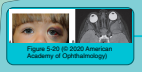
7.5.8. Orbital Neoplasms and Malformations (VII)

Metastatic Tumors

children

Neuroblastoma

- adults more frequently have metastases to the choroid
- in children, distant tumors metastasize to the orbit more frequently than to the globe
- abrupt ecchymotic proptosis
  - eyelid ecchymosis may lead to the mistaken impression of injury
  - 
    Figure 5-20 (© 2020 American Academy of Ophthalmology)
- a bilateral
- a Horner syndrome
- bone destruction
  - particularly lateral orbital wall or sphenoid marrow
  - CT
- orbital metastases typically occur late in the course of the disease
  - primary tumor can be detected readily in abdomen, mediastinum, or neck
- treatment
  - chemotherapy
  - radiotherapy
    - for cases of impending vision loss due to compressive optic neuropathy
- survival rate
  - related to the patient's age at diagnosis
    - <1 year
      - 90% survival
    - >1 year
      - 10%
- congenital neuroblastoma of the cervical ganglia
  - ipsilateral Horner syndrome
  - heterochromia

Leukemia

- acute lymphoblastic leukemia
  - most common leukemia to metastasize to the orbit
  - unilateral or bilateral proptosis
- primary leukemic orbital mass
  - granulocytic sarcoma or chloroma
  - rare variant of myelogenous leukemia
  - metastasis to the subarachnoid space of the optic nerve
    - rare
    - sudden vision loss
    - swelling of the optic nerve
  - emergency
    - treated with orbital radiotherapy
  - special stains for cytoplasmic esterase
    - Leder stain
    - granulocytic precursor cells
  - typically, orbital lesions present before blood or bone marrow signs of leukemia
    - almost invariably follow within several months
    - chances for survival are improved if chemotherapy is instituted before the discovery of leukemia in bone marrow or peripheral blood
- references

adults

- any carcinoma of the internal organs and cutaneous melanoma can metastasize to the orbit
- breast and lung tumors are most common

clinical presentation

- pain
- proptosis
- inflammation
- bone destruction
- early ophthalmoplegia
  - extraocular muscles are frequently involved
    - have abundant blood supply !

bone marrow space of the sphenoid bone

- second most common site
- relatively high volume of low-flow blood
- lytic destruction of this part of the lateral orbital wall is highly suggestive of metastatic disease

diagnostic workup

- known primary tumor
  - 75%
- unknown primary tumor
  - 25%
- elevation of serum carcinoembryonic antigen
- fine-needle aspiration biopsy
  - can be performed in the office
- orbitotomy and open biopsy

breast carcinoma

- most common primary source of orbital metastases in women
- may occur many years after the breast has been removed
- enophthalmos
- may elicit a fibrous response
  -
    Figure 5-22 (© 2020 American Academy of Ophthalmology)
- restriction of ocular motility
- hormonal therapy
  - most likely to help patients whose tumors are receptor-positive
- fresh tissue should be submitted for estrogen receptor assay
  - even if this test was previously performed because estrogen receptor content may differ between the primary and the metastatic lesions

bronchogenic carcinoma

- most frequent origin of orbital metastasis in men
- primary lesion may be quite small
- CT of suspicious lung lesions

prostate carcinoma

- can produce a clinical picture resembling that of acute nonspecific orbital inflammation
- lytic bone lesion
  -
    Figure 5-21 (© 2020 American Academy of Ophthalmology)

management of orbital metastases

- palliative
- local radiation therapy
- wide excision
  - carcinoid
  - renal cell carcinoma
- some patients may survive for many years following resection of isolated metastases from these primary tumors

Secondary Orbital Conditions

eyelid origin

- extend into the orbit from contiguous structures
- globe origin
  - choroidal melanomas
  - retinoblastomas
- sebaceous gland carcinoma
- squamous cell carcinoma
- basal cell carcinoma
- references

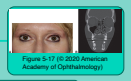
sinus origin

- proptosis and globe displacement

mucoceles and mucopyoceles

- cystic structures with pseudostratified ciliated columnar (respiratory) epithelium resulting from obstruction of the sinus excretory ducts
- invade the orbit by expansion and erosion of the bones of the orbital walls
- mucoceles
  - most mucoceles arise from the frontal or ethmoid sinuses
  -
    Figure 5-16 (© 2020 American Academy of Ophthalmology)
  - filled with thick mucoid secretions
- pyoceles
  - filled with pus
- surgical treatment
  - evacuation of the mucocele
  - reestablishment of drainage of the affected sinus
    - OR
    - obliteration of the sinus by mucosal stripping and packing with bone or fat

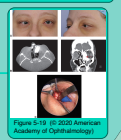
silent sinus syndrome

- chronic subclinical sinusitis
  - causes thinning of the bone of the involved sinus
    - collapse of the orbital floor
      - may occur in association with a recent significant change in atmospheric pressure
        - during airplane travel
        - scuba diving
- enophthalmos
  - 
    Figure 5-17 (© 2020 American Academy of Ophthalmology)
- upper eyelid may appear relatively retracted
- a transient diplopia
- treatment
  - restoration of normal sinus drainage
  - reconstruction of the orbital floor

squamous cell carcinoma and adenocarcinoma

-
  Figure 5-18 (© 2020 American Academy of Ophthalmology)
- usually arise within the maxillary sinuses
  - followed by the nasopharynx or the oropharynx
- nasal obstruction
- epistaxis
- epiphora
- treatment
  - surgical excision
  - radiation therapy
  - exenteration if the periorbita is traversed by tumor

nonepithelial tumors from the sinuses, nose, and facial bones

- osteomas
  - 
    Figure 5-19 (© 2020 American Academy of Ophthalmology)
- fibrous dysplasia
- miscellaneous sarcomas

survival rate

- opposite of histiocytoma tumors
- only 50%
  - <2 years
- 80%
  - >2 years

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