7.5.1. Orbital Neoplasms and Malformations (I): Vascular Tumors (I)

Lymphatic Malformation

- "lymphangiomas"
- vascular dysgenesis
  - disruption of initially pluripotent vascular anlage leads to aberrant development and congenital malformation
- clinical presentation
  - first or second decade of life
  - may also occur in
    - conjunctiva
    - eyelids
    - oropharynx
    - sinuses
  - noncontiguous intracranial vascular malformations
    - ≤25% of patients
  - low rate of spontaneous hemorrhage
    - not treated prophylactically
  - natural history
    - localized/slow progression
    - unpredictable
    - diffuse infiltration/inexorable enlargement
    - sudden spontaneous hemorrhage from interstitial capillaries
      - abrupt proptosis or mass lesion
    - may enlarge during upper respiratory tract infections
      - response of lymphoid tissues within lesion
- histology
  - large, serum-filled channels lined by flat endothelial cells that have immunostaining patterns consistent with lymphatic capillaries
    - not neoplasms!
    - endothelial cells do not proliferate
  - contain both venous and lymphatic components
  - scattered follicles of lymphoid tissue
  - not encapsulated
    - infiltrative pattern
- MRI
  - multiple grapelike cystic lesions with fluid–fluid layering of serum and red blood cells
  - pathognomonic
  - Figure 5-3 (© 2020 American Academy of Ophthalmology)
- management
  - treatment of choice
    - lower adverse effects compared with surgical resection
  - intralesional sclerosing agents
    - successful in treating larger cystic components
      - reducing tumor bulk
    - technique
      - cyst is drained
      - extent is evaluated with contrast media
      - sodium morrhuate
        - introduced, withdrawn, and rinsed
      - absolute alcohol (other sclerosing agents)
        - introduced, withdrawn, and rinsed
      - >1 treatment may be required
    - retained sclerosing foams of doxycycline and bleomycin
      - successful in treating the microcysts
  - surgical intervention
    - deferred unless vision affected
    - subtotal resection
      - to avoid sacrificing important structures
  - indications for treatment
    - significant proptosis
    - optic neuropathy
    - corneal ulceration
  - acute hemorrhage
    - mild hematomas may resolve spontaneously
    - ultrasound-guided aspiration of blood
    - open surgical exploration

Hemangiopericytoma

- uncommon
- appear in midlife
- resemble cavernous hemangiomas on CT and MRI
- appear bluish intraoperatively
- pathology
  - hypervascular
  - hypercellular
  - encapsulated
  - plump pericytes that surround a rich capillary network
- treatment
  - complete excision
    - may recur, undergo malignant degeneration, or metastasize
  - no correlation between mitotic rate and clinical behavior
    - microscopically "benign" lesions may recur and metastasize
    - microscopically "malignant" lesions may remain localized

Infantile (Capillary) Hemangioma

- risk factors
  - female
  - low birth weight
  - prematurity
  - maternal chorionic villus sampling
- natural course
  - present at birth or in the first few weeks of life
  - enlarge dramatically over 6–12 months
  - involute after the first year of life
    - 75% resolve during the first 3–7 years
- clinical presentation
  - superficial (cutaneous)
    - bright red
    - soft
    - dimpled texture
  - subcutaneous
    - bluish
  - deep (within the orbit)
    - enlarging mass without overlying skin change
  - superonasal orbit & medial upper eyelid
    - most common
  - ± hemangiomas of other parts of the body
    - lesions involving neck can compromise airway
    - multiple large visceral lesions can produce thrombocytopenia
      - Kasabach-Merritt syndrome
- ocular complications
  - amblyopia
  - strabismus
  - anisometropia
  - severe disfigurement
- MRI
  - characteristic fine intralesional vascular channels and high blood flow
  - distinguish infantile hemangiomas from other vascular malformations
- management
  - first line of management
    - observation
    - refractive correction
    - amblyopia therapy
    - treatment may be deferred until it is clear that the natural course will not lead to the desired result
  - indications for treatment
    - disruption of vision
    - severe disfigurement
  - β-blockers
    - initial therapy
    - topical timolol gel
      - for superficial tumors
      - limited systemic adverse effects
      - for larger and deeper lesions
    - oral propranolol
      - fewer adverse effects than systemic steroids
        - hypoglycemia
        - cardiovascular adverse effects
    - Figure 5-1 (© 2020 American Academy of Ophthalmology)
    - for lesions unresponsive to or showing limited improvement with β-blockers
  - steroids
    - topical
    - local injection
      - skin necrosis
      - subcutaneous fat atrophy
      - systemic growth retardation
      - retinal embolic vision loss
    - oral
  - surgical excision
    - for nodular lesions that are smaller, subcutaneous, or refractory to steroids
    - meticulous hemostasis
  - radiation therapy
    - cataract formation
    - bony hypoplasia
    - future malignancy
  - pulsed-dye laser therapy
    - superficial components of the hemangioma
  - sclerosing solutions
    - severe scarring
    - not recommended !

Cavernous Hemangioma

- most common benign neoplasm of the orbit in adults
- F>M
- clinical presentation
  - slowly progressive proptosis
    - Figure 5-2 (© 2020 American Academy of Ophthalmology)
  - strabismus
  - optic nerve compression
  - retinal striae
  - hyperopia
  - increased intraocular pressure (IOP)
  - growth may accelerate during pregnancy
- orbital imaging
  - MRI
    - small intralesional vascular channels containing slowly flowing blood
  - well-encapsulated mass
  - homogeneously enhancing
  - radiodense phleboliths
    - chronic lesions
  - coronal imaging
    - important in determining the position of the cavernous hemangioma relative to the optic nerve
  - arteriography and venography
    - not useful because of limited communication with the systemic circulation
- histology
  - encapsulated
  - large cavernous spaces containing red blood cells
  - walls of the spaces contain smooth muscle
- surgical excision
  - indications
    - compromised ocular function
    - significant proptosis
    - significant growth
  - cavernous hemangiomas have surgical planes that allow easier separation from normal orbital tissues
- rarely involute spontaneously

Ophthalmology Mind Maps/Arman Mashayekhi, MD