Learning objectives:
1. Demonstrate how appropriate localization of a finding will determine the differential.
2. See how a few pieces of clinical information can affect the differential.
3. Focus on using signal characteristics and enhancement patterns to come to a favored diagnosis.

Case Studies In the Orbit

Case # 1

- 63 year old female with progressive painless right visual field loss

Axial T1 FS pre and post

Axial T2 FS and Sagittal T1 FS post

Differential

- Optic nerve sheath meningioma
- Optic pathway glioma
- Lymphoma
- Orbital pseudotumor
**Optic nerve glioma**

- Enlargement of nerve itself and not around the nerve
- Infiltrative, typically low T2 signal intensity

**Orbital lymphoma**

- Painful condition that affects multiple structures

**Orbital pseudotumor**

- Benign tumor from arachnoid cap cells
- DEEP to the dura and thus are contained by and spread along dura
- F>M
- Mean age ~40 years old
- Associated with NF2 (MISME)

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**Differential**

- Optic nerve sheath meningioma
- Optic pathway glioma
- Lymphoma
- Orbital pseudotumor

**Case # 1 = Optic Nerve Sheath Meningioma**
Optic Nerve Sheath Meningioma Presentation

- Painless visual field loss in >90% of cases
  - Worse with lesions near apex
- Painless proptosis present in majority
  - Worse with lesions near globe

Optic Nerve Sheath Meningioma Findings

- T1 isointense, T2 iso/hyperintense to nerve
- Homogenous enhancement
  - Tubular most common, exophytic rarer
- Calcifications on CT
- Perioptic cysts can develop between globe and tumor

Case # 2

- 35 year old male with rapid onset bilateral painless vision loss

Axial T2 FS and Coronal T1 FS post

- Nerves normal in size/signal, no enhancement

Axial DWI and ADC

- Restricted diffusion of bilateral retrobulbar optic nerves
- No STIR signal or enhancement
Differential

- Optic neuritis
- Optic pathway glioma
- Ischemic optic neuropathy

Patient underwent partial resection of large cervical meningioma.

Case # 2: Posterior Ischemic Optic Neuropathy

- Patient underwent prolonged spinal surgery in the prone position and awoke with complete vision loss
- Rarer than AION
- Associated with prolonged surgeries (CABG)
  - Prolonged hypotension
  - Cardiac risk factors
Posterior Ischemic Optic Neuropathy

- Anterior optic nerve
- Short posterior ciliary arteries
- Posterior optic nerve
  - Circumferential branches of ophthalmic/retinal arteries

Posterior Ischemic Optic Neuropathy Findings

- High diffusion signal of the retrobulbar optic nerve and/or chiasm
- ADC maps may be difficult to interpret
- T2/STIR signal may be normal
- Enhancement may occur in subacute/late stages

Posterior Ischemic Optic Neuropathy Findings

- High diffusion signal of the retrobulbar optic nerve and/or chiasm
- ADC maps may be difficult to interpret
- T2/STIR signal may be normal
- Enhancement may occur in subacute/late stages

Anterior Ischemic Optic Neuropathy

- 45 year old female with slowly progressive painless proptosis and now with right visual field loss

Case # 3

Axial CTA and T1 FS post

- Mass at orbital apex, eccentric enhancement
- Displaces ophthalmic vein
- Low signal intensity rim
- Cor performed after Axial, progressive enhancement

Coronal STIR and T1 FS post

- STIR hyperintense with low intensity septae
- Low signal intensity capsule/soft tissue
- Intraconal
- Circumscribed/Non-agressve
- Extraconal/Extraoral tissues normal
- Progressive, centripetal enhancement

Case courtesy of Dr. Karen Salzman MD.
**Differential**

- **Schwannoma**  Homogenously enhancing, symptomatic when small
- **Cavernous malformation**
- **Orbital apex meningioma**
- **Dermoid/Epidermoid**
- **Venolymphatic malformation/Varix**

**Oculomotor Schwannoma**

**Differential**

- **Schwannoma**  Homogenously enhancing, symptomatic when small
- **Cavernous malformation**
- **Orbital apex meningioma**
- **Dermoid/Epidermoid**
- **Venolymphatic malformation/Varix**

