4.14.5. Orbit (V): Neoplasia (III)

Nerve sheath tumors

Neurofibroma

- most common nerve sheath tumor
- clinical
  - slow-growing
  - firm/rubbery
- histology
  - cells
    - endoneural fibroblasts
    - Schwann cells
    - axons
  - arranged in ribbons/cords
  - contains axons
  - matrix of myxoid tissue and collagen
  - not encapsulated
- chromosome 9p rearrangement
- isolated neurofibromas
  - no systemic involvement
- plexiform neurofibromas
  - neurofibromatosis type 1
  - S-shaped deformity of upper eyelid
  - connect cell surface and extracellular matrix
    - matricellular proteins
      - CCNs
        - CCN1 gene

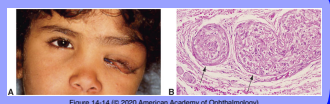

Figure 14-14 (© 2020 American Academy of Ophthalmology)

Neurilemoma (schwannoma)

- arise from Schwann cells
- slow growing
- encapsulated
- associations
  - solitary
  - associated with neurofibromatosis
- patterns
  - Antoni A pattern
    - interlacing whorls/cords/palisades of spindle cells
    - fibrils resembling sensory corpuscles
      - Verocay bodies
  - Antoni B pattern
    - mucoid stroma
    - stellate cells
- histology
  - cysts
  - hemorrhagic necrosis
  - prominent thick-walled vessels
  - no axons
    - in contrast to neurofibromas

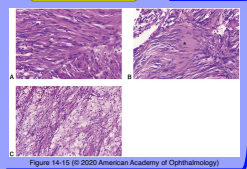

Figure 14-15 (© 2020 American Academy of Ophthalmology)

- immunohistochemistry
  - S-100
  - vimentin
  - CD68

Tumors with fibrous proliferation

Fibrous histiocytoma (fibroxanthoma)

- adults
  - median age=43 years
  - 6 mo - 85 years
- upper nasal orbit
- histology
  - fibroblasts
    - storiform/whorly/matlike pattern
  - histiocytes
  - immunohistochemistry
    - CD45
    - CD68

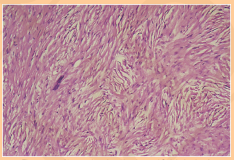

Figure 14-11 (© 2020 American Academy of Ophthalmology)

- clinical behavior
  - benign
    - most common
  - intermediate
  - malignant
    - high mitosis rate
      - >1/HPF
    - nuclear pleomorphism
    - necrosis
    - <10% have metastatic potential
  - locally aggressive
- difficult to distinguish, clinically and histologically, from hemangiopericytoma

Hemangiopericytoma

- similar to embryonal RMS
- solitary fibrous tumor is part of hemangiopericytoma spectrum
- uncommon
- adults
  - median age=42 years
    - similar to fibrous histiocytoma
- clinical
  - proptosis
    - similar to adenoid cystic carcinoma of lacrimal gland
  - pain
  - diplopia
  - decreased visual acuity
- pathology
  - hypervascular
    - staghorn vascular pattern
  - hypercellular
    - plump pericytes that surround a rich capillary network
    - densely packed spindle-shaped cells
    - reticulin stain
      - individual tumor cells wrapped in collagenous material
  - encapsulated
  - no correlation between mitotic rate and clinical behavior
    - microscopically "benign" lesions may recur and metastasize
    - microscopically "malignant" lesions may remain localized
  - immunohistochemistry
    - CD34

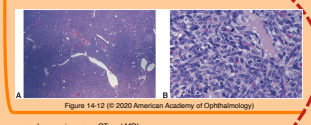

Figure 14-12 (© 2020 American Academy of Ophthalmology)

- resembles cavernous hemangiomas on CT and MRI
- clinical behavior
  - benign
    - most common
  - intermediate
  - malignant
    - infiltrating border
    - anaplasia
    - high mitosis rate
    - necrosis
- treatment
  - complete excision
    - may recur, undergo malignant degeneration, or metastasize
  - appear bluish intraoperatively

Bony lesions

Fibrous dysplasia

- types
  - monostotic
  - polyostotic
- orbital involvement is monostotic
- first 3 decades
- may cross suture lines
- narrowing of optic canal
- narrowing of lacrimal drainage system
- plain radiograph
  - ground-glass appearance
  - lytic foci
  - cysts containing fluid
- histology
  - immature woven bone
    - bony trabeculae
      - C-shaped
  - highly vascularized fibrous stroma

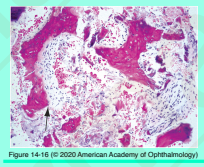

Figure 14-16 (© 2020 American Academy of Ophthalmology)

Fibro-osseus dysplasia (juvenile ossifying fibroma)

- histology
  - rimmed by osteoblasts
    - bone spicules
  - cellular fibrous stroma

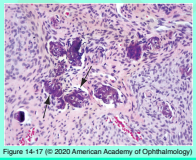

Figure 14-17 (© 2020 American Academy of Ophthalmology)

- psammomatous meningioma
- differential diagnosis

osseous/cartilaginous tumors

- osteoma
  - most common
  - slow growing
  - well circumscribed
  - mature bone
  - frontal sinus
- osteoblastoma
- giant cell tumor
- chondroma
- Ewing sarcoma
- osteogenic sarcoma
- chondrosarcoma

Smooth muscle tumors

- Leiomyoma
  - rare
  - 4th and 5th decade
  - proptosis
    - slowly progressive
  - spindle cells
    - nuclei
      - blunt-ended
      - cigar-shaped
    - cytoplasm
      - filamentous
      - trichrome-positive
- Leiomyosarcoma
  - 7th decade
  - histology
    - have more
      - cellularity
      - necrosis
      - nuclear pleomorphism
      - mitotic figures

Ophthalmology Mind Maps/Arman Mashayekhi, MD