COMMON ACUTE VISION LOSS OFFENDERS

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GIANT CELL ARTERITIS

- Inflammation of the lining of the arteries
- GCA typically occurs in older adults, usually those over the age of 50
- GCA affects blood flow to the eyes and can lead to vision loss
- Usually associated with polymyalgia rheumatica

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PATHOPHYSIOLOGY

- Unknown exact cause
- Diseases of cell-mediated immunity
- T-Cell mediated immune response to an unrecognized vessel wall protein

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PATHOPHYSIOLOGY

- Most common ocular sign of GCA is vision loss and secondary to arteritic anterior ischemic optic neuropathy (AAION)
- Non-arteritic ischemic optic neuropathy (NAION) is usually not associated with GCA but is a similar phenomenon
- Biopsies have confirmed AAION secondary to GCA with segmental disk edema without pallor, mimicking NAION
- GCA can impact visual pathway starting from retina all the way to the occipital lobe

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PATHOPHYSIOLOGY

- Less common causes of ischemic visual loss
  - Posterior ischemic optic neuropathy
  - Cilioretinal artery occlusion
  - Choroidal infarction
- Cortical visual loss may lead to visual hallucinations
- Patients often experience headaches and have scalp tenderness

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ETIOLOGY

- Genetic predisposition has been suspected for GCA
- Infectious factors could trigger immune response
- Linked genes related to cytokine and chemokine expression → alters clinical presentation in different patients
- GCA affects three-layered vessels (outer adventitia, muscular medial layer, and elastic lamina)

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EPIDEMIOLOGY

- GCA is most common in those who are 50 years or older
- Prevalence of GCA depends on the number of people who are 50 years or older; mean age of onset is 75 years
- Study shows that GCA incidence is highest in Scandinavia
- Risk factors include smoking, low BMI, early menopause

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EPIDEMIOLOGY

- Age is most important risk factor
- GCA is most common systemic vasculitis affecting elderly patients
- Results of studies have shown that GCA primarily affects whites, specifically those of European descent

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PHYSICAL EXAMINATION

- Ophthalmologic examination that evaluates visual acuity and pupils
- Visual fields tests
- Auscultation of carotid artery
- Auscultation of heart for aortic regurgitation murmur

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AAION

- Acute monocular vision loss with optic disc edema
- Optic disk edema may be accompanied by retinal whitening, disc hemorrhage, cotton wool spots
  - Retinal whitening and cotton wool spots raise suspicions for GCA and indicate concurrent retinal ischemia
- In posterior ischemic optic neuropathy, patients have similar symptoms to those of AAION
  - Posterior ischemic optic neuropathy is a much less common cause of GCA
  - Big difference is that optic disc appears normal

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CLINICAL PRESENTATION

- Headache (usually localized to temporal/occipital area)
- Fatigue
- Jaw claudication
- Systemic manifestations of malaise
- Weight loss
- Fever
- Night sweats
- Depression
- Proximal muscle pain/stiffness due to polymyalgia rheumatica
- Pain/tenderness due to local inflammation
- Unilateral visual blurring/vision loss
- Hallucinations preceding to permanent vision loss

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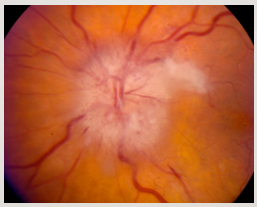

Patient with arteritic ischemic optic neuropathy with pallid disk edema, hemorrhages, and cotton wool spot

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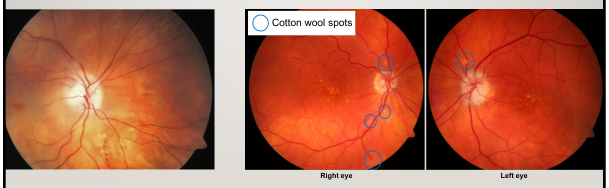
EYE PRESENTATION



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CLINICAL DIAGNOSIS

- Diagnosis should be considered in patients over age of 50 with new headaches, visual changes, symptoms of polymyalgia rheumatica, and jaw claudication
- Inhiation of corticosteroid treatment is recommended for patients where GCA is suspected based off elevated ESR, C-reactive protein/thrombocytosis, temporal artery biopsy, or other findings

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DIAGNOSIS

- Blood tests
  - Erythrocyte sedimentation rate
  - C-reactive protein
- Imaging Tests
  - Doppler ultrasound
  - MRI
  - PET scan: this technique is combined with computed tomography (PET-CT)

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TEMPORAL ARTERY BIOPSY

- "Diagnostic gold standard"
- Superficial temporal artery biopsy (TAB) is a standard for diagnosing GCA
- TAB is crucial and necessary for any patient who is suspected of having GCA
  - Important due to the long treatment for GCA and the complications that arise
- Positive TAB = 100% specificity but low sensitivity. (15%-87%) for diagnosing GCA

