Evaluation of ONH Pallor in Glaucoma Patients and Suspects

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I. Abstract

This case report will evaluate a young glaucoma suspect with unilateral sectoral optic nerve pallor. A discussion will highlight four cases of optic nerve pallor with differing etiologies and clinical features.

II. Case History

- Patient Demographics: 29 year old black female
- Chief Complaint: Referred from an outside provider for glaucoma evaluation due to suspicious optic nerve head appearance.
- Ocular/Medical History: Unremarkable
- Medications: Vitamin D3 supplement
- Other: (+) Fam Hx of Glaucoma- grandfather

III. Pertinent Findings

- Clinical: Large, physiologic cupping with fully intact neuroretinal rims with no glaucomatous features OU; Superior temporal optic nerve pallor OS. IOP 12 mm Hg OS; CCT 493 OD, 488 OS; RAPD OS; Abnormal RNFL OS; normal RNFL OD. Mild visual field loss inferior OS corresponding to area of disc pallor.
- Physical: None
- Lab/Imaging Studies: MRI: Unremarkable, Bloodwork: Unremarkable,
- Other: (-) Red cap abnormalities, (-) Color vision defect, (-) Retinal vascular abnormalities, Gonioscopy open without abnormalities OD, OS

IV. Differential Diagnosis
• **Primary/Leading:** Compressive lesion of anterior visual pathway.

• **Other differential diagnoses:** Occult optic neuritis, ischemic optic neuropathy, inflammatory, infiltrative, infectious, or metabolic optic neuropathy.

V. **Diagnosis/Discussion**

• **Discussion:** Optic nerve pallor is not a common feature of glaucoma and optic atrophy must be thoroughly investigated. Optic atrophy has many different etiologies with differing presentations. Primary optic atrophy refers to a condition not preceded by inflammation, secondary optic atrophy occurs secondary to inflammation, and consecutive optic neuropathy occurs secondary to retinal or degenerative conditions. It is important to determine the etiology of optic nerve atrophy as some causes may be associated with significant visual morbidity. Pallor can be difficult to diagnose especially in patients with end-stage glaucoma; therefore, it is important to understand diagnostic features between glaucomatous and nonglaucomatous optic atrophy.

A retrospective study was conducted at the University of Benin Teaching Hospital where they collected medical records from patients that had nonglaucomatous optic atrophy. The most common etiologies were unknown (45.2%), chorioretinal disease (23.1%), trauma (13.5%), toxic-nutritional (7.7%), and compressive lesions (4.8%).

Although compressive lesions only accounted for 4.8% of patients, optic atrophy was the most common (23.9%) ocular sign among patients with intracranial tumors in Nigeria.

A second study discussed key differences between glaucomatous and nonglaucomatous optic disc cupping. Changes in the optic disc from glaucoma typically include: focal enlargement where vertical diameter change is larger than horizontal change, deepened excavation, increased exposure of lamina cribosa, wedge-shaped NFL defects, flame-shaped disc hemorrhages, and beta-zone peripapillary atrophy.

Optic nerve color, diffuse vs. focal rim loss, VF defects, IOP, visual acuity, and the presence of peripapillary atrophy should all be considered when differentiating glaucomatous optic atrophy versus nonglaucomatous.

A third study compared optic nerve cup depths of glaucomatous optic neuropathy and compressive optic neuropathy. The mean and maximum cup depths were significantly smaller in those with compressive optic neuropathy versus glaucomatous neuropathy.
A fourth study looked at the incidence of positive neuroradiologic studies in consecutive patients with glaucoma associated with normal IOP compared to disc cupping associated with intracranial masses. Younger age, lower levels of visual acuity, vertically aligned visual field defects, and neuroretinal rim pallor may increase the likelihood of identifying an intracranial mass lesion.

- **Unique features:** This presentation will additionally review four cases of optic atrophy with different etiologies.
  - **Case 1:** A 54 year old male with POAG presents with ONH pallor. Visual field test shows a bi-temporal hemianopic defect. The patient was sent for an MRI and was diagnosed with a pituitary adenoma.
  - **Case 2:** A 58 year old female with POAG presents with ONH pallor. MRI indicates a chiasmal mass, likely pituitary macroadenoma or possibly craniopharyngioma.
  - **Case 3:** A 77 year old male with a systemic history of diabetes, hypertension, and hypercholesterolemia presents with a 2+ APD and optic nerve pallor OS. VF represents inferior arcuate scotoma OS. The patient was diagnosed with a longstanding BRAO.
  - **Case 4:** A 55 year old female presents with elevated intraocular pressure, an RAPD and optic disc pallor OS with an unremarkable systemic history. Subsequent neuroimaging and evaluation failed to reveal a cause.

**VI. Treatment/Management**

- **Treatment:** None. Monitor closely with photos.

**VII. Conclusion**

- **Clinical Pearls:**
  - Although compressive lesions account for a small percentage of patients with optic atrophy, optic atrophy is the most common ocular sign for compressive lesions.
Visual field testing serves as a diagnostic test for many intracranial lesions; therefore, it should be utilized in patients with optic atrophy.

Patients with optic nerve pallor should have a comprehensive examination that includes: a complete medical history, visual acuity, color vision, confrontation fields, pupil reaction, red cap desaturation, and dilated fundus evaluation.

Patients diagnosed or suspected of having glaucoma also demonstrating optic nerve head pallor in excess of cupping need to be further evaluated for alternate conditions or comorbidities.

References:


