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Optic Nerve Head Findings: How a Neuro-ophthalmologist can Aid the Neurologist?

ABSTRACT

Context: Specific pathological patterns of neurological diseases can be studied in the optic nerve. **Aims:** The aims of study were to study the demographics, etiology, and clinical characteristics in neuro-ophthalmic optic nerve head disorders with their systemic correlation in adults in India. **Settings and Design:** In the study period of 18 months, all patients presenting to our institute with unilateral or bilateral optic nerve head disease were included in the study. **Materials and Methods:** This study was retrospective review of prospectively collected data. **Statistical Analysis Used:** The results were analyzed using Microsoft Office Excel (Microsoft Corporation, 2010, Louisville KY) software with filters applied on the Master Chart to enable a Sub-Group Analysis. **Results:** A total of 195 patients received specialist neuro-ophthalmology consultations during the study period, of which 170 patients met the inclusion-exclusion criteria and were reviewed. The age range was 16–72 years. There were 98 (57.64%) males and 72 (42.35%) females. Headache was the presenting complaint in 97 (57.05%) patients. Etiological diagnosis was achieved in 161 (94.70%) patients. From a neurological perspective, most common diagnosis was benign intracranial hypertension in 32 (18.82%) patients. From a neurosurgical perspective, pituitary adenoma was the most common space-occupying lesion (SOL) which was seen in 18 of the 44 patients with SOLs (i.e., 25.88% of the total 170 patients). **Conclusions:** This study highlights how a neuro-ophthalmic examination can provide sight and life-saving insight to the neurologist and the neurosurgeon into underlying disease processes in the optic nerve in neurologic diseases.

Key words: Neuro-ophthalmology, Optic nerve head disorders, Visual field analysis

Key Messages: Specific pathological patterns seen in the optic nerve can help the neuro-ophthalmologist provide insight to the neurologist and the neurosurgeon underlying disease processes in the optic nerve. This helps affords early detection of neurologic disease and can help have better outcomes while managing them.

INTRODUCTION

Optic nerve head disorders result from various etiologies which may be intraorbital or intracranial optic nerve related causes or systemic diseases which include life as well as vision threatening conditions. Many of these optic nerve head disorders are seen predominantly in specific gender, age-groups, and regions. The etiology of optic neuropathy, can rarely be found on the basis of only a single clinical finding. There is considerable overlap of clinical features between even the relatively common optic neuropathies. In optic neuropathies, very often, specific pathological patterns of disease seen in the optic nerve can help the neuro-ophthalmologist provide sight and life-saving insight to the neurologist and the neurosurgeon into the underlying disease process, and thus, afford early detection of the disease and help have better outcomes, not only in relation to the eye but also in managing and treating other secondary systemic manifestations of the disease. An appropriately made clinical impression of the pathology in the optic nerve is likely to be reliable and sufficient to detect these specific patterns of the disease in the optic nerve and, thus, guide appropriate investigations and lead to the correct diagnosis of the disease and treatment.

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Before we proceed further, we understand that, the term "disc edema" describes a pathological "pattern" in the optic nerve which represents a pathology which may be either "Papilledema" or "Optic Neuritis (ON)"/"Papillitis." It is then up to the neuro-ophthalmologist to use appropriate clinical findings including best-corrected visual acuity, pupillary reaction – direct and consensual, color vision and visual field to pinpoint the pathology (Papilledema/ON) to the neurologist. Using appropriate investigations including among

others a lumbar puncture or an magnetic resonance imaging (MRI) scan the neurologist can then accurately pinpoint the etiology (say benign intracranial hypertension [BIH]) which leads to the genesis of the pathology (Papilledema) in the optic nerve which is seen as a pathological pattern (Disc edema) bilaterally. Similarly, multiple sclerosis (MS) (etiology) leads to the genesis of retrobulbar/ON (pathology) which sometimes leads to disc edema (pathological pattern) in the optic nerve. Thus, it is relevant to study of etiopathogenesis of the non-glaucomatous optic nerve disorders.

