Orbital Inflammatory Disease

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Disclosure

- I have no financial disclosures that would be a potential conflict of interest with this presentation.
- I will not be discussing off label uses of medications or unapproved uses of a commercial product or device.

Goals

- Review orbital infections and inflammatory conditions.
- Discuss the Chandler classification of orbital infection. Knowledge of this system aids communication with our referring physicians.
- Review potential complications of orbital infections – what to watch out for!

Organization

- Pertinent orbital anatomy.
- Idiopathic orbital inflammatory disease.
- Review remaining inflammatory diseases on an anatomic basis.

Osteology

- Zygomatic
- Maxillary
- Frontal
- Lacrimal
- Ethmoid
- Sphenoid
- Palatine

- Constructed of 7 separate bones
Orbital Septum
- Originates at the confluence of the periorbita and periosteum.
- Inserts at or near the tarsal plates, deep to the orbicularis oculi muscle.
- Separates the orbital fat from the subcutaneous fat - barrier for infection.

Globe and Optic Nerve
- Anterior Chamber
- Iris
- Lens
- Vitreous Chamber
- Dural Sheath
- Optic Nerve
- CSF
The venous system in and about the orbit is “valveless”, allowing bidirectional flow and can be a conduit for spread of infection.

Two main venous channels:
- Superior ophthalmic vein
  - Originates near superior oblique, courses posteriorly and laterally through orbit to enter the cavernous sinus.
- Inferior ophthalmic vein
  - Drains a venous plexus along the floor of the orbit, can terminate in the pterygoid plexus, superior orbital vein or cavernous sinus.
Superior Ophthalmic Veins
Superior Ophthalmic Veins

Superior Ophthalmic Veins
Idiopathic Orbital Inflammatory Disease

- The disease previously known as “pseudotumor”.
- 5% of orbital conditions, behind thyroid orbitopathy, and lymphoproliferative disease.
- Diagnosis of exclusion, no definitive pathologic or clinical criteria.
- My pragmatic definition: No response to abx, typically responds to steroids, no identifiable systemic autoimmune condition.

Mimics of IOID

- IgG4 Related Disease.
- Thyroid Orbitopathy
- Sarcoidosis
- Amyloidosis
- Granulomatosis with Polyangiitis
- Crohns disease
- Systemic Lupus Erythematous
- Scleroderma
- Giant Cell Arteritis
- Lymphoma
- Metastatic disease
- Orbital Cellulitis
IgG4 Related Disease

- Tumor like fibroinflammatory condition which can affect multiple organ systems.
- Often associated with elevated serum IgG4 levels, and/or IgG4 positive lymphocytes on biopsy.
- First reported in association with autoimmune pancreatitis, now associated with almost every organ system.

IgG4 Related Disease

- Head and Neck diseases with IgG4 contributions
  - Idiopathic Orbital Inflammatory Disease
  - Idiopathic Hypertrophic Pachymeningitis
  - Mikulicz Syndrome (Salivary and Lacrimal Glands)
  - Riedel’s Thyroiditis
  - Kuttner’s Tumor (Submandibular Glands)

Idiopathic Orbital Inflammatory Disease

**Tolosa Hunt –**

**Description:**

“Episodic orbital pain associated with paralysis of one or more of the third, fourth and/or sixth cranial nerves which usually resolves spontaneously but tends to relapse and remit.”

**Diagnostic criteria:**

- One or more episodes of unilateral orbital pain
- Paresis of one or more of cranial nerves III, IV or VI and/or demonstration of granuloma by MRI or biopsy
- Paresis coincides with the onset of pain or follows it within 2 weeks
- Pain and paresis resolve within 72 hours when treated adequately with corticosteroids
- Other causes have been excluded by appropriate investigations

Normal Cavernous Sinus

Uniformly Enhancing
Concave, smooth borders.

29 yo female: L retro-orbital pain & progressive cranial neuropathies 3, 4, V1, V2, & 6

Case Courtesy of Christopher Wood, MD

Chandler Classification

Classification of orbital infection based on anatomic site of involvement, development of a defined abscess, and etiology.

Chandler I through Chandler V.
  - The categories are not sequential.
  - The Chandler classification has been very effective in helping guide appropriate diagnosis and therapy for orbital infections.

Anatomic Sites

- Preseptal / Periorbital Cellulitis
- Postseptal / Orbital Cellulitis
- Subperiosteal Abscess
- Cavernous Sinus
- Dacryoadenitis
- Dacryocystitis
- Globe
- Optic Nerve
- Myositis

Preseptal / Periorbital Cellulitis

- Infection of the soft tissues anterior to the orbital septum.
- Most common in pediatric patients – often subsequent to trauma, acne or insect bite.
- If treated, it is relatively uncommon to spread into the postseptal orbit.
- Typically caused by staph, strep, or HiB.

- Most are treated as outpatients.
- The imaging goals are to exclude postseptal cellulitis, identify drainable abscess, exclude intracranial complications and causative sinus disease.

Preseptal Cellulitis

- Often secondary to sinus infection – spreading to the orbit via veins and/or through the thin bony septae.
- Clinical hallmarks of proptosis, chemosis, ophthalmoplegia and visual acuity loss.
- Infection in a closed space – increased pressure, lack of drainage.
- Initial tx is usually medical: abx, +/- steroids, +/- sinus irrigation, +/- sinus vasoconstrictors.
Postseptal Cellulitis

- Often secondary to sinus infection – spreading to the orbit via veins and/or through the thin bony septae.
- Clinical hallmarks of proptosis, chemosis, ophthalmoplegia, and visual acuity loss.
- Infection in a closed space – increased pressure, lack of drainage.
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Postseptal Cellulitis

- Typically inpatient tx. Repeat CT and/or MRI with persistent symptoms or worsening exophthalmos.
- Surgical decompression is typically advised with persistent or worsening symptoms after 48 hours.
- Imaging goals are to document abscess formation, venous thrombosis, cavernous sinus thrombosis.

15 yo with IDDM 1, poorly controlled
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Subperiosteal Abscess

- Typically from extension of ethmoid sinusitis through the lamina papyracea.
- The abscess characteristics guide medical versus surgical tx.
Subperiosteal Abscess

- Medical treatment can be attempted if the abscess...
  - is medial in location.
  - is less than 1 cm in size.
  - is not dental or frontal sinus in origin.
  - presents without optic nerve compromise.
  - presents without gas within the abscess.

Subperiosteal Abscess

- Surgical decompression is typically via endoscopic sinus surgery, with drainage into the sinonasal cavity.
- This can be a challenge in young patients.
- Alternative procedure involves a medial canthotomy to access the abscess.
- Imaging assists with guidance, abscess characterization, and exclusion of remote abscess pockets which might warrant an external approach.

Subperiosteal Abscess + Gas

- 7 year old with headache and left "eye swelling".

Subperiosteal + Epidural Abscess

- 14 year old with headache, fever, and left periorbital swelling.
Subperiosteal + Epidural Abscess

14 year old with headache, fever, and left periorbital swelling.

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Cavernous Sinus Thrombosis
- A feared complication of orbital cellulitis.
- Mortality of 30%. 50% of survivors will have cranial nerve deficits.
- Presents with chemosis, visual acuity loss, and ophthalmoplegia – nonspecific signs similar to orbital cellulitis.
- May spread to the contralateral side. Radiologists must be vigilant!
- Look for an expanded cavernous sinus with convex lateral border, filling defects, thrombosed SOV.

Cavernous Sinus Thrombosis
- Tx with abx, +/- anticoagulation.
- High incidence of intracranial infection and meningitis – remember to evaluate the adjacent brain and meninges!

Normal Cavernous Sinus
- Uniformly Enhancing
- Concave, smooth borders.

Cavernous Sinus Thrombosis
- 18 year old with headache, left facial swelling and ophthalmoplegia.
Cavernous Sinus Thrombosis

18 year old with headache, left facial swelling and ophthalmoplegia.
Cavernous Sinus Thrombosis

18 year old with headache, left facial swelling and ophthalmoplegia.
Cavernous Sinus Thrombosis

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Dacryoadenitis
- Can be viral, bacterial, or due to inflammatory conditions such as Sarcoid, Sjögrens, GPA, or Idiopathic Orbital Inflammatory Disease.
- Use bilaterality as a clue – viral, autoimmune, or neoplastic causes are more likely with bilateral disease.

