



OPHTHALMOLOGY
ON CALL

January 2023

George Washington University

Department of Ophthalmology

Table of Contents

Introduction.....	Page 3
Optic Neuritis.....	Page 4
Branched Retinal Artery Occlusion (BRAO).....	Page 10
Acknowledgements.....	Page 15

Introduction

GW Ophthalmology On Call is a monthly case report publication presented by medical students and residents at the GW Department of Ophthalmology. The purpose of this publication series is to highlight educational content seen at the GW Department of Ophthalmology. This content is for teaching purposes only and should not be used to guide treatment. All case presentations were written in accordance with HIPAA guidelines. Case details have been modified for patient privacy purposes.

Left Eye Vision Loss

Chief Complaint

Vision loss in the left eye

History of Present Illness

A 28-year-old female presented to the emergency department with 2 weeks of painless, decreased vision of the left eye. Additionally, she reported a left sided headache for the first week and endorsed discomfort with left eye movement. She denied head trauma, fever, paresthesias, weakness, changes in hearing, tinnitus, pain when chewing, watery eyes, eye redness, or recent illness. She recalled a similar episode of vision loss in the left eye that resolved in 3 weeks with no intervention 4 months ago.

Ocular History

None

Past Medical History

G6PD deficiency

Medications

None

Allergies

No known drug allergies

Family History

None

Social History

Everyday e-cig user

Review of Systems

Noted in HPI, otherwise negative

Ocular Exam

Physical Exam		
	OD	OS
Visual Acuity	20/20	Count Fingers
Intraocular Pressure (mmHg)	13	12
Pupils	Round and reactive	2+ APD
Extraocular Motility	EOMI	EOMI

Slit Lamp Exam		
	OD	OS
External	Unremarkable	Unremarkable
Lids and Lashes	Normal, no lesions	Normal, no lesions
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	Clear	Clear
Anterior chamber	Deep and quiet	Deep and quiet
Iris	Round and reactive	Round and reactive
Lens	Clear	Clear

Dilated Fundus Exam		
	OD	OS
Cup to Disc	0.3	0.3
Optic Nerve	Pink/sharp	Pink/sharp
Vessels	Normal caliber and course	Normal caliber and course
Macula	Normal	Normal
Periphery	Flat	Flat
Vitreous	Clear	Clear

Imaging

MRI Face/neck/orbit with and without contrast

- Abnormal appearance of the left optic nerve including prominent asymmetric increased T2 signal throughout the left optic nerve which appears most pronounced intracranially where there is also mild enlargement and prominent enhancement of the nerve which extends slightly into the left side of the optic chiasm. There is lesser enhancement of the orbital portion of the left optic nerve without apparent enlargement. These findings are indicative of acute optic neuritis. Normal MRI examination of the right optic nerve performed with and without contrast.

