



**Differential Diagnosis of
Optic Nerve Disorders**

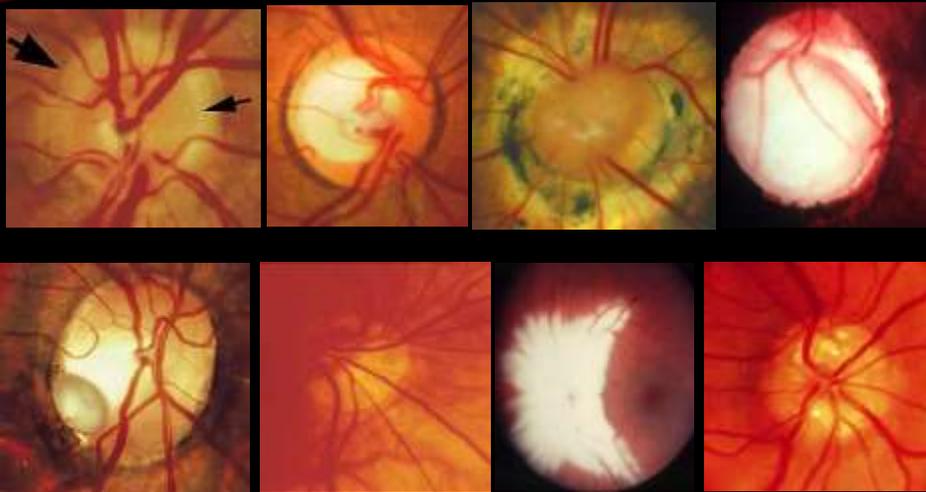
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OPTIC NERVE DISORDERS

1. Congenital
2. Hereditary
3. Ischemic
4. Demyelinating
5. Inflammatory
6. Nutritional
7. Toxic
8. Radiation
9. Traumatic
10. Tumors
11. Compressive
12. Papilledema
13. Glaucomatous

CONGENITAL ANOMALIES



Hereditary Optic Atrophy

1. Dominant Optic Atrophy
2. Recessive Optic Atrophy
3. Leber's Hereditary Optic Neuropathy (LHON)

DOMINANT OPTIC ATROPHY

- Begins before the age of 10 years
- Bilateral but it can be asymmetrical
- Mild visual loss from 6/9 to 6/36
- Fields: subtle central or cecocentral scotoma
- Dyschromatopsia
- Temporal optic disc pallor with triangular excavation



RECESSIVE OPTIC ATROPHY

- Rare
- Begins before the ages of 5 years
- Bilateral severe visual loss
- Nystagmus in 50%
- Pale discs with vascular attenuation but normal ERG
- Mental retardation, spasticity, hypertonia, and cerebellar ataxia (Behr's syndrome)

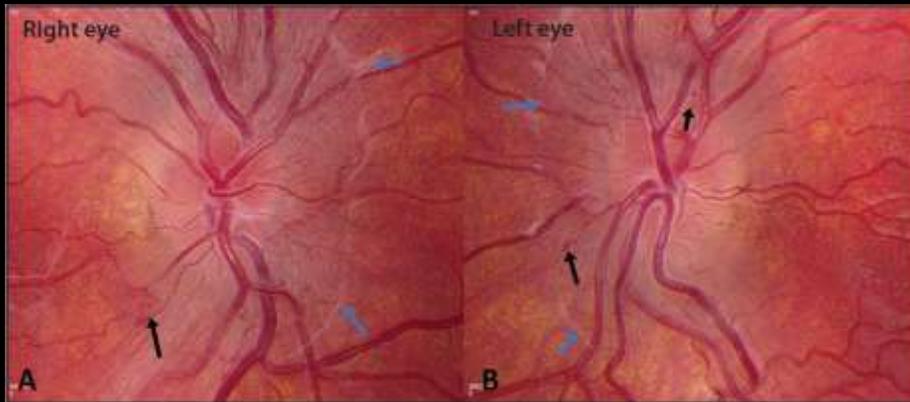
LEBER'S HEREDITARY OPTIC NEUROPATHY (LHON)

