

Commentary: Neuro-ophthalmological conditions mimicking glaucoma – A diagnostic challenge

Glaucoma is a chronic progressive optic neuropathy. It is characterized by typical changes in the optic nerve head (ONH) including enlarged vertical cup to disc ratio (CDR) of >0.6 , asymmetry of >0.2 in the CDR, notching or neuro retinal rim (NRR) loss, disc hemorrhages and nerve fiber layer defects. The first step in establishing the diagnosis of glaucoma is a careful and systematic evaluation of ONH, and retinal nerve fiber layer (RNFL). This is best achieved by an indirect ophthalmoscopy, using slit lamp bio-microscopy and a 90 or 78 Diopter lens. The systematic approach for evaluating the ONH for glaucoma becomes simple with the five rules (5 Rs)^[1] namely,

1. Observing the scleral **R**ing to identify the optic disc size
2. Identifying the size and color of neuroretinal **R**im
3. Examination of **R**etinal nerve fiber layer
4. Examination of the **R**egion outside the optic disc for parapapillary atrophy
5. Watching for **R**etinal and optic disc hemorrhages.

Once the clinical evaluation is completed, the findings need to be correlated with field changes. Optical coherence tomography (OCT) also plays an important role in establishing the diagnosis and monitoring the progression. Though elevated intraocular pressure (IOP) is the most important risk factor for glaucoma, a significant proportion of patients may present with IOP in the normal range.^[2] A definitive diagnosis is not always simple and straight forward in case of normal tension glaucoma (NTG).^[3] Because IOP is not a criteria, there is a high possibility of misdiagnosing various non-glaucomatous optic neuropathies as NTG.

NTG should be considered as a diagnosis of exclusion, after ruling out the other possibilities like primary open angle glaucoma with wide fluctuations in the IOP, secondary glaucoma, such as steroid induced glaucoma, burnt out stage of pigmentary glaucoma. Apart from these glaucoma, it is essential to rule out non-glaucomatous optic neuropathies like optic atrophy following anterior ischemic optic neuropathy, compressive optic neuropathy, demyelinating optic neuritis,

toxic optic neuropathies, hereditary optic neuropathies and congenital disc anomalies.

It is not uncommon to see many of these conditions being treated as glaucoma. Choudhari *et al.* reported a case series of six patients with neuro-ophthalmic optic neuropathies, misdiagnosed as glaucoma, managed not only with medications but also had undergone surgical interventions for glaucoma.^[4] In another cross-sectional study, fundus photographs and Humphrey visual fields of 102 eyes with neuro-ophthalmic conditions mimicking glaucoma and 42 eyes with NTG were graded by a masked glaucoma expert. They found that about one quarter of the neuro-ophthalmic conditions were misdiagnosed as glaucoma.^[5] Hence, a meticulous history taking and a comprehensive eye examination is a must to differentiate these life-threatening conditions from glaucoma.

Glaucoma is a highly asymptomatic condition, which is more common in elderly. In glaucoma, visual acuity and central vision is maintained well even in the advanced stage of the disease. It is often bilateral and symmetric. ONH will reveal a vertically enlarged cup, NRR remains pink and the colour vision remains intact till the advanced stage. The field defects typically respect the horizontal meridian with characteristic arcuate pattern.

Unlike glaucoma, a young patient with cupping, presenting with a history of sudden loss of vision or rapidly progressive vision loss, headache or diplopia, should prompt the physician to do a complete neuro-ophthalmic work up. In these neuro-ophthalmic conditions, the vision loss is usually profound, will not correspond to the cup. There can be a relative afferent pupillary defect. ONH will reveal pallor more than cup especially in the temporal aspect and the color vision is usually impaired. Studies have demonstrated that OCT in these eyes had more diffuse RNFL loss, with more thinning in the nasal and temporal part as compared to glaucomatous eye with similar average RNFL thickness.^[6] The field defects usually will respect the vertical meridian. There can be central, centrocecal, altitudinal, bitemporal, quadrantanopic, or hemianopic field loss depending upon the underlying pathology. With these signs and symptoms one should proceed with neuroimaging to establish the diagnosis. Routine neuroimaging is not mandatory in all patients with NTG as the yield is low.^[7]

In case of congenital disc anomalies it is essential to establish good baseline tests including fundus photography and fields. This will help to assess progression if any during the follow-up visits.

Though glaucoma is the commonest cause of cupping, nearly 20% of the cupping can be non-glaucomatous.^[8] Discriminating glaucoma from non-glaucomatous cupping can be a difficult task even in the hands of an expert. We need to remember that it is both clinically and financially important. Subjecting every patient with suspected NTG to neuroimaging should not be our goal. Misdiagnosis can have a serious impact not only on the visual status but also on the overall wellbeing of the patients. Let us hone our clinical skills, and neither overdiagnose glaucoma nor miss the possible life-threatening conditions.

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References

1. Fingeret M, Medeiros FA, Susanna R Jr, Weinreb RN. Five rules to evaluate the optic disc and retinal nerve fiber layer for glaucoma. *Optometry* 2005;76:661-8.
2. Collaborative Normal-Tension Glaucoma Study Group. The effectiveness of intraocular pressure reduction in the treatment of normal-tension glaucoma. *Am J Ophthalmol* 1998;126:498-505.
3. Sharma J, Jain A, Bhagat PR. Importance of neuroimaging in normal tension glaucoma. *Indian J Ophthalmol* 2020;68:1163-5.
4. Choudhari NS, Neog A, Fudnawala V, George R. Cupped disc with normal intraocular pressure: The long road to avoid misdiagnosis. *Indian J Ophthalmol* 2011;59:491-7.
5. Dias DT, Ushida M, Battistella R, Dorairaj S, Prata TS. Neurophthalmological conditions mimicking glaucomatous optic neuropathy: Analysis of the most common causes of misdiagnosis. *BMC Ophthalmol* 2017;17:2.
6. Gupta PK, Asrani S, Freedman SF, El-Dairi M, Bhatti MT. Differentiating glaucomatous from non-glaucomatous optic nerve cupping by optical coherence tomography. *Open Neurol J* 2011;5:1.
7. Greenfield DS, Siatkowski RM, Glaser JS, Schatz NJ, Parrish RK II. Cupped disc. Who needs neuroimaging? *Ophthalmology* 1998;105:1866-74.
8. Trobe JD, Glaser JS, Cassady JC. Optic atrophy. Differential diagnosis by fundus observation alone. *Arch Ophthalmol* 1980;98:1040-5.

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