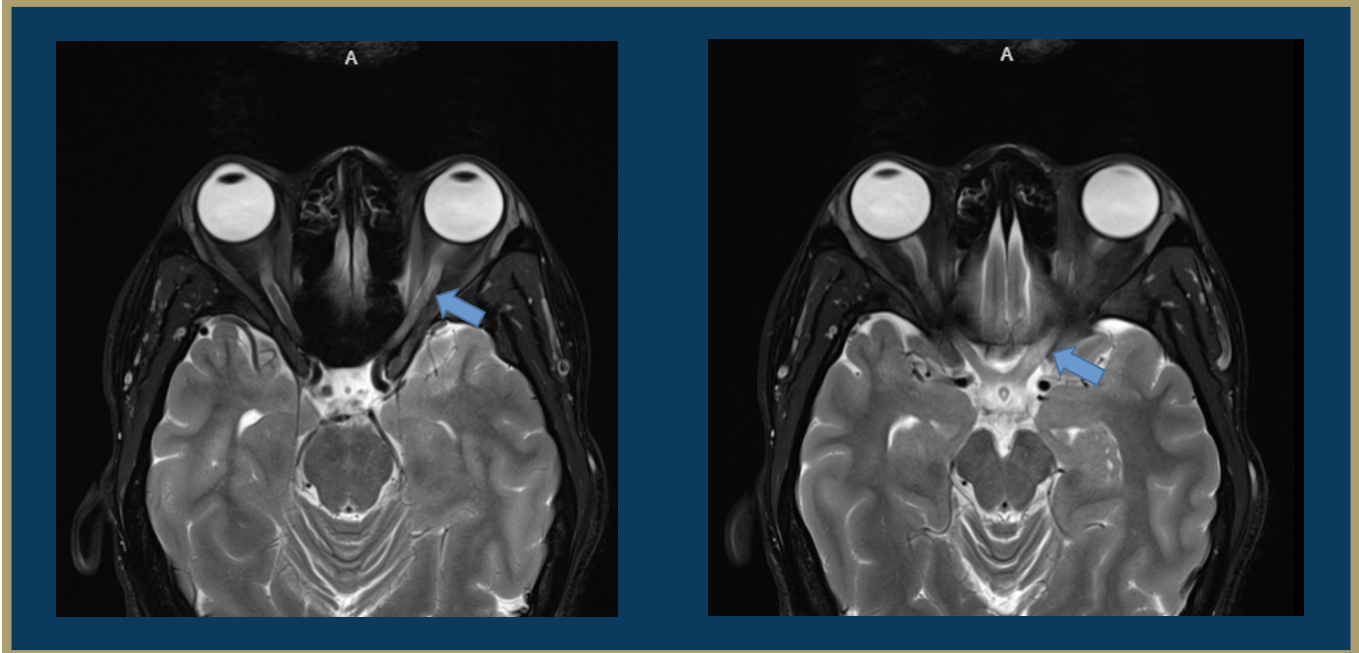


Figure 1: MRI Orbit showing asymmetric increased T2 signal throughout the left optic nerve.

MRI Brain with and without contrast

- There is a single high T2 signal intensity lesion in the left precentral gyrus suspicious for demyelinating disease given its appearance and location, particularly given the patient's clinical presentation and apparent left optic neuritis on the concurrent MRI examination of the orbits. The remaining brain is within normal limits.

MRI MRV Head without contrast

- Normal MR venogram of the brain. Specifically, there is no evidence of dural venous sinus thrombosis.

MRI Cervical Spine and Thoracic with and without contrast

- Essentially normal MRI of the cervical and thoracic spine.

Test Results

- ESR, CRP, IgG, HIV negative/unremarkable
- ANA- negative
- **CSF Oligoclonal bands- positive (type 2)**
- CSF ACE- negative
- NMO AQP4-IgG- negative
- MOG-IgG- negative
- LP with cell count 4WBC (89% lymphs), glucose: 89, IgG: 1,232.

- CSF with cx: mild pleocytosis, normal protein, no polymorphonuclear lymphocytes
- CSF IgG index 1.6 (high), IgG synthesis rate 4.2 (high)
- Meningoencephalitis panel- negative

Differential Diagnosis

- Clinically isolated syndrome (CIS) of optic neuritis (ON)
- CIS of ON prodromal of multiple sclerosis (MS)
- neuromyelitis optica spectrum disorder
- MAG
- Lupus

Clinical Course

The patient is a 26 year old female with a past medical history of G6PD deficiency who was admitted for recurrent subacute left-sided visual loss with APD and contrast-enhancing left optic nerve on imaging, concerning for optic neuritis. MRI of the brain showed central vein sign consistent with demyelinating disease. The patient received a 3-day course of SoluMedrol with improvement in vision and was discharged with a steroid taper. The patient followed up with the neurology outpatient clinic after discharge. Oligoclonal IgG bands (type 2) were positive and the patient was diagnosed with Multiple Sclerosis (MS).

Discussion

Optic neuritis is an acute, inflammatory condition of the optic nerve associated with asymmetric vision loss and eye pain. The etiology of optic neuritis, while often idiopathic, can be secondary to demyelination, self-directed antibodies, autoimmune inflammation, infections, and post-infectious/post-vaccination. Central to the pathophysiology are T and B cell lymphocytes that initiate a faulty immune response by recognizing neural autoantigens as foreign proteins. The resulting inflammatory response leads to demyelination, axonal loss, and the subsequent ocular symptomatology.

Optic neuritis classically presents in a young female (age 20-40) with symptoms of unilateral vision loss, pain with eye movement, a deficiency in color vision, and a history of a prior episode that had spontaneously improved. Optic Neuritis is a clinical diagnosis ascertained by physical exam findings such as decreased Snellen visual acuity, decreased color vision on Pelli-Robson or Vistech test, RAPD on swinging flashlight test, pain on extraocular motility exam, nerve fiber layer or central loss shown on visual field exam, and optic disc edema on dilated fundus exam (present in up to 35% of patients). The presentation of this patient generally supports the body of literature that describes epidemiology and symptomatology commonly associated with optic neuritis. A patient of female sex in her early 20s presenting with recurrent neurologic deficits and unilateral, red-denatured paints the clinical picture (Toosy et al, 2014).

For diagnostics, MRI of the brain and Orbit can help elucidate the etiology of optic neuritis. In typical Optic Neuritis, MS shows on MRI as a segment of contrast enhancement of the optic nerve and a high intensity T2 weighted lesion white matter lesions located in the periventricular, juxtacortical, infratentorial, and/or spinal cord white matter. In non-typical ON,

white matter lesions may appear outside of typical locations and optic nerve enhancement is more posterior in the optic chiasm and tract. This patient underwent an MRI of the brain and orbit to validate suspicion of a CNS process that might have affected white matter structures. Asymmetric increase of T2 signal throughout the left optic nerve supports suspicion of local inflammation.

Additional testing includes CSF analysis for oligoclonal bands to confidently diagnose MS. In this patient, the oligoclonal bands detected on electrophoresis and the recurrent nature further support MS, a commonly observed etiology of optic neuritis. For non MS etiologies, VDRL and FTA-ABS can test for syphilis, antibody titers can test for Lyme disease, chest x-ray and serum angiotensin-converting enzyme levels test for sarcoidosis, and ANA for Systemic lupus erythematosus and vasculitis.

The clinical evidence in this case indicates an inflammatory process, thus initial treatment of methylprednisolone was prescribed. Treatment is guided by the The Optic Neuritis Treatment Trial which evaluated the efficacy and safety of an oral placebo, oral prednisone, and IV methylprednisolone followed by oral prednisolone in treating acute ON. The 1000 mg IV methylprednisolone daily for 3 days and 11 day oral prednisone taper was found to accelerate visual recovery fastest in the first 4-6 weeks post onset but did not improve long-term outcomes. Additionally, patients who took oral steroids in conventional doses alone were found to have higher rates of recurrence of ON and are thus contraindicated in acute ON.

Overall, Optic neuritis is a complicated pathology with several etiologies and diagnostic algorithms. While multiple sclerosis is the most common typical etiology for optic neuritis, immediate treatment is the same for most causes followed by further investigation to treat the underlying cause. In this case, the patient's clinical presentation and MRI and lab results lead towards the diagnosis and treatment of Multiple Sclerosis.

References

Beck RW, Cleary PA, Anderson MM, et al. A randomized, controlled trial of corticosteroids in the treatment of acute optic neuritis. The Optic Neuritis Study Group. *N Engl J Med*. 1992;326(9):581-588. doi:10.1056/NEJM199202273260901

Chen JJ, Pittock SJ, Flanagan EP, Lennon VA, Bhatti MT. Optic neuritis in the era of biomarkers. *Surv Ophthalmol*. 2020;65(1):12-17. doi:10.1016/j.survophthal.2019.08.001

Horton JC. Disorders of the Eye. In: Jameson JL, Fauci AS, Kasper DL, Hauser SL, Longo DL, Loscalzo J, eds. *Harrison's Principles of Internal Medicine*. 20th ed. McGraw-Hill Education; 2018. Accessed January 7, 2023.
accessmedicine.mhmedical.com/content.aspx?aid=1187230393

Papadakis MA, McPhee SJ, Bernstein J. Optic Neuritis. In: *Quick Medical Diagnosis & Treatment 2022*. McGraw-Hill Education; 2022. Accessed January 7, 2023.
accessmedicine.mhmedical.com/content.aspx?aid=1186004205

Toosy AT, Mason DF, Miller DH. Optic neuritis. *Lancet Neurol*. 2014;13(1):83-99. doi:10.1016/S1474-4422(13)70259-X

