Slaying the Giant
GCA and the Eye
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Goals

1. Understand Diagnostic Strategies
2. Learn Nuances of GCA
3. Develop Referral Strategy

Diagnosis

Possible
Probable
Prognostic
Pragmatic
Optic Nerve Edema

V - Vascular
O - Ophthalmic
I - Inflammatory
C - Compressive

Optic Nerve Edema

Vascular

AION
NAION

Optic Nerve Edema

Ophthalmic

Drusen
CRVO
Hypotony
Optic Nerve Edema

**Inflammatory**
- Infectious (syphilis, etc)
- Non-infectious (collagen vascular)
- Optic Neuritis

**Compressive**
- Tumor
- Graves' Disease
- Chiasmal

Optic Nerve Edema

1. Possible
2. Probable
3. Prognostic
4. Pragmatic
Optic Nerve Edema

Possible

Probable - based on age

Prognostic

Pragmatic

Probable

Based on Age

40 and younger -- Optic Neuritis

50-60 -- mostly atypical

60+ -- NAION

Optic Nerve Edema

Possible

Probable

Prognostic - GCA

Pragmatic - GCA
What is GCA?

Immune-Mediated Vasculitis

Focal arteritic lesions — Ischemia

Affects medium, large arteries

~18 per 100,000


How does the Temporal Artery connect to the eye?
Why is it so important?

Profound Vision Loss
Bilateral in 14 days in 1/3 if Untreated
Systemic Complications
Treatable

Ophthalmic

Anterior Ischemic Optic Neuropathy (AION)
Central Retinal Artery Occlusion (CRAO)
Amaurosis Fugax
Diplopia

Systemic

- Headache
- Jaw Claudication
- Scalp Tenderness
- Neck Pain
- Anorexia/Weight Loss

A-AION

- Sudden, Painless Vision Loss
- Amaurosis Fugax
- Occurs ≥ 50 years of age
- 1 out of 10
Ocular Symptoms

Vision loss

Amaurosis Fugax

Amaurosis Fugax

From 7% to 50% of patients with GCA

Hayreh found 30%

In sharp contrast to NAION (2.5%)

Transient ischemia to ONH


Systemic Symptoms

Jaw Claudication (Odds Ratio 9.0)
Neck Pain (Odds Ratio 3.4)
Anorexia (Odds Ratio 2)


Less Predictable

Table 3. Incidence of Systemic Signs and Symptoms in Patients with Histologically Confirmed Temporal Arteritis

<table>
<thead>
<tr>
<th>Symptom/Sign</th>
<th>Positive</th>
<th>Negative</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>20/30</td>
<td>60/70</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Fever</td>
<td>15/30</td>
<td>55/70</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Scalp Tenderness</td>
<td>10/20</td>
<td>60/70</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Malaise</td>
<td>10/20</td>
<td>60/70</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Headache

A-AION - 46% had Headache
NA-AION – 54% had Headache

Could Mislead


Average Number of Symptoms = 3
Occult GCA

- Between 5 and 38% of cases
- No systemic symptoms


Contralateral Eye

- Important for 2 reasons
  - Gives us clues about diagnosis
  - Make sure other eye stays healthy

C/D Ratio

- Average C/D in Population = 0.4
- Contralateral C/D in NA-AION
  - 75% are ≤ 0.3
  - 33% are ≤ 0.15
C/D Ratio

A-AION

\[ \leq 0.3 = \frac{50}{725} = \frac{1}{15} \]

\[ \geq 0.4 = \frac{50}{275} = \frac{1}{5} \]

C/D Ratio

Some evidence says:
90% of C/D in NA-AION is \( \leq 0.3 \)

Then…

\[ \leq 0.3 = \frac{50}{860} = \frac{1}{17} \]

\[ \geq 0.4 = \frac{50}{140} = \frac{1}{3} \]
Testing

Labs — ESR, CRP, CBC, Platelets
Fluorescein Angiography
Ultrasound, PET, MRI — Limited Benefit
Temporal Artery Biopsy

