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No Financial Disclosures

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Goals

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

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Diagnosis

1. Possible
2. Probable
3. Prognostic
4. Pragmatic

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Optic Nerve Edema

V - Vascular

O - Ophthalmic

I - Inflammatory

C - Compressive

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Optic Nerve Edema

Vascular

AION
NAION

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Optic Nerve Edema

Ophthalmic

Drusen
CRVO
Hypotony

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Optic Nerve Edema

Inflammatory

Infectious (syphilis, etc)
Non-infectious (collagen vascular)

Optic Neuritis

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Optic Nerve Edema

Compressive

Tumor
Graves' Disease
Chiasmal

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Optic Nerve Edema

1. Possible
2. Probable
3. Prognostic
4. Pragmatic

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Optic Nerve Edema

1. Possible
2. **Probable - based on age**
3. Prognostic
4. Pragmatic

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Probable

Based on Age

40 and younger -- Optic Neuritis

50-60 -- mostly atypical

60+ -- NAION

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Optic Nerve Edema

1. Possible
2. Probable
3. **Prognostic - GCA**
4. **Pragmatic - GCA**

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What is GCA?

Immune-Mediated Vasculitis

Focal arteritic lesions — Ischemia

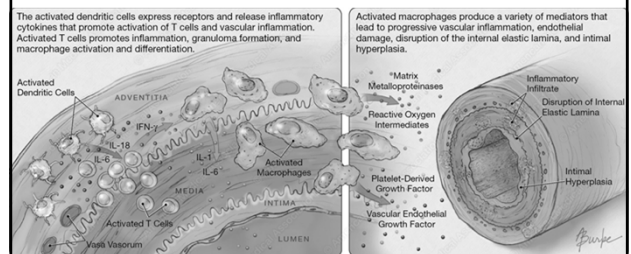
Affects medium, large arteries

~18 per 100,000

Rahman, et al. Giant Cell (Temporal) Ateritis: An Overview and Update. Surv Ophthalmol 50:415-428, 2005

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What is GCA?



<https://goo.gl/images/Tdi5Tr>

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How does the Temporal Artery connect to the eye?

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Why is it so important?

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

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Ophthalmic

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

Hayreh SS, et al. Ocular Manifestations of Giant Cell Arteritis. Am J Ophthalmol 1998;125:509-520

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Systemic

Headache
 Jaw Claudication
 Scalp Tenderness
 Neck Pain
 Anorexia/Weight Loss

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A-AION

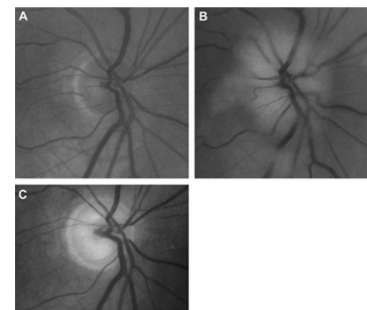
Sudden, Painless Vision Loss
 Amaurosis Fugax
 Occurs \geq 50 years of age
 1 out of 10

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

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

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Ocular Symptoms

Vision loss
Amaurosis Fugax

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Amaurosis Fugax

From 7% to 50% of patients with GCA
Hayreh found 30%
In sharp contrast to NAION (2.5%)
Transient ischemia to ONH

Hayreh SS, et al. Amaurosis Fugax in Ocular Vascular Occlusive Disorders. Retina 2013;0:1-8
Hayreh SS, et al. Ocular Manifestations of Giant Cell Arteritis. Am J Ophthalmol 1998;125:509-520

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Systemic Symptoms

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

Hayreh SS, et al. Giant Cell Arteritis: Validity and Reliability of Various Diagnostic Criteria. Am J Ophthalmol 1997;123:285-96

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Less Predictable

Headache
Fever
Scalp Tenderness
Malaise

Hayreh SS, et al. Giant Cell Arteritis: Validity and Reliability of Various Diagnostic Criteria. Am J Ophthalmol 1997;123:285-96

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V | vwhp If#M | p swp v

Table 3. Incidence of Systemic Signs and Symptoms in Patients With Positive and Negative Temporal Artery Biopsy for Giant Cell Arteritis

Signs and Symptoms	Temporal Artery Biopsy (No. [%])		P Value
	Positive (n = 106)	Negative (n = 257)	
Headache	59 (55.7)	117 (45.5)	.084
Anorexia/weight loss	55 (51.9)	84 (32.7)	.0005
Jaw claudication	51 (48.1)	22 (8.6)	<.0001
Malaise	40 (37.7)	78 (30.4)	.177
Myalgia	31 (29.2)	68 (26.5)	.606
Fever	23 (21.4)	42 (16.3)	.040
Abnormal temporal artery	21 (19.8)	33 (12.8)	.105
Scalp tenderness	19 (17.9)	27 (10.5)	.058
Neck pain	17 (16.0)	11 (4.3)	.0003
Anemia	14 (13.2)	31 (12.1)	.730