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TEMPORAL ARTERY BIOPSY

- Negative biopsy does not confirm a negative diagnosis
- False negatives are common (5%-13%) due to "skip lesions" (areas without disease within vessels)
  - Biopsy samples are recommended to be 1 cm-2.5 cm in length
- Temporal artery biopsy: positive TAB = 100% specificity, low sensitivity = 87%

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ANCILLARY IMAGING TESTS

- Color Doppler Ultrasound (CDUS)
  - Non-invasive and safe
  - Halo sign
  - Compression sign
- Magnetic Resonance Imaging (MRI)
  - Blood vessel wall thickening/enhancement
- Ultrasound biomicroscopy (UBM)
  - Detect the halo sign
- CT & PET
  - Evaluate aorta and other large vessels

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LABORATORY TESTING

- Erythrocyte Sedimentation Rate (ESR): measures how fast red blood cells falls to a the bottom of a test tube of blood; inflammation indicated by red cells that drop rapidly
- C-reactive protein (CRP)
- CRP exceeds 50 mm/h
- ESR exceeds 100 mm/h

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TREATMENT

- High doses of corticosteroid drug (prednisone)
  - Should be started immediately after suspicions of GCA are confirmed
  - Higher doses should be given to higher risk patients
  - Suggested dose ranges from 40 mg-100 mg per day
  - Dose should be increased to 80 mg- 100 mg if GCA is confirmed with biopsy
- Calcium and vitamin D supplements may be prescribed to counter the effects of the corticosteroids

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TAKEAWAY

- Mainly affects arteries in head
- Leads to blindness if left untreated
- 50% of people with GCA tend to have polymyalgia rheumatica

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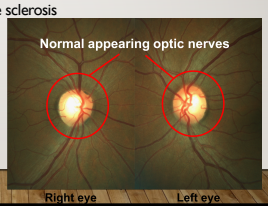
OPTIC NEURITIS

- Inflammation that damages optic nerve
- Can be the first indication of multiple sclerosis



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PATHOPHYSIOLOGY

- Inflammatory demyelination of optic nerve
- Pathology is like multiple sclerosis plaques in brain
- Demyelination is immune mediated
- Systemic T cell activation is recognized at symptom onset
- T cell activation leads to the release of cytokines and inflammatory agents
- B cell activation against myelin basic protein can be shown in CSF of patients diagnosed with optic neuritis

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EPIDEMIOLOGY

- Two-thirds of cases occur in women
- Patients are typically between the ages of 20 and 40
- Highest in populations located at higher latitudes (northern United States and western Europe, New Zealand)
- Annual incidence of ON is as estimated to be as high as 6.4 per 100,000

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GENERAL PATHOLOGY

- Immune-mediated inflammatory demyelination of optic nerve
- Myelin gets destructed which causes axons poorly conduct impulses → leads to axons being damaged
- Retinal ganglion cell axons usually become damaged in optic neuritis

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ETIOLOGY

- Idiopathic
- Demyelinating diseases
  - Multiple Sclerosis – immune system attacks protective covering of nerves
  - Neuromyelitis optics (Devic's Disease) – inflammation and demyelination of the central nervous system, mainly of spinal cord and optic nerve
  - Schilder's Disease – rare progressive demyelinating disorder usually beginning at childhood
  - Acute disseminated encephalomyelitis (ADEM) – attack of inflammation in brain and spinal cord which damages myelin

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ETIOLOGY CONT.

- Infectious Diseases
  - Ethmoiditis: leads to pressure/pain between eyes
  - Lyme Disease
  - Syphilis
- Viral Infections
  - Measles and mumps
- Autoimmune Diseases
  - Sarcoidosis
  - Lupus
  - Neuromyelitis optics

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CLINICAL TYPES

- Papillitis – deterioration and inflammation of optic disk
  - Individuals may experience vision loss in one eye which may occur within many hours of onset
- Retrobulbar neuritis - optic nerve (back of eye) becomes inflamed
  - Visual signaling to brain is interrupted and there are visual impairments
- Neuroretinitis – inflammation of neural retina and optic nerve

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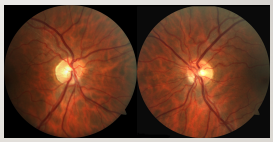
CLINICAL PRESENTATION

- Pain in and around eye
- Loss of vision in one eye
- Visual field loss
- Loss of color vision (red/dull/faded colors)
- Flashing lights



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SYMPTOMS

- Prodromal viral illness (could be present)
- Phosphenes may occur
- "Washed out" color vision
- Uhthoff and Pulfrich phenomenon