With this background, we decided to study the spectrum including demographics, etiology, and the clinical characteristics in non-glaucomatous optic nerve head disorders with their systemic correlation in adult age group in the neuro-ophthalmic clinic of our hospital which is a tertiary care hospital in India.[1]

SUBJECTS AND METHODS

Approval for this retrospective analysis was obtained from the Ethics and Research Committee of our institute. In the study period of 18 months, all patients presenting to our institute with symptoms and clinical signs suggestive of unilateral or bilateral optic nerve head disease were included in to the study. A retrospective review of this prospectively collected data from the medical records of all these patients was then performed.

Inclusion criteria for the study

All patients:
1. Aged 16 years and above
2. With ophthalmoscopic findings of optic neuropathy
3. Or with visual dysfunction (visual acuity, color vision, and/or visual field loss) attributable to the optic nerve (e.g., relative afferent pupillary defect)
4. And normal intraocular pressure was included in the study.

Thus, patients with a diagnosis of disc edema, papilledema, ON, optic neuropathy, and optic atrophy were included in the study.

Exclusion criteria for the study

Patients with:
1. Glaucomatous optic neuropathy
2. Ocular hypertension
3. Congenital optic nerve disorders
4. Hereditary optic neuropathy were excluded from the study.

Study conduct

With proper informed written consent, patients with various optic nerve disorders were assessed with respect to the presenting complaints. A detailed presenting, past, systemic, and family history was noted. Thereafter, a detailed ophthalmic evaluation including visual acuity, color vision, ocular alignment, and extraocular movements was done. This was followed by assessment of direct and consensual pupillary reactions, slit-lamp biomicroscopy of the anterior segment with intraocular pressure using Applanation tonometry, and assessment of the angles with a 4-mirror goniolens (if required). Subsequently, a post-dilated assessment of the ocular fundus using a 20 D lens (indirect ophthalmoscopy) and an assessment of the optic nerve using a 90 D lens using slit lamp biomicroscopy was done. A fundus photograph was done for each patient. A visual field examination, visual-evoked potential, electro-retinograms, optic coherence tomography, and appropriate neuroimaging and laboratory investigations were done when required. A follow-up visit within 1–2 weeks with appropriate investigations and other specialists' referrals was done to look for systemic corelations. Data on age, gender, laterality of optic atrophy, visual function, and etiology of optic atrophy were obtained. The results were analyzed using Microsoft Office Excel (Microsoft Corporation, 2010, Louisville KY) software with "Count if" and "Sum" being the commonly used functions with filters applied on the Master Chart to enable a Sub-Group Analysis.

RESULTS

A total of 195 patients received specialist neuro-ophthalmology consultations during the study period, of which 170 patients met the inclusion-exclusion criteria and were reviewed. The age range was 16–72 years. The mean age was 39 years. There were 98 (57.64%) males and 72 (42.35%) females (M:F ratio = 1.3:1). The two most common presenting complaints were headache and progressive deterioration of vision; with headache as the presenting complaint in 97 (57.05%) of the patients.

For the purpose of analysis, both eyes of each patient were considered separately. Thus, we have analyzed 340 eyes of 170 patients. Of the 340 eyes (of 170 patients) analyzed, 54 eyes (15.88%) had a presenting visual acuity of <6/60. The color vision was affected in 94 (27.64%) of the 340 eyes. The worse affected eye showed a relative afferent pupillary defect in 43 (25.29%) of the 170 patients.

Major findings

Optic nerve head findings

The most common optic nerve finding/pathological pattern seen by the neuro-ophthalmologist was "edema" (ranging from mild disc hyperemia to severe disc edema) at presentation in 208 (61.17%) of the 340 eyes, of which 15 eyes (4.41%) had severe splinter hemorrhages at the optic nerve. Table 1 shows the examination findings of the optic nerve in the 340 eyes of the 170 patients.