**Orbital Apex Meningioma**

**Differential**

- **Schwannoma**  Homogenously enhancing, symptomatic when small
- **Cavernous malformation**  Homogenous, non-progressive enhancement
- **Orbital apex meningioma**
- **Dermoid/Epidermoid**  Fat attenuation/intensity, non-enhancing
- **Venolymphatic malformation/Varix**

**Superficial Angular Dermoid**
### Differential

- **Schwannoma**
  - Homogenously enhancing, symptomatic when small
- **Cavernous malformation**
- **Orbital apex meningioma**
  - Homogenous, non-progressive enhancement
- **Dermoid/Epidermoid**
  - Fat attenuation/intensity, non-enhancing
- **Venolymphatic malformation/Varix**
  - Fluid-fluid levels, trans-spatial, follows course of vein

### Orbital varix

- T1 FS with and without Valsalva

### Case # 3: Cavernous Malformation

- Most common vascular orbital lesion
  - ISSVA \(\rightarrow\) Slow flow venous/vascular malformation
- Dilated vascular spaces with fibrous capsule and septae
- No dominant arterial supply
- F>M, average age 40

### Cavernous Malformation Presentation

- Slowly progressive painless proptosis
- Diplopia may occur from EOM compression or CN compression
- Visual field deficits may occur from compression of ON if near orbital apex

### Cavernous Malformation Findings

- Non-aggressive intraconal mass
- T1 isointense, T2/STIR hyperintense
  - T2 hypointense capsule and septae
- Progressive irregular centripetal enhancement
- May have areas of T1 hyperintense thrombus

### Centrifugal enhancement

Sequential axial T1 sequences obtained before contrast and at 60, 100, and 300 seconds after contrast administration.
Case # 4

- 72 year old male with “fuzzy vision” on the left
- Patient unable to clarify further

Axial T1 pre and T1 FS post

Differential

- Choroidal/Retinal detachment Possibly … but enhancement?
- Uveal nerve sheath tumor Very rare, high T2 signal
- Uveal melanoma
- Uveal metastases
- Retinoblastoma

Differential

- Choroidal/Retinal detachment Possibly … but enhancement?
- Uveal nerve sheath tumor Very rare, high T2 signal
- Uveal melanoma
- Uveal metastases
- Retinoblastoma

Metastatic prostate adenocarcinoma to the globe with associated retinal detachment

Disease of the young
Retinoblastoma with retinal detachment
Axial T2, T1 pre, and T1 post contrast sequences in a 7 year old male.

Case # 4: Uveal Melanoma

- Most common primary intra-ocular malignancy
- White population
- Incidence increases with age, rarely seen before age 20

Uveal Melanoma Presentation

- Depends on location, can occur anywhere uveal tissue is present
- Iris lesions visually apparent and diagnosed early
- Choroidal lesions found on fundoscopy, often after retinal detachment has occurred

Uveal Melanoma Findings

- Intrinsic T1 hyperintense mass
  - Melanin and hemorrhage
  - Variable but robust enhancement
  - Variable T2 signal
  - Look for exudative retinal detachment
  - T1 bright, T2 bright, non-enhancing

Case # 5

- 76 year old male with dementia, family reports he’s “not seeing as well lately”
- No other clinical information provided
- Patient could not get contrast due to ESRD
Axial T2 and T2 FS

- V-shaped linear filling defect within the globe
- Circumferential, T2 hyperintense on both sides
- Lines converge to optic disc
- No extension past 2:00 or 10:00

Axial DWI and Sagittal T1

- Contained T1 hyperintensity of deep aspect
- High diffusion signal of deep aspect
- Linear, circumferential vitreous filling defect
- Lines converge to optic disc
- No extension beyond 2:00 or 10:00
- No definite mass

Differential

- Choroidal detachment
- Retinal detachment
- Intra-vitreous hemorrhage
- Ocular melanoma/metastasis

Main differential:

Amorphous, no linear filling defect

Large Intra-Vitreous Hemorrhage

Hematocrit effect

Differential

- Choroidal detachment
- Retinal detachment
- Intra-vitreous hemorrhage
- Ocular melanoma/metastasis

Main differential:

Amorphous, no linear filling defect

Should be considered, no contrast enhanced images acquired

Metastatic prostate adenocarcinoma to the globe with associated retinal detachment
Case # 5: Retinal Detachment

- Separation of the retina from the underlying choroid
- May occur due to:
  - Trauma
  - Exudative fluid accumulation
  - Traction band formation
  - Post-operative complications