- Maternally inherited disease due to mitochondrial DNA (mtDNA) point mutations → defective oxidative phosphorylation
- Usually male in 2nd to 4th decade
- Papillomacular bundle becomes atrophic (impairments in the mitochondria)

- Progressive bilateral (sequential or simultaneous) severe, painless central vision loss (6/60 to CF).
- Acquired red/green dyschromatopsia
- Central or centrocentral scotomas
- Pupillary reactions are usually normal, even in the early monocular stages of LHON

3-Triad:

1. Circumpapillary telangiectatic microangiopathy
2. Pseudoedema of the disc (swelling of the NFL)
3. Absence of fluorescein staining



- Later on, mild temporal disc pallor
- Systemic associat: cardiac arrhythmia
- Molecular genetic analysis of mtDNA from leukocytes confirms diagnosis

ANTERIOR ISCHEMIC OPTIC NEUROPATHY (AION)

- The most common cause of acute optic neuropathy in old age.
- Non-arteritic [NAION] or arteritic
- Arteritic [AION] is associated with giant cell arteritis.

NON-ARTERTIC AION

- NAION affects men and women in their 5th and 6th decades.
- Sudden painless visual loss is usually apparent upon awakening (nocturnal hypotension??).

On Examination:

- A small cup disc ratio is usually noted.
- Disc pallor and swelling (diffuse or sectorial especially of the superior disc)
- Hyperemia and disc hemorrhage may occur acutely
- Later on → optic atrophy and retinal arteriolar narrowing.



- The visual loss is usually irreversible.
- Progressive deterioration in VA and visual field may occur after six weeks.
- The fellow eye may be affected in up to 25-40% of patients, with young diabetic males being at highest risk.

ARTERITIC (GIANT CELL ARTERITIS) AION

- A disease of the elderly
- Its prevalence increases from the 6th through the 8th decades of life.
- Females are affected **3** times as often as males

ARTERITIC (GIANT CELL ARTERITIS) AION

- Sudden, painless, and profound visual loss (CF to NPL). Bilateral presentation is common
- Malaise, weight loss, fever, abdominal pain, anorexia, headache, scalp and temples tenderness, jaw claudication, muscle pain, and swelling.

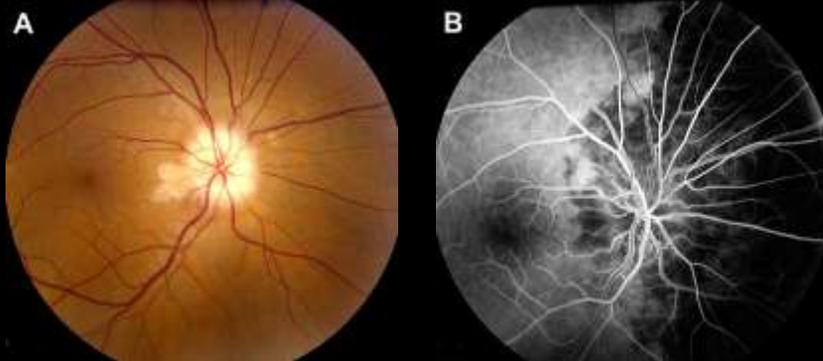
On Examination:

- The disc is chalky white, pale, and swollen.
- **Tenderness over the TA and it may be pulseless.**
- Oral, tongue, or even scalp ulcers may be seen.



On Examination:

➤ Multiple ischemia (eg, choroidal infarction, anterior segment ischemia, EOM ischemia → diplopia).



➤ Later on , often years later, → much higher incidence of abdominal aortic aneurysm.

Other causes of Arteritic AION

- | | |
|-----------------------------|------------------------|
| 1. herpes zoster | 7. Behçet's disease |
| 2. relapsing polychondritis | 8. Crohn's disease |
| 3. polyarteritis nodosa | 9. systemic lupus |
| 4. rheumatoid arthritis | 10. Rarely, infections |
| 5. Wegener's vasculitis | by <i>Rickettsia</i> |
| 6. Takayasu's arteritis | <i>conorii</i> |

DEMYELINATING OPTIC NEUROPATHY

- 20-50 years, mean of 30-35 y
- ♀ > ♂ 77% are women
- 85% Caucasian
- Family history of DM may be present

DEMYELINATING OPTIC NEUROPATHY

- **Loss of vision:** abrupt, over hours to days. Worsening of vision for up to 14 days thereafter V/A improves over several weeks.
- Tends to be monocular, but occasionally both eyes are affected simultaneously.
- Vision loss exacerbated by heat or exercise (Uhthoff phenomenon).

DEMYELINATING OPTIC NEUROPATHY

- **Dyschromatopsia** : may be more prominent than the decreased vision.
- **Pain** in or around the eye (more than 90% of patients usually is exacerbated by eye movement).
- History of **previous episodes** in the same or the fellow eye, previous history of **neurologic problems**

ON EXAMINATION:

- RAPD in **unilateral** cases
- **Color** vision and **contrast** sensitivity are impaired out of proportion to V/A
- **Visual loss** varies from a mildly reduced V/A to NoPL
- 85% of patients with 1st attack will recover to 20/40 or better

OPTIC NEURITIS

Viral and Postviral Syndromes (children)
Infections of orbits or paranasal sinuses