Clinical impression based on neuro-ophthalmic examination

Before any further investigations, based on the history, visual assessment, color vision, and examination of the

pupils and the optic nerves a preliminary clinical impression of the pathology were formed in all the 170 patients. Papilledema (84 patients–49.41%) was the most common clinical impression/pathology, followed by optic atrophy (38 patients–22.35%) and ON (16 patients–9.41%). Table 2 shows the clinical impression of the pathology made by the neuro-ophthalmologist after a clinic-based examination of the 170 patients.

Visual field analysis of the patients

Visual field analysis of the central 30 degrees (CVF 30-2) was requested in 230 eyes of the 170 patients; of which it was not accurately performed and, thus, was not interpreted in 19 eyes. Of the 211 eyes in which the visual field was interpreted, 50 eyes had a normal visual field. Of the remaining 161 eyes, 62 eyes showed an advanced field defect; 34 eyes an enlarged blind spot; 29 eyes showed a hemianopic field; and 26 eyes had a central scotoma. Table 3 shows an analysis of the visual field defects in 211 eyes of the 170 patients.

Table 1: Examination findings of the optic nerve in the 340 eyes of the 170 patients. 14 optic nerves were healthy

Disc findings	Number of eyes
Healthy disc	14
Hyperemia	21
Mild edema	134
Moderate edema	31
Severe edema	22
Splinter hemorrhages	15
Temporal pallor	53
Primary optic atrophy	5
Secondary optic atrophy	4
Global pallor	32
Pallid edema	9

Table 2: Clinical impression of the pathology made by the neuro-ophthalmologist after a clinic based examination of the 170 patients

Clinical impression	Number of patients
Papilledema	84
Optic neuritis	16
Optic atrophy	38
Hypertensive retinopathy	13
Pallid disc edema	2
Optic neuropathy	8
Traumatic optic neuropathy	3
Ischemic optic neuropathy	3
Inflammatory optic neuropathy	1
Lymphoproliferative disorder	1
Temporal optic nerve pallor	1

Neuroimaging findings of the patients

Neuroimaging using a MRI scan was requested and performed by 128 of the 170 patients. Radiological evidence of BIH was seen in 32 patients; a space-occupying lesion (SOL) in 38 patients; MS/ON in 22 patients; a cerebral infarct in 13 patients; and cortical venous sinus thrombosis in eight patients.

Table 4 shows the MRI findings in 128 of the 170 patients.

Final diagnosis as achieved by the neurologist

A diagnosis of the etiological cause of the non-glaucomatous optic nerve disorders was achieved in 161 (94.70%) of the 170 patients. A diagnosis of BIH was made in 32 (18.82%) patients. Pituitary adenoma was the most common SOL which was seen in 18 of the 44 patients (25.88% of the 170 patients) with SOLs. Relatively rare disorders like POEMS syndrome, gas geyser syndrome and chronic relapsing inflammatory optic neuropathy were seen in our series. Table 5 summarizes the various final etiological diagnoses in our series.

Table 3: Analysis of the visual field defects in 211 eyes of the 170 patients

Visual field analysis	Number of eyes
Normal field	50
Enlarged blind spot	34
Altitudinal field defect	6
Hemianopia	29
Central scotoma	26
Pie in sky	2
Pie on floor	2
Advanced field defect	62

Table 4: MRI findings in 128 of the 170 patients

Neuroimaging findings	Number of patients
Benign intracranial hypertension	32
Space-occupying lesion	38
Multiple sclerosis	9
Cerebral infarct	13
Cerebral hemorrhage	5
Non-specific lesions	3
Optic neuritis	13
Traumatic optic atrophy	2
TB meningitis	4
Inflammatory optic neuropathy	1
Cortical venous sinus thrombosis	8

