidermoid) cyst
    - no adnexal structures
    - lined by
      - respiratory epithelium
      - conjunctival epithelium
      - apocrine epithelium

Amyloid

- primary systemic amyloidosis
- can involve extraocular muscles & nerves
  - ophthalmoplegia
  - ptosis

Orbital inflammatory diseases

- types
  - idiopathic
  - secondary
    - systemic inflammatory disease
    - foreign body
    - infections
- differential diagnosis
  - congenital mass lesions
  - neoplastic diseases
    - lymphoma
    - rhabdomyosarcoma

Thyroid eye disease

- most common cause of unilateral or bilateral proptosis in adults
- terminology
  - Graves disease
  - thyroid ophthalmopathy
  - thyroid-associated orbitopathy
- pathogenesis
  - inflammation of orbital connective tissue
    - lymphocytes
      - T-cell-bound CD154 — attaches & activates
    - plasma cells
      - generally found on B-cells
    - fibroblasts
      - express CD40 receptors
        - upregulate fibroblast inflammatory genes
          - IL-6
          - IL-8
          - PGE2
        - increased synthesis of hyaluronan & glycosaminoglycans
      - express TSH-R
        - higher numbers in early disease
          - decreases with disease duration
          - correlates with serum TSH-R antibody levels
          - up-regulates TSH-R mRNA
            - promotes adipocyte differentiation/ adipogenesis
      - express IGF-I receptor
        - activated by circulating IgG
        - secrete
          - glycosaminoglycans
          - cytokines
          - chemoattractants
        - insulin-like growth factor
  - inflammation and fibrosis of extraocular muscles
    - firm white muscles
    - TED does not involve muscle tendons!
    - fusiform enlargement
    - inferior & medial rectus muscles
  - adipogenesis
- Figure 14-1 (© 2020 American Academy of Ophthalmology)

Ophthalmology Mind Maps/Arman Mashayekhi, MD