Idiopathic orbital inflammation

SYPHILIS, Tuberculosis, LYME

SARCOIDOSIS

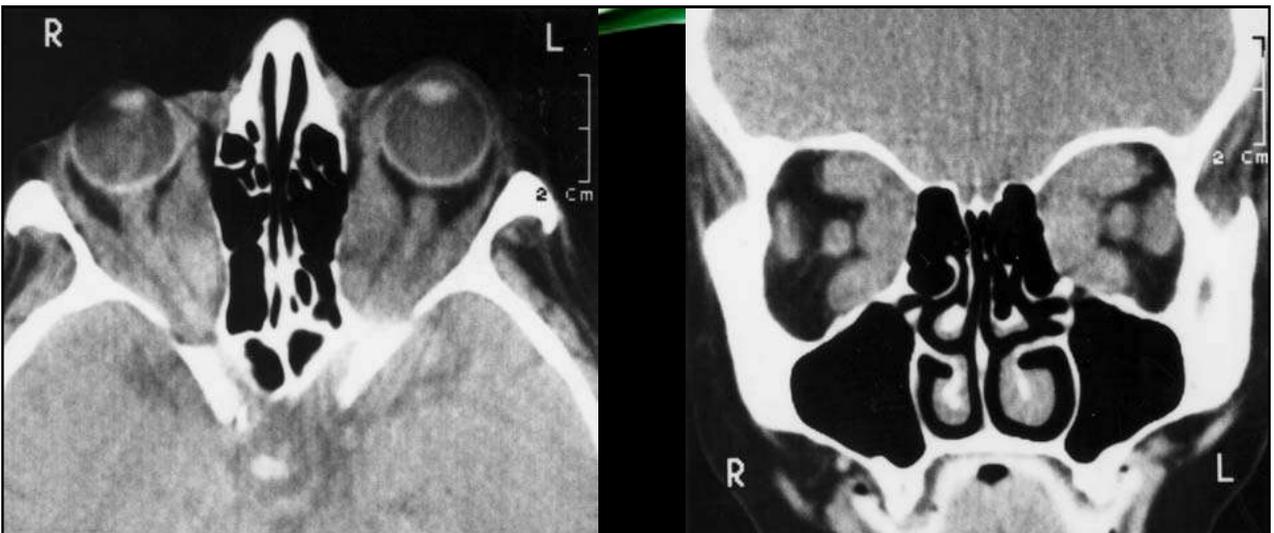
CVDs: WEGNER'S, SLE

Compressive optic neuropathy

<p>*Primary tumors or metastases</p> <ul style="list-style-type: none"> •ON gliomas •ON meningiomas 	<p>Thyroid ophthalmop.</p> <ul style="list-style-type: none"> *Orbital hemorrhage <u>Inflammatory processes</u> Orbital pseudotumor Sarcoidosis *Fibrous dysplasia 		<p>*<u>Intracranial causes</u></p> <ul style="list-style-type: none"> <u>Pituitary adenoma</u> <u>Craniopharyngioma</u> Metastatic carcinoma Aneurysms
<p>*<u>infiltrative processes</u></p> <p>Leukemia Lymphoma</p>	<p><u>Orbital tumors</u></p> <ul style="list-style-type: none"> •Meningiomas •Cavernous Hemangiomas •Schwannoma 	<p><u>Orbital Cysts</u></p> <ul style="list-style-type: none"> •Dermoid cysts •Mucocoeles 	<p>*<u>Carcinomas extending from sinuses</u></p>

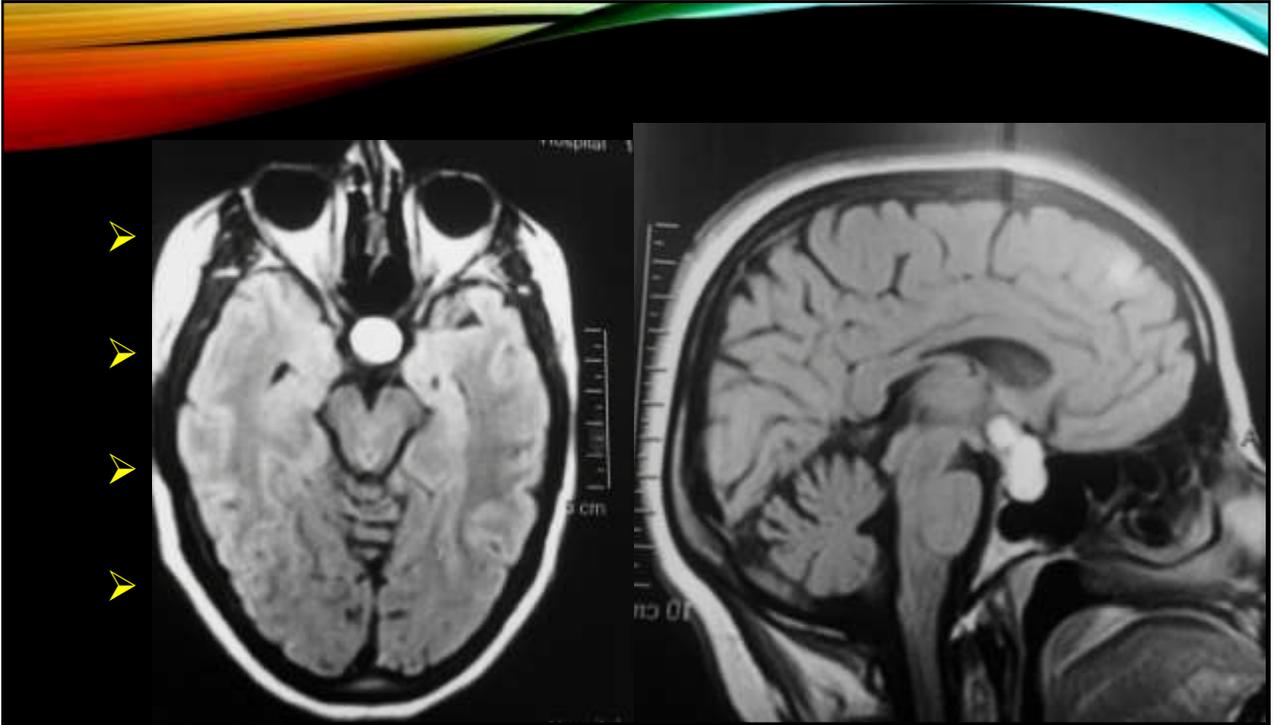
THYROID OPTIC NEUROPATHY

- Occurs in about 5%
- Early defective color vision
- Usually normal disc appearance
- May occur in the absence of significant proptosis



A grading scale is used to assess the degree of optic nerve compression by the enlarged EOM at the orbital apex on coronal images.