FA

Management of ischemic optic neuropathies
Indian Journal Ophthalmol 2011 Vol 59, 2, 123-136

Labs

ESR

≥ 33 mm/h
Sensitivity 92%
Specificity 92%

Labs

CRP

≥ 2.45 mg/dl
Sensitivity 100%
Specificity 82%


Labs

CBC includes

WBC
RBC
Platelets


ESR + CRP

Sensitivity 100%
Specificity 97%
Platelets

Odds Ratio:

ESR $\geq 47$ mm/hr = 1.5
CRP $> 2.45$ mg/dL = 5.3
Platelets $> 400,000/\mu$L = 4.2
All 3 elevated = 8


Additional Testing

Temporal Artery Biopsy
  Gold Standard
  Case by Case
Side Effects
  Necrosis, Infection, Nerve Damage
Bilateral?

Temporal Artery Biopsy

If case is equivocal
ESR + CRP +
ESR + CRP -
ESR - CRP +
ESR - CRP -
Clinical Picture

Unilateral Optic Disc Edema
Age
Systemic Symptoms
Labs (ESR, CRP, Platelets)
Other Eye
Temporal Artery Biopsy

American College of Rheumatology

Need 3 of the following 5
Over 50 years of age
New onset of Headache
Scalp tenderness
ESR > 50 mm/h
(+ ) Temporal Artery Biopsy

American College of Rheumatology

Meeting this criteria yields:

94% Sensitivity
91% Specificity

NOT good enough with swollen optic nerve
Study of ACR Criteria

112 Patients in Neuro-Ophthalmology Clinic
25% with + TAB missed by ACR
28% with - TAB met criteria


Case

62 Year Old Male
Sudden Vision Loss
Unilateral Optic Nerve Edema
What now?

More Information

More details….
Other eye = C/D 0.5
Systemic symptoms = Headache, Neck Pain
Labs: ESR = 70 mm/h
    CRP = 3.2 mg/dl
What If...

More details…
- Other eye = C/D 0.2
- Systemic symptoms = Headache
- Labs: ESR = 20 mm/h
  - CRP = 0.8 mg/dl

Treatment

Oral Steroid
- 80-100+ mg
- VERY long taper
  - 75% reached 5 mg/day at year

IV Steroid no better
Limited Evidence for Immune Modulators
- TNF Blockers
- Methotrexate
Clinical Picture

Unilateral Optic Nerve Edema
Systemic Symptoms
Lab Results
Other Eye

Referral

If AION suspect
Labs
Case History
Possible referral

Referral

Ophthalmology
Rheumatology
Neurologist
Urgent!!!
Giant Cell Arteritis

Affects Eyes

ION, CRAO, Diplopia, Amaurosis Fugax

Affects Systemic

Elevated Labs, Symptoms

Emergency!

MR. A

63 y/o White Male

New onset headache, temporal pain, neck pain

NO vision complaints

EOM full

NO APD
MR.A

Labs

ESR = 56
CRP = 1.37

Pending Temporal Artery Biopsy
MR.A

Started on 40 mg Pred
When he tapers, headaches return
What about vision?
20/70 and VF reduction
MR. A

When Pred resumed, vision returned to 20/25

Visual Field improved
MR. A

Has been on low dose Prednisone for 4 years!

MR. T

63 Year Old White Male
Presents to ER with vision loss OS
Progressed over the day

MR. T

20/20 OD
NLP OS
MR. T

Anterior Segment normal
0.2 C/D OD
Edematous OS

MR. T

Scalp Tenderness
No headache, jaw claudication, neck pain
Intermittent diplopia a month ago

MR. T

ESR = 96 mm/h
CRP = 6.9 mg/dL
MR. T

Admitted and started on IV corticosteroids
Temporal Artery Biopsy +

12 years later on 5 mg/day Prednisone
If tapers off, symptoms start OD

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Thank You!

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