Clinical Pathological Correlation of Proptosis and Globe Dystopia

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Disclosures

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Proptosis

- Most common cause of unilateral and bilateral proptosis in an adult is thyroid eye disease
- Most common cause of unilateral proptosis in a child is cellulitis
- Most common cause of bilateral proptosis in a child is metastatic neuroblastoma and leukemia
Proptosis

- In cases of unilateral proptosis must carefully differentiate between true protrusion and enophthalmos of the other eye
- Most common causes of enophthalmos in a child is microphthalmos
- Most common cause of enophthalmos in an adult is old trauma but must always rule out scirrhous breast cancer mets and sclerosing pseudotumor
IgG4 Orbital Inflammation Variant

- Systemic variant of sclerosing orbital inflammation
- First recognized as a discrete entity in 2003 with extra pancreatic manifestations in autoimmune pancreatitis\(^1\)
- Now has been identified in virtually every organ system including the orbit\(^1\)
- Dense lymphoplasmacytic infiltration with plasma cells rich in IgG4\(^1\)
- Produces elevated levels of IgG4 in the blood and can respond to systemic immune modulation/suppression\(^1\)

When evaluating a patient with proptosis, it is also important to see if the eye is being moved in a specific direction or if the movement is purely axial.

The differential diagnosis is significantly impacted by relative movement of the globe.

Globe typically moves opposite the location of the lesion.
Case #1 Superonasal Mass

- 8 y.o old white male presented with one month history of rapid proptosis and inferolateral displacement of the globe
- First though to be a “bug bite”
- Then thought to be pre-septal cellulitis that did not respond to oral antibiotics
- When proptosis, dystopia, and decreased motility readily apparent, referred to SEI
Case #1 Superonasal Mass

Case # 1 Superonasal Mass

- MRI showed a large superonasal mass extending to the apex
- Lid crease incision and anterior orbitotomy performed
- Large bluish, highly vascular tumor identified, debulked, and biopsied
- Tumor was mostly solid but seemed to have areas that were liquefied
- Most of incision closed but external drain placed due to vascular nature of mass
Case #1 Superonasal Mass

Alveolar Rhabdomyosarcoma

H&E, 100x
Alveolar Rhabdomyosarcoma

H&E, 400x
Orbital Rhabdomyosarcoma

- Usually presents as a rapid onset of proptosis with inferior / infero-temporal displacement of the globe
- Past history of trauma may delay diagnosis
- Often simulates an orbital cellulitis due to rapid onset
- 4 histopathological subtypes:
  - Embryonal - most common
  - Alveolar - worst prognosis
    - Usually has rearrangement of FOXO1A (1;13 or 2;13)
  - Differentiated (pleomorphic) - least common, best prognosis
  - Botryoid (a variant of embryonal)
Differential Diagnosis: Superonasal Lesions

- Mucocele / encephalocele
- Rhabdomyosarcoma - commonly superior or superonasal location
- Hemangiopericytoma - commonly superior location
Case #2 Superolateral Mass

- 84 y.o A.A. female presented with history of intermittent redness, swelling, and pain of left eye
- Patient had what appeared to be post-inflammatory hyperpigmentation of skin around left eye
- Patient had palpable mass in superolateral quadrant
- CT scan with contrast demonstrated lacrimal gland asymmetry with molding to the globe on the left
- Anterior orbitotomy with lid crease incision performed with incisional biopsy since lesion did not radiographically appear to be an epithelial lesion
Case #2 Superolateral Mass
Recurrent Dacryoadenitis

- Several weeks post-op, patient presented with proptosis, decreased motility, redness, and chemosis
- Started on 50mg of oral prednisone with rapid response and improvement
- When tapered to 10mg inflammation recurred and dose elevated to 50mg again with rapid response
- Currently on slow taper and if cannot taper effectively will re-image and biopsy again to rule out lymphoproliferative disease
Recurrent Dacryoadenitis
Dacryoadenitis

H&E, 40x
Dacryoadenitis

H&E, 200x
Dacryoadenitis

H&E, 400x
Dacryoadenitis

- **Acute:**
  - Infectious: bacterial, viral, or fungal
  - Generally presents with pain, redness, and rapid onset

- **Chronic:**
  - Differential includes sarcoid, Sjogrens, Graves disease, nonspecific orbital inflammatory disease (NSOI), chronic infections
  - Generally presents with painless enlargement of the lacrimal gland for >1 month (although our case showed a relapsing/remitting course), can be bilateral
Differential Diagnosis: Superolateral Lesions

- Dermoid
- Lacrimal gland lesions:
  - Dacryoadenitis (inflammation of the lacrimal gland)
  - Dacryops (lacrical duct cysts)
  - Benign mixed adenoma / pleomorphic adenoma
  - Adenoid cystic carcinoma
  - Lymphoma
Other considerations for inferior displacement of the globe:

- Lymphangioma (usually superior or nasal quadrants)
- Orbital extension of a plexiform neurofibroma
- Schwannoma
- Orbital floor fracture
74 y.o. white male presented to SEI with long standing history of painless proptosis and superonasal globe dystopia

- Palpable mass in inferolateral quadrant pushing eye up and in
- CT scan revealed large well circumscribed extraconal mass
- Transconjunctival swinging eyelid inferior orbitotomy performed with excisional biopsy of lesion
Case # 3 Inferolateral Mass
Case # 3 Inferolateral Mass
Schwannoma

H&E, 40x
Schwannoma

S100, 40x
Schwannoma

- Rare, benign tumor composed of the myelinating cells of peripheral nerves (Schwann cells)
- Often located in the superior orbit... but not always
- Well-circumscribed lesion on imaging
- Histopathology notable for Antoni A (with Verocay bodies) and Antoni B patterns

Histopathology of Schwannomas

- Antoni A and Antoni B patterns:
  - Antoni A = cellular areas
  - Antoni B = paucicellular areas, possibly degenerating cells from Antoni A
- Verocay bodies = palisading nuclei surrounding pink areas, resembling sensory corpuscles
- Staining patterns:
  - Positive for S100
Schwannoma

H&E, 200x
Differential Diagnosis: Inferolateral Lesions

- Cavernous hemangioma (also common in axial location)
- Varix (thrombosed)
Case # 4 Inferomedial Lesion

- 9 y.o. white female presented to MUSC with 3 day history of cold symptoms to include stuffy nose and fatigue
- On day of presentation awoke with significant proptosis, pain, and decreased motility
- Seen at local ED where CT scan demonstrated large subperiosteal abscess with concern for intracranial extension
- Patient flown from Florence to MUSC for management
Case # 4 Inferomedial Lesion

- CT scan repeated that showed ethmoid and maxillary sinusitis, large SPA, and no evidence of intracranial spread
- Afferent system intact but patient taken emergently to OR for endoscopic drainage of SPA and sinuses
- Significantly better on post op day one but worsens on post op day 2
- Re-imaging shows orbital SPA better but new SPA below inferior turbinate
- Patient taken back to OR for re-drainage
Case # 4 Inferomedial Lesion
Orbital Cellulitis and SPA

- Smaller SPAs in kids can be watched on IV antibiotics and many respond without drainage.
- Large SPAs should be drained.
- Preferred method is to drain the sinuses and abscess at the same time to prevent recurrence.
- This case shows that careful follow up also important and if does not respond promptly should re-image!
Differential Diagnosis: Inferomedial Lesions

- Subperiosteal abscess
- Lacrimal sac lesions - including tumors and infectious processes
Axial Proptosis

- 75 yo A.A. female presents to USC with sudden onset of axial proptosis and decreased motility of the left eye
- CT scan shows large intraconal mass with extension inferolaterally with air-fluid line
- Afferent system intact and patient admitted overnight for observation and then transferred to SEI
- MRI demonstrated heterogeneous internal contents of lesion consistent with blood breakdown products
- Afferent system still intact so decision to follow clinically before decision made to obtain tissue
Axial Proptosis
Axial Proptosis

- Over next several weeks patient improved significantly with decreased pain and proptosis and improved motility
- Most likely spontaneous hemorrhage into orbital varix
Differential Diagnosis of Axial Proptosis

- Cavernous hemangioma
- Fibrous histiocytoma
- Solitary fibrous tumor
- Schwannoma
- Hemangiopericytoma
Cavernous hemangioma
References