Grade 0 = no effacement of perineural fat planes by enlarged EOM, whereas grade 4 reflects more than 75% effacement



Optic Nerve Glioma

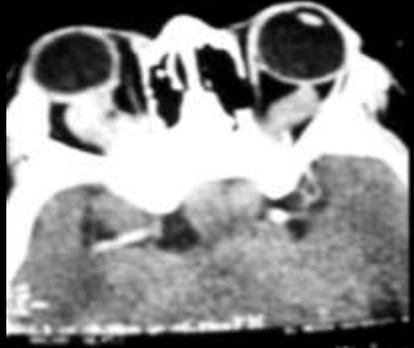






Most common in children under 10 years
No sex predilection

2/3 of all 1ry ON tumors and 5% of all intracranial tumors.



OPTIC NERVE SHEATH MENINGIOMA

Most common in **middle-aged** (20-60 mean =40 and are rare below twenty)

Women :men 3:2

the 2nd most common ON tumor, but only **1%** of all meningioma



TOXIC AND NUTRITIONAL NEUROPATHY

Nutritional deficiency: Vitamins (e.g. B12, thiamine or folic acid) and amino acids used in mitochondrial metabolism (e.g. homocysteine or methionine).

Tobacco-alcohol amblyopia: heavy drinking and smoking → slow, progressive bilateral visual field loss due to cyanide (from tobacco) and low levels of B12 2ry to poor nutrition and poor absorption due to alcohol consumption



Toxins that induce optic neuropathy

Drugs

Ethambutol	Oral hypoglycemics (chlorpropamide & tolbutamide)
Isoniazid	
Chloramphenicol	Quinine
Digitalis	Thallium
D-penicillamine	

Others

Methanol
Carbon
monoxide
Cyanide
Lead

TOXIC AND NUTRITIONAL NEUROPATHY

- Bilateral, insidious loss of central vision
- Dyschromatopsia
- Bilateral cecocentral scotomas

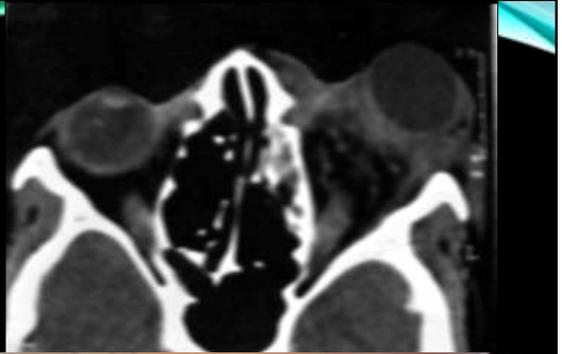
RADIATION OPTIC NEUROPATHY(RION)

- RION is a complication of radiotherapy caused by radiation necrosis of anterior visual pathway → acute, profound and irreversible visual loss.
- Unilateral or bilateral; simultaneous or sequential.
- Occurs between 10-20 months after treatment (but the onset may range from 3 months to 9 years).

TRAUMATIC OPTIC NEUROPATHY (TON)

- Transmitted shock from an orbital impact to the intracanalicular portion of optic nerve
- Penetrating injury ,FB or bony fragments
- Orbital hemorrhage and ON sheath hematoma

➤ Optic nerve shearing or avulsion



papilledema:

1. Blurring of disc margin
2. Disc hyperemia
3. Loss of venous pulsations
4. Venous distention
5. Deflection of the vessels
6. Filling of the cup
7. Elevated optic disc
8. Hemorrhage and exudate
9. Patonoe line
10. Optociliary shunt vessels