Retinal Detachment Presentation

- More commonly older patients with other ocular pathology (prior cataract surgery)
- Myopic patients more at risk, longer globe
- Sudden onset vision loss if traumatic/hemorrhagic
- Senile patients or those with pre-existing vision loss, may not be expected

Retinal Detachment Findings

- Curvilinear defect
- Limited posteriorly by the optic disk
  - Lines CONVERGE posteriorly
- Limited anteriorly by ora serrata
  - Lines do not extend beyond 2:00,10:00
- Fluid in the subretinal space
  - Simple fluid or blood products

Case # 6

- 47 year old female with progressive painless proptosis bilaterally, now with visual field loss

Retinal Detachment Findings

- Ora serrata = Junction between retina and ciliary body

Subretinal hemorrhage (Left)
Subchoroidal hemorrhage (Right)

Subretinal hemorrhage (Left)

Coronal T1 FS and FS post
Axial STIR and T1 FS post

- T2/STIR signal of muscles, mild proptosis
- Crowding at orbital apex
- Spared of distal insertion sites
- Intraconal fat prominent but not involved
- Orbital pseudotumor
- Myositic Orbital Pseudotumor
- Intraconal fat prominent but not involved
- Crowding of apex

Differential
- Orbital pseudotumor
- Thyroid associated orbitopathy
- Sarcoidosis
- Lymphoma
- Erdheim-Chester

Myositis Orbital Pseudotumor

- Enlarged EOMs, proptosis
- Tendons spared
- Prominent but not inflamed intraconal fat

Differential
- Orbital pseudotumor
- Thyroid associated orbitopathy
- Sarcoidosis
- Lymphoma
- Erdheim-Chester

Orbital Sarcoidosis Causing Chronic Ptosis

- Patient with chronic ptosis and systemic sarcoidosis. Infiltrative enlargement and enhancement of superior rectus and LPS.

Differential
- Orbital pseudotumor
- Thyroid associated orbitopathy
- Sarcoidosis
- Lymphoma
- Erdheim-Chester

Infiltrative processes, T2 hypointense
Orbital Erdheim-Chester
Infiltrative enhancing low T2 signal intraconal lesions.
Darin A. et al., Cerebral, facial, and orbital involvement in Erdheim-Chester disease. CT and MR imaging findings. Radiology 2010 255:2586-594

Case # 6: Thyroid Associated Orbitopathy

- Most common cause of painless proptosis
- Patients with Graves disease, rarely lymphocytic thyroiditis (Hashimoto’s)
  - Women > Men by 5:1, middle-aged
- Mechanism still not fully understood
  - Inflammatory cells in muscles ramped up by TSH antibody cross-reaction

Thyroid Orbitopathy Presentation

- Patient with thyroid disease
  - May be iatropic stimulus
- Painless proptosis, chemosis, corneal dryness
  - Bilateral 75-90%, symmetric 60-80%
- Diplopia and vision loss when severe
  - Compression of orbital apex

Thyroid Orbitopathy Findings

- Bilateral symmetric muscle enlargement
  - SPARING of anterior myotendinous junction
- IMSLO:
  - Inferior→Medial→Superior→Lateral→Obliques
- Fatty infiltration of muscle
- Increased volume of intraconal fat

Thyroid Orbitopathy = Inflammatory Process

Patient with Graves’ disease and painless proptosis. CT images before and after treatment with TNF-α inhibitors.

Case # 7

- 17 year old male with history of horizontal diplopia and now with painless proptosis on the right
Axial T1 pre and T1 FS post

- Circumscribed mass, fluid-fluid level
- Faint peripheral enhancement
- Medial rectus displaced, no fat stranding

Axial T2 FS and DWI

- No significant diffusion signal
- Non-dependent aspect saturates out on FS
- Extracranal
- Circumscribed/Non-aggressive mass
- Lipid/Fat containing
- Fluid-fluid level
- No diffusion abnormality

Differential

- Cavernous malformation
  - Intracranal, progressive enhancement
- Orbital dermoid
- Orbital epidermoid
- Venolymphatic malformation/Varix

Orbital Cavernous Malformation

Differential

- Cavernous malformation
  - Intracranal, progressive enhancement
- Orbital dermoid
- Orbital epidermoid
- Venolymphatic malformation/Varix

Main differential, no diffusion restriction

External Angular Orbital Epidermoid
Differential

- Cavernous malformation
  - Intracanal, progressive enhancement
- Orbital dermoid
  - Main differential, no diffusion restriction
- Orbital epidermoid
- Venolymphatic malformation/Varix
  - Fluid-fluid levels, trans-spatial, follows course of vein

Orbital varix
T1 FS with and without Valsalva


Case # 7: Deep Orbital Dermoid

- Congenital lesion, most common orbital tumor in children
- Abnormal ectodermal remnant lined by squamous epithelium
- Has epidermal components: hair, sweat, sebaceous glands
- Has NO macroscopic fat, T1 signal from oily sebaceous material

Orbital Dermoid Presentation

- Child/Adolescent
- Superficial dermoid more common
  - External angular dermoid occurs at lacrimal fossa
- Slow growing, may be diagnosed later in life if deep
- Painless mass
- Painless proptosis, ophthalmoplegia

External Angular Dermoid
Incidental finding on CT obtained for headache. Fat attenuation lesion of the lacrimal fossa.

Orbital Dermoid Findings

- Circumscribed mass
  - Most commonly lacrimal fossa
- Non-enhancing
- Calcifications on CT rare
- Lipid signal characteristics
- Minimal to no diffusion abnormality
- Inflammatory changes if rupture occurs
Case # 8

- 58 year old male with slowly growing painless mass, now with painless proptosis and ptosis on the left
- Of note, patient admitted to picking at lesion for the last week

Axial T1 FS pre and post

Low T1 signal extraconal mass
Localizes to lacrimal gland, minimal to no enhancement of mass
Superficial inflammatory changes

Axial T2 and ADC

Low T2 signal
Low ADC signal
Extraconal
Lacrimal gland, both lobes
Low T1/T2, minimal enhancement
Restricted diffusion
No locally invasive features

Differential

- Infectious dacryoadenitis
- Sarcoidosis
- Orbital pseudotumor
- Sicca syndrome (Sjögren)
- Pleomorphic adenoma
- Malignant salivary gland neoplasm
- Lymphoma

Infectious Dacryoadenitis

- Painful, rapid progression, infectious markers, T2 bright, enhancement
- Painful process, enhancement
- Typically bilateral, more common in women, enhances

Differential

- Infectious dacryoadenitis
- Sarcoidosis
- Orbital pseudotumor
- Sicca syndrome (Sjögren)
- Pleomorphic adenoma
- Malignant salivary gland neoplasm
- Lymphoma
Sicca Syndrome

- Infectious dacryoadenitis
- Sarcoidosis
- Orbital pseudotumor
- Sicca syndrome (Sjögren)
- Pleomorphic adenoma
- Malignant salivary gland neoplasm
- Lymphoma

Differential

- Painful, rapid progression, infectious markers, T2 bright, enhancement
- Enhancement, no systemic disease
- Painful process, enhancement
- Typically bilateral and more common in women
- Enhancement, usually one lobe, invasive

Lacrimal Gland Adenoid Cystic Carcinoma

- Lacrimal gland masses, rule of thirds
  - Inflammatory ~1/3
  - Lymphoproliferative ~1/3
  - Salivary type neoplasm ~1/3
- Involvement of both orbital and palpebral lobes in inflammation/lymphoma
- MALT type most common, follicular next
- MALT = good prognosis

Case # 8: Lacrimal Gland Lymphoma

- Older patients, F>M
- May not have systemic disease
- Progressive painless proptosis
- Ptosis → Orbital and palpebral lobes involved
  - Lobes separated by levator palpebrae superioris aponeurosis
- Unilateral disease

Lacrimal Lymphoma Presentation

- Unilateral enlarged lacrimal gland
  - Both lobes
- Low T1 and low T2
- Minimal enhancement
- Diffusion restriction → Cellularity
- Cannot differentiate lymphoma types

Lacrimal Lymphoma Findings
Lymphoid Hyperplasia of Lacrimal Gland
Essentially identical appearance to lymphoma.


Closing Points

1. Determine the location of the lesion in question.
2. Know the chronicity of the problem and the presence/absence of pain.
3. Factor in demographics.
4. Knowledge of the “most common” at a given location gets you there most of the time.

Case Studies
In the Orbit

Ethan Neufeld MD
Department of Radiology and Imaging Sciences
University of Utah Health Sciences Center
Salt Lake City, Utah, USA