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UHTHOFF'S PHENOMENON

- Worsening of multiple sclerosis symptoms which leads to an increase in temperature
  - Elevated temperature leads to visual impairments
  - Occurs due to damage to the optic nerve

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PULFRICH PHENOMENON

- Perception that an object is moving linearly along 2D plane appears to follows an elliptical 3D trajectory
- Found to occur in patients with ocular/neurological conditions where the visual pathway has been affected

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SIGNS

- Decreased visual acuity
- RAPD unless both eyes are affected
- Efferent lesions may be present (ocular dysmetria or internuclear ophthalmoplegia)
- Retinal vascular sheathing (periphlebitis occurs in roughly 5%-10% of patients with MS)

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DIAGNOSIS

- MRI
- Blood tests – check for specific antibodies/infections
- Optical Coherence Tomography (OCT) – measures thickness of eye's retinal nerve fiber layer
- Visual Evoked Response
- Chest X-Ray

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CLINICAL DIAGNOSIS

- Diagnosed by symptoms of acute unilateral decrease in vision, eye pain, RAPD, decrease in color vision/contrast/brightness
- Patients could have severe optic disc swelling with hemorrhages/retinal exudates, but this is less common
- In this case, an MRI indicates the presence/future development of MS
  - Patients with acute ON who had gadolinium-enhanced fat-suppressed cranial MRI scans within 20 days of visual loss had an enhancement of the orbital optic nerve
  - This was seen in 94% of the observed patients

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DIAGNOSTIC PROCEDURE

- Optical coherence tomography (OCT)
  - Helpful measurement of nerve function
  - Can quantify the onset of optic disc pallor
  - OCT is useful for detection/quantification of optic atrophy

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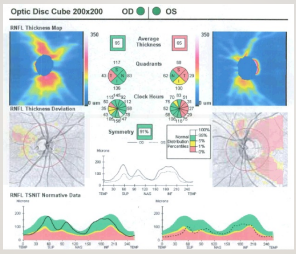

OCT image from patient's right eye 2.5 months after and left eye 3.5 months after

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ADDITIONAL LABORATORY TESTS

- Further blood tests for NMOSD, MOG and other infectious/inflammatory diseases may be considered
- If vision loss has occurred in young male with a family history of maternally-related males with bilateral vision loss, then genetic counseling and testing would be considered for Leber hereditary optic neuropath (LHON)
  - Patients with LHON are less likely to recover their vision

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PHYSICAL EXAMINATION

- Color vision/contrast measurement (Pelli-Robson/VisTech)
- Swinging flashlight test for RAPD
- Evaluation of extraocular movement
- Biomicroscopy/direct ophthalmoscopy of optic nerve and retina

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DIFFERENTIAL DIAGNOSIS OF RETROBULBAR OPTIC NEURITIS

- With Relative Afferent Pupillary Defect
  - Compressive lesions like meningioma
  - Posterior ischemic optic neuropathy: acute optic neuropathy due to ischemia in retrobulbar portion of optic nerve
  - Paracentral acute middle maculopathy: optical coherence tomography finding in patients with retinal capillary ischemia and persistent scotomas
  - Central serous chorioretinopathy: fluid builds up under retina which can distort vision

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DIFFERENTIAL DIAGNOSIS OF RETROBULBAR OPTIC NEURITIS

- No Relative Afferent Pupillary Defect
  - Visual field defects from lesions beyond lateral geniculate body
  - Retinal degeneration (retinitis pigmentosa)
  - Macular disease
    - Age-related macular degeneration
    - Macular edema (post-cataract surgery, diabetic)
    - Macular hole (traumatic, idiopathic)
    - Acute macular neuroretinopathy (AMNR)

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DIFFERENTIAL DIAGNOSIS OF OPTIC NEURITIS (UNILATERAL OPTIC DISC EDEMA)

- NMOSD (Devic disease)
- MOG immunoglobulin G-associated disorder
- Anterior ischemic optic neuropathy (usually painless)
- Neuroretinitis
- Chronic relapsing inflammatory optic neuropathy (painful)
- Pending central retinal vein occlusion (painless)
- Diabetic papillopathy (painless)

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DIFFERENTIAL DIAGNOSIS OF OPTIC NEURITIS (UNILATERAL OPTIC DISC EDEMA)

- Vasculitis
- Malignant hypertension
- Leber hereditary optic neuropathy
- Radiation induced optic neuropathy
- Multiple evanescent white dot syndrome

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MEDICAL THERAPY

- Low dose (60 mg/day) oral steroids are contradicted in acute ON patients
- Neuro-opthalmologists are using a methylprednisolone smoothie
  - Dilute the powder in a smoothie drink
  - Patients drink one dose a day for 3 days
- Repository corticotropin (intramuscular/subcutaneous) injection at 80 units/1 ml has also been used to treat acute ON