29 yo female, IDDM 1, renal osteodystrophy, renal transplant

16 year old female with orbital swelling
Dacryoadenitis

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Dacryocystitis
- An infection of the lacrimal or nasolacrimal ducts.
- Can develop as a consequence of a dacryocystocele – a dilated lacrimal duct formed following narrowing or obstruction of the lacrimal duct.
- Initial tx via antibiotics, relief of obstruction via probing the duct, or dacryocystorhinostomy.
Dacryocystitis

3 year old male, 10 days epiphora, concern for orbital cellulitis

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Inflammation Involving the Globe

- Subdivided by anatomic site.
- Not commonly imaged.
- Can be due to instrumentation, foreign body, trauma, virus (CMV), parasites (toxo), or autoimmune reactions.

Conjunctivitis
Keratitis
Scleritis
Uveitis
  - Anterior
  - Posterior
Retinitis
Papillitis
Endophthalmitis

Strep Pneumococcus Endophthalmitis

60 year old with left sided pain and vision loss.

Recurrent Idiopathic Scleritis

60 year old with left sided pain and vision loss.
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Optic Neuritis / Neuropathy
- Described by segment – intraorbital, intracanalicular, prechiasmatic, and chiasmatic.
- Optic neuritis is commonly associated with demyelinating conditions.
- Some authors reserve the term for demyelination, preferring to use inflammatory optic neuropathy for non-demyelinating conditions.

Optic Neuritis
- Most common in females, 30 to 40 years old.
- ~38% go on to develop clinical MS.
- A single >3mm white matter lesion increases risk of subsequent MS to 56%. With no lesions 22% go on to clinical MS.
- Neuromyelitis Optica / Devics disease is a demyelinating condition caused by anti-aquaporin-4 IgG. Presents with ON (sometimes bilateral) and myelitis (>3 segments).

Inflammatory Optic Neuropathy
- Can be caused by a variety of conditions:
  - Bacterial: Lyme, Syphilis, Bartonella
  - Autoimmune: Sjögrens, Behçets, GPA, IBD, Idiopathic Perineuritis (IOID), Lupus, or Sarcoidosis.
  - Viral: CMV, Varicella
- Often involves the nerve sheath “perineuritis”, as well as the optic nerve.

Herpes Zoster Ophthalmicus
- 60 year old woman with left sided periorbital pain and V1 distribution vesicles.

Inflammatory Optic Neuropathy
- 31 yo male. Painful left orbit, near complete vision loss. HSV, lyme, bartonella serologies negative with multiple LPs, negative C-ANCA
Inflammatory Optic Neuropathy

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- **Myositis**

**Myositis**
- Thyroid orbitopathy
  - Classically bilateral, relatively symmetric, sparing myotendinous junction.
  - Isolated muscle involvement in 5%.
- Idiopathic Orbital Inflammatory Disease
  - Typically unilateral, equal distribution, involves myotendinous junction.
  - Bilateral or multiple muscle involvement increases frequency of recurrence.

**Myositis**
- Autoimmune processes are relatively common, can be confused with IOID. Sarcoid, GPA, Crohns disease have all been reported.
- Bacterial infection is rare, but can occur in conjunction with orbital cellulitis.
- Extraocular enlargement can occur with a cavernous sinus-carotid fistula.
- Lymphoma and metastasis must be also be considered.

**Postseptal Abscess / Myositis**

Chandler IV 34 yo F with orbital pain, DM II

**Postseptal Abscess / Myositis**

Chandler IV 34 yo F with orbital pain, DM II
IgG4 Related Disease

52 year old female with 3 weeks of right eye pain and diplopia.

IgG4 Related Disease

52 year old female with 3 weeks of right eye pain and diplopia.

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52 year old female with 3 weeks of right eye pain and diplopia.

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Take Home Points

- Chandler Classification – First step to guiding clinical therapy.
- IOID – Diagnosis of exclusion, responsive to steroids, consider IgG4, GPA, other autoimmune conditions.
- Always check carefully for subperiosteal and epidural abscesses!
- Be extremely vigilant in immunocompromised or diabetic patients with orbital pain.

Take Home Images