Wilhelm H, Schabet M. The Diagnosis and Treatment of Optic Neuritis. *Dtsch Arztebl Int.* 2015;112(37):616-625; quiz 626. doi:10.3238/arztebl.2015.0616

Diagnostic Approach to Atypical Optic Neuritis - EyeWiki. eyewiki.aao.org. Accessed January 15, 2023.

https://eyewiki.aao.org/Diagnostic_Approach_to_Atypical_Optic_Neuritis#cite_note-:0-3

Demyelinating Optic Neuritis - EyeWiki. [Eyewiki.aao.org](https://eyewiki.aao.org). Accessed January 15, 2023.

https://eyewiki.aao.org/Demyelinating_Optic_Neuritis

Right Eye Blind Spot

Chief Complaint

New blind spot in the right eye

History of Present Illness

A 49-year old male presented complaining of a new blind spot in the right eye. This symptom began 2 weeks ago described as flashes located inferotemporally in the right eye along with metamorphopsia and a scotoma. The flashes resolved and his symptoms progressed to an inferotemporal scotoma. Of note, he reported a blood pressure of 169/99 on last check which has been fluctuating with reports of low blood pressure.

Ocular History

Zoster ophthalmicus (several years ago)

Pertinent Past Medical History

Hypertension

Hyperlipidemia

Asthma

HyperIgE (Job) syndrome

DVT after knee surgery at age 16

Shingles/Ramsay Hunt Syndrome

Medications

Amlodipine

Aspirin

Atorvastatin

Duloxetine

Lasix

Pantoprazole

Ranitidine

Saline (ocular)

Allergies

Ciprofloxacin

Codeine derivatives

NSAIDs

Penicillin

IV vancomycin

HCL in dextrose

Zosyn SOLR

Family History

Maternal: glaucoma, HTN/HLD, heart failure, breast cancer

Paternal: myocardial infarction (deceased)

Social History

History of tobacco use (1 pack per day), current status unknown

No alcohol use

Review of Systems

Denies fever, chills, headache, chest pain, palpitations, shortness of breath, abdominal pain, nausea, vomiting, focal weakness, or sensation loss

Ocular Exam

Physical Exam		
	OD	OS
Visual Acuity	20/40	20/50+2
Intraocular Pressure (mmHg)	10	19
Pupils	PERRL, no APD	PERRL, no APD
Confrontational Visual Field	Total inferotemporal deficit	normal
Extraocular Motility	EOMI	EOMI

Slit Lamp Exam		
	OD	OS
External	Unremarkable	Unremarkable
Lids and Lashes	Normal, no lesions	Normal, no lesions
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	Clear	Clear
Anterior chamber	Deep and quiet	Deep and quiet
Iris	Round and reactive	Round and reactive
Lens	Clear	Clear

Dilated Fundus Exam		
	OD	OS
Cup to Disc	0.25	0.25
Optic Nerve	sharp disc	sharp disc
Vessels	AV nicking, Plaque in vessel off nerve superiorly	AV nicking
Macula	Cotton-wool spot superior temporal with whitening along superior macula and superior temporal arcade	Normal
Periphery	Flat, media clear, no other inflammatory or retinal findings	Flat, media clear, no other inflammatory or retinal findings
Vitreous	Clear	Clear

Imaging

- OCT macula (Heidelberg)
 - OD: distortion/thickening of inner retina
 - OS: normal foveal contour and thickness
- Optos Fundus Photos and Fundus Autofluorescence
 - OD: Hollenhorst plaque in retinal artery off superior nerve, hypertensive vascular changes, punctate hemorrhages in periphery
 - OS: hypertensive vascular changes