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TREATMENT

- High-dose steroid drugs through an IV
- Intravenous Immune Globulin (IVIG) – plasma exchange
- Vitamin B12 shots

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MEDICAL FOLLOW-UP

- Visual acuity testing
- Fluorescein angiogram can rule out optic nerve edema if LHON was suspected
- Visual field testing at 3, 6, and 12 months if possible

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COMPLICATIONS

- Visual prognosis is excellent
- Decreased brightness sense, contrast deficit, and loss of stereopsis may exist
- Possibility of
  - Permanent visual loss (20/30) to (20/200)
  - Permanent scotomas = limiting driving
  - Recurrences

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COMPLICATIONS

- IV steroids can lead to insomnia, mood changes, dyspepsia, weight gain, vomiting, and spiked blood pressure
- Patients tend to have more side effects from oral prednisone taper

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PROGNOSIS

- 94% of patients recover vision to 20/40 or better
- Visual recovery occurs usually at 1 month after onset
- Continuous pain with eye movement, lack of recovery, recurrence would lead to re-evaluation for causes of ON

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TAKEAWAYS

- Optic Neuritis is usually the first indication of multiple sclerosis
- Common with infections/immune diseases
- Most regain vision within 6 months after ON episode
- Some may have permanent optic nerve damage after ON episode and there may be decreased visual acuity
- MRI scan of brain if ON is suspected

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WORKS CITED

- "Giant Cell Arteritis." Edited by Marcy Bolster, *Rheumatology.org*, www.rheumatology.org/I-Am-A/Patient-Caregiver/Diseases-Conditions/Giant-Cell-Arteritis.
- Li, Katherine J., et al. "A Meta-Analysis of the Epidemiology of Giant Cell Arteritis across Time and Space." *Arthritis Research & Therapy*, BioMed Central, 11 Mar. 2021, arthritis-research.biomedcentral.com/articles/10.1186/s13075-021-02450-w.
- Mythili Seetharaman, MD. "Giant Cell Arteritis (Temporal Arteritis)." *Practice Essentials, Pathophysiology, Etiology*, Medscape, 29 July 2021, emedicine.medscape.com/article/332483-overview#a5.
- JG;, Jones. "Clinical Features of Giant Cell Arteritis." *Baillière's Clinical Rheumatology*, U.S. National Library of Medicine, pubmed.ncbi.nlm.nih.gov/1807819/.
- Mythili Seetharaman, MD. "Giant Cell Arteritis (TEMPORAL Arteritis) WORKUP: Approach Considerations, Temporal Artery BIOPSY, ULTRASONOGRAPHY." *Giant Cell Arteritis (Temporal Arteritis) Workup: Approach Considerations, Temporal Artery Biopsy, Ultrasonography*, Medscape, 29 July 2021, emedicine.medscape.com/article/332483-workup#c1.
- Osborne, Benjamin, and Laura J Balcer. "Optic Neuritis: Pathophysiology, Clinical Features, and Diagnosis." *Uptodate*, 17 July 2018, www.uptodate.com/contents/optic-neuritis-pathophysiology-clinical-features-and-diagnosis.
- Pau, D, et al. "Optic Neuritis." *Eye (London, England)*, Nature Publishing Group, July 2011, www.ncbi.nlm.nih.gov/pmc/articles/PMC3178158/.

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WORKS CITED

- "Optic Neuritis." *Mayo Clinic*, Mayo Foundation for Medical Education and Research, 12 Feb. 2020, www.mayoclinic.org/diseases-conditions/optic-neuritis/symptoms-causes/syc-20354953.
- "Papillitis." *NORD (National Organization for Rare Disorders)*, rarediseases.org/rare-diseases/papillitis/.
- "Retrobulbar Neuritis Guide: Causes, Symptoms and Treatment Options." *Drugs.com*, 12 Jan. 2021, www.drugs.com/health-guide/retrobulbar-neuritis.html.
- "Optic Neuritis." *Mayo Clinic*, Mayo Foundation for Medical Education and Research, 12 Feb. 2020, www.mayoclinic.org/diseases-conditions/optic-neuritis/diagnosis-treatment/drc-20354958.
- Rath, Linda. "Optic Neuritis: Symptoms, CAUSES, Tests, DIAGNOSIS, TREATMENT." *WebMD*, WebMD, www.webmd.com/multiple-sclerosis/optic-neuritis-ms-vision.
- "Demyelinating Optic Neuritis." *EyeWiki*, 12 Mar. 2021, eyewiki.aao.org/Demyelinating_Optic_Neuritis#Etiology.
- "Giant Cell Arteritis." *EyeWiki*, 1 Feb. 2021, eyewiki.org/Giant_Cell_Arteritis.

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