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Headache

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

Could Mislead

Hayreh SS, et al. Giant Cell Arteritis: Validity and Reliability of Various Diagnostic Criteria. Am J Ophthalmol 1997;123:285-96

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Average Number of Symptoms = 3

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Occult GCA

Between 5 and 38% of cases

No systemic symptoms

Rahman, et al. Giant Cell (Temporal) Arteritis: An Overview and Update. Surv Ophthalmol 50:415--428, 2005

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Contralateral Eye

Important for 2 reasons

1. Gives us clues about diagnosis
2. Make sure other eye stays healthy

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C/D Ratio

Average C/D in Population = 0.4

Contralateral C/D in NA-AION

- 75% are ≤ 0.3
- 33% are ≤ 0.15

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C/D Ratio

A-AION

- $\leq 0.3 = 50/725 = 1/15$
- $\geq 0.4 = 50/275 = 1/5$

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C/D Ratio

Some evidence says:
90% of C/D in NA-AION is ≤ 0.3

Then...
 $\leq 0.3 = 50/860 = 1/17$

$\geq 0.4 = 50/140 = 1/3$

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Testing

Labs – ESR, CRP, CBC, Platelets

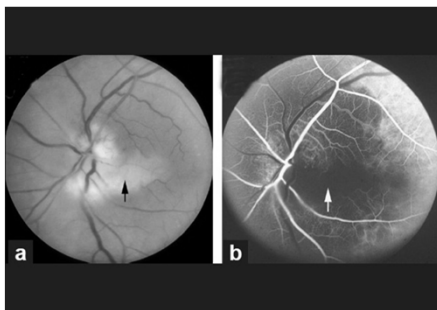
Fluorescein Angiography

Ultrasound, PET, MRI – Limited Benefit

Temporal Artery Biopsy

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FA



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

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Labs

ESR
CRP
CBC



Hayreh SS, et al. Giant Cell Arteritis: Validity and Reliability of Various Diagnostic Criteria. Am J Ophthalmol 1997;123:285-96

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Labs

ESR

- ≥ 33 mm/h
- Sensitivity 92%
- Specificity 92%

Hayreh SS, et al. Giant Cell Arteritis: Validity and Reliability of Various Diagnostic Criteria. Am J Ophthalmol 1997;123:285-96

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Labs

CRP

- ≥ 2.45 mg/dl
- Sensitivity 100%
- Specificity 82%

Hayreh SS, et al. Giant Cell Arteritis: Validity and Reliability of Various Diagnostic Criteria. Am J Ophthalmol 1997;123:285-96

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Labs

CBC includes

WBC
RBC
Platelets

Hayreh SS, et al. Giant Cell Arteritis: Validity and Reliability of Various Diagnostic Criteria. Am J Ophthalmol 1997;123:285-96

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ESR + CRP

Sensitivity 100%

Specificity 97%

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Platelets

Odds Ratio:

ESR \geq 47 mm/hr = 1.5

CRP > 2.45 mg/dL = 5.3

Platelets > 400,000/ μ L = 4.2

All 3 elevated = 8

Walvick MD, Walvick MP. Giant Cell Arteritis: Laboratory Predictors of a Positive Temporal Artery Biopsy. Ophthalmology 2011;118:1201-1204

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Additional Testing

Temporal Artery Biopsy

Gold Standard

Case by Case

Side Effects

Necrosis, Infection, Nerve Damage

Bilateral?

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Temporal Artery Biopsy

If case is equivocal

ESR + CRP +

ESR + CRP -

ESR - CRP +

ESR - CRP -

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Clinical Picture

Unilateral Optic Disc Edema

Age

Systemic Symptoms

Labs (ESR, CRP, Platelets)

Other Eye

Temporal Artery Biopsy

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American College of Rheumatology

Need 3 of the following 5

1. Over 50 years of age
2. New onset of Headache
3. Scalp tenderness
4. ESR > 50 mm/h
5. (+) Temporal Artery Biopsy

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American College of Rheumatology

Meeting this criteria yields:

94% Sensitivity

91% Specificity

NOT good enough with swollen optic nerve

50

Study of ACR Criteria

112 Patients in Neuro-Ophthalmology Clinic

25% with + TAB missed by ACR

28% with - TAB met criteria

Murchison, AP, et al. Am J Ophthalmol 2012;154:722-729

51

Case

62 Year Old Male

Sudden Vision Loss

Unilateral Optic Nerve Edema

What now?

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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

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What If...

More details....

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

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Treatment

Oral Steroid
80-100+ mg
VERY long taper



75% reached 5 mg/day at year

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Treatment

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

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Treatment

Actemra



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Study

4 Groups

Actemra weekly vs bi-weekly (with 26 week pred taper)

Placebo (with 26 week pred taper)

Placebo (with 52 week pred taper)

1 - Stone, J H, et al. Trial of Tocilizumab in Giant Cell Arteritis. N Engl J Med 2017; 377:317-328

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Sustained Remission at 52 weeks

Actemra weekly	56%
Actemra bi-weekly	53%
Placebo 26 week	14%
Placebo 52 week	18%

59

Cumulative Dose of Prednisone

Actemra weekly	1862 mg
Actemra bi-weekly	1862 mg
Placebo 26 week	3296 mg
Placebo 52 week	3818 mg

60

Many adverse reactions

Mostly non-serious (like injection site reaction)

More serious reaction in prednisone alone

25% vs 15%

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Clinical Picture

Unilateral Optic Nerve Edema

Systemic Symptoms

Lab Results

Other Eye

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Referral

If AION suspect

Labs

Case History

Possible referral

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Referral

Ophthalmology

Rheumatology

Neurologist

Urgent!!!

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Giant Cell Arteritis

Affects Eyes

ION, CRAO, Diplopia, Amaurosis Fugax

Affects Systemic

Elevated Labs, Symptoms

Emergency!

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MR. A

63 y/o White Male

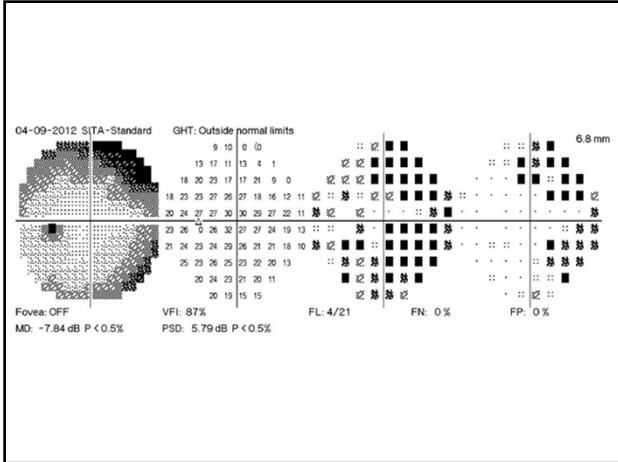
New onset headache, temporal pain, neck pain

NO vision complaints

EOM full

NO APD

66



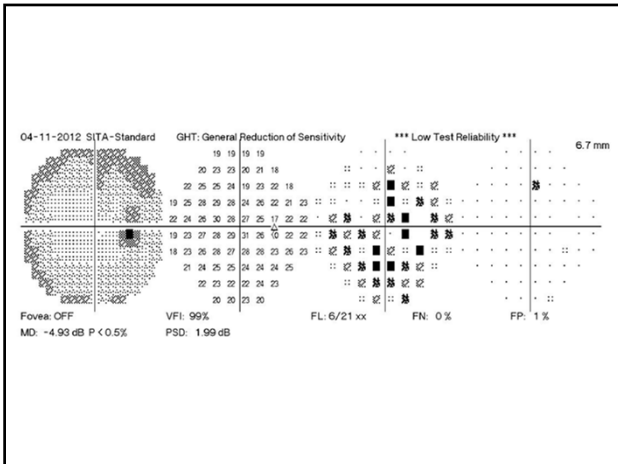
73

MR. A

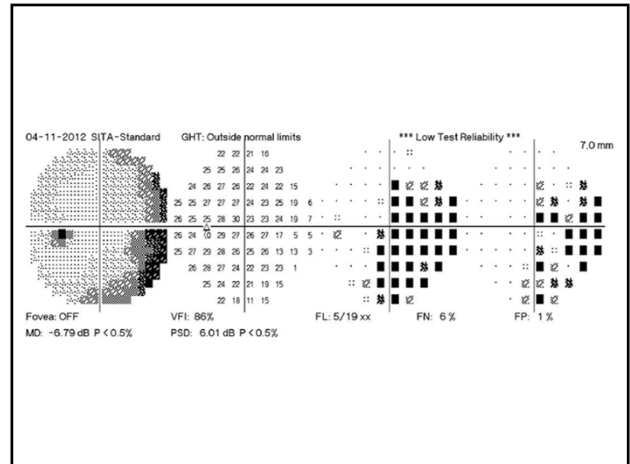
When Pred resumed, vision returned to 20/25

Visual Field improved

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MR. A

Has been on low dose Prednisone for 4 years!

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MR. T

63 Year Old White Male

Presents to ER with vision loss OS

Progressed over the day

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MR. T

20/20 OD

NLP OS

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MR. T

Anterior Segment normal

0.2 C/D OD

Edematous OS

80

MR. T

Scalp Tenderness

NO headache, jaw claudication, neck pain

Intermittent diplopia a month ago

81

MR. T

ESR = 96 mm/h

CRP = 6.9 mg/dL

82

MR. T

Admitted and started on IV corticosteroids

Temporal Artery Biopsy +

83

MR. T

12 years later on 5 mg/day Prednisone

If tapers off, symptoms start OD

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Optic Nerve Edema

1. Possible
2. Probable
3. Prognostic
4. Pragmatic

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Goals

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

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Thank You!

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