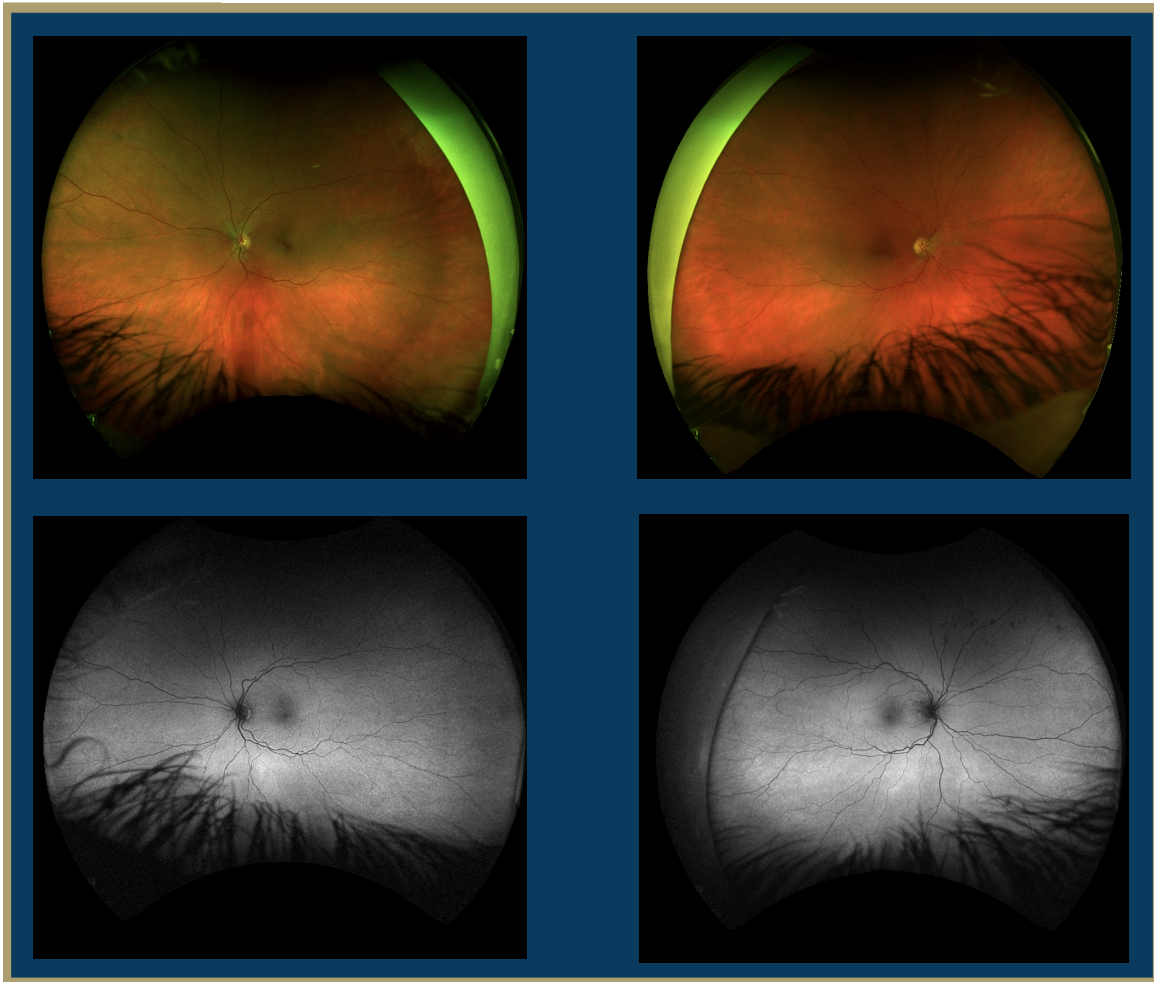


Figure 2: Optos Fundus Photo and Fundus Autofluorescence showing Hollenhorst plaque in the retinal artery off the superior nerve, hypertensive vascular changes, and punctate hemorrhages in the periphery of the right eye and hypertensive vascular changes in the left eye.

- MRI brain with contrast
 - multiple small subacute intracranial infarcts within the left frontal and parietal lobes, right caudate and right inferior cerebellum and abnormal FLAIR hyperintensity with associated susceptibility hypointensity within the left parietal and occipital lobe sulci concerning for subarachnoid hemorrhage
- Carotid Ultrasound
 - Unremarkable, minimal bilateral plaque

Differential Diagnosis

- Acute branch retinal artery occlusion OD, involving macula
 - Patient has several cardiovascular risk factors, but no history of heart murmur and TTE negative in 2018
- Transient monocular visual loss due to retinal artery vasospasm
- Hypertensive retinopathy OU

Clinical Course

This patient is a 49-year old male who was originally referred to the ophthalmology clinic by his primary care provider for concern of retinal detachment after experiencing transient floaters and a persistent inferotemporal blind spot in the right eye. Clinical exam revealed a BRAO and he underwent subsequent stroke work-up. Carotid ultrasound was unremarkable. Brain MRI revealed CVA with multiple subacute infarcts. The patient underwent echocardiography, which revealed a patent foramen ovale (PFO). He was subsequently started on dual antiplatelet therapy. He also underwent hypercoagulability and inflammatory work-up, which were negative. He was discharged with scheduled follow-up for cardiology and neurology. The patient received a cardiology evaluation for percutaneous closure of the patent foramen ovale, but was determined to be a poor candidate for this intervention due to a recent hospitalization with sepsis. He continues to follow up with cardiology on a regular basis.

Discussion

Branch retinal artery occlusion is a cerebrovascular accident (CVA) or stroke involving the retinal vasculature. It leads to decreased blood flow to the retina and presents clinically as acute, painless monocular vision loss. On visual field testing, BRAO most commonly presents as an acute scotoma. In older adults, the etiology is often an atherosclerotic plaque/valvular emboli. In adults under 50 years of age, the etiology is often a cardiac embolic source or hypercoagulable state.

BRAO causes acute retinal ischemia and is therefore considered a vascular emergency. In this case, the patient was sent immediately to the emergency department upon diagnosis to receive a stroke work-up. This patient had pre-existing risk factors for stroke and acute coronary events. A robust work-up must be done to rule out any undiagnosed life-threatening conditions.

This patient was ultimately found to have a Patent Foramen Ovale (PFO) on imaging. PFO is a risk factor for life-threatening emboli and PFO-related BRAO have been reported in several cases in young adults (Shoebi et al. 2018). Referral to cardiology is necessary for further management of the condition.

In conclusion, BRAO is an uncommon diagnosis, especially in a patient under 50. Patients who present with retinal artery occlusion should undergo further work-up as this may ultimately reveal underlying systemic etiology. In this case, BRAO was the presenting sign that led to the diagnosis of patent foramen ovale (PFO) and the finding of multiple intracranial infarcts.

References

Biousse V, Nahab F, Newman NJ. Management of Acute Retinal Ischemia: Follow the Guidelines! *Ophthalmology*. 2018;125(10):1597-1607. doi:10.1016/j.ophtha.2018.03.054

Chatziralli IP, Parikakis EA, Mitropoulos PG. Undiagnosed patent foramen ovale as a rare cause for branch retinal artery occlusion. *Eur J Ophthalmol*. 2015;25(5):e88-90. doi:10.5301/ejo.5000608

Greven CM, Slusher MM, Weaver RG. Retinal Arterial Occlusions in Young Adults. *American Journal of Ophthalmology*. 1995;120(6):776-783. doi:10.1016/S0002-9394(14)72731-X

Hayreh SS. Ocular vascular occlusive disorders: Natural history of visual outcome. *Progress in Retinal and Eye Research*. 2014;41:1-25. doi:10.1016/j.preteyeres.2014.04.001

Iqbal P, Shams A, Ali M, Muthanna B, Hussain T. Rare Association of Patent Foramen Ovale and Atrial Septal Aneurysm Leading to Branch Retinal Artery Occlusion in a Young Healthy Man. *Cureus*. 2020;12(7). doi:10.7759/cureus.8994

Wieder MS, Blace N, Szlechter MM, Shulman E, Thankenchen J, Mbekeani JN. Central retinal artery occlusion associated with patent foramen ovale: a case report and literature review. *Arquivos Brasileiros de Oftalmologia*. 2021;84(5). doi:10.5935/0004-2749.20210073

Acknowledgments

Case Report Authors

Optic Neuritis

Devin Hill, MS3

Tyler Muser, MS2

BRAO

Sahak Hovsepian, MS3

Shaleen Arora, MS2

Resident Editors

Masumi Asahi, MD

Braedon Murdock, MD

Will Foos, MD

Attending Reviewer

David Belyea, MD MBA

Editors in Chief

Jason Dossantos, MS3

Julie Thomasian, MS4