Table 5: Final etiological diagnoses in our series

Final diagnosis	Number of patients
Benign intracranial hypertension	32
Pituitary adenoma	18
Other CNS space-occupying lesions	26
Optic neuritis/multiple sclerosis	15
Cortical venous sinus thrombosis	15
Malignant hypertension	12
Ischemic optic neuropathy	6
Toxic optic neuropathy	7
Meningitis with optic nerve swelling	8
Traumatic optic neuropathy	5
Cerebral infarct	3
Cerebral hemorrhage	1
Inflammatory optic neuropathy	4
Hereditary optic neuropathy	1
AV malformation	1
Chronic relapsing inflammatory optic neuropathy	2
Gas geyser syndrome	1
POEMS	1
Neuromyelitis optica	1
Tolosa hunt syndrome	1
ADEM	1
Unclassified	9

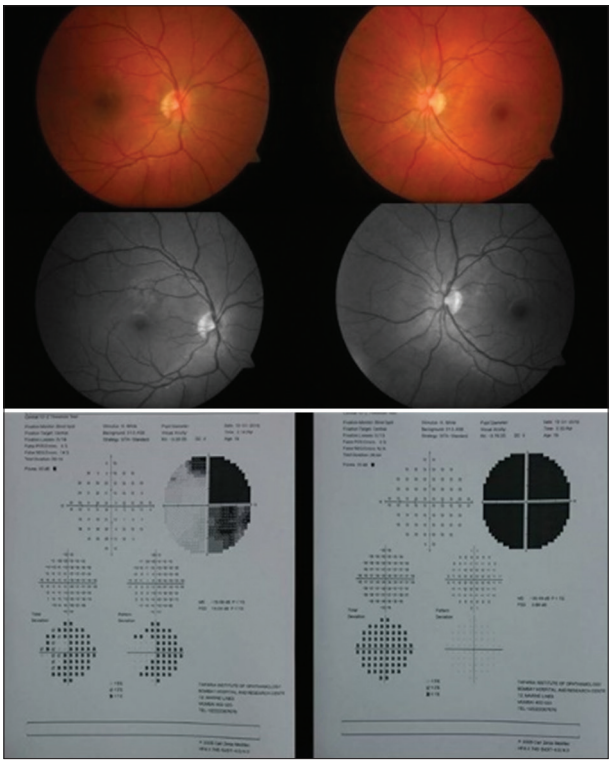

Figure 1: Fundus picture of both eyes of a 19-year-old lady showing mild disc hyperemia with edema. Visual field of the same 19-year-old lady showing an advanced field loss in the left eye and a predominantly superior defect involving the central 10 degrees in the right eye

Sub-group analysis of the clinical impression based on neuro-ophthalmic examination

Papilledema

There were 84 patients with a clinical impression of papilledema.

Of the 168 eyes of these 84 patients, 152 eyes (90.47%) had a vision of 6/12 or better with a normal color vision in 144 eyes (85.71%). Of the 101 eyes who had an interpretable visual field, 41 eyes had a normal visual field. An enlarged blind spot was seen in 32 eyes (53.33%) and was the most common visual field defect seen in the 60 eyes that had a visual field defect. Of these 84 patients, 29 (34.52%) had a BIH. Pituitary adenoma was the most common SOL in our series but accounted for only (1.19%), that is, one of the 84 patients who had papilledema. The other SOLs with 19 patients (22.61%) and cortical venous sinus thrombosis with 15 patients (17.85%) accounted of the remaining patients who presented with papilledema.

ON

There were 16 patients in our series with a clinical impression of ON.

A part from conspicuously having an affected color vision in 17 (53.12%) of the 32 eyes, these patients also predominantly showed a central scotoma in their visual fields – 15 of the 26 eyes (57.69%) who performed a interpretable visual field. Of these 16 patients, 15 had an etiological diagnosis with ON/MS being the most common diagnosis made in ten patients (62.5%), followed by toxic optic neuropathy in four patients and anterior ischemic optic neuropathy in one patient.

Optic atrophy

Of the 38 patients with varying degrees of optic atrophy, color vision affection was noted in 32 of the 76 eyes (42.10%). Herein, it was noted that pituitary adenoma was the most common cause of optic atrophy, with 16 of the 38 patients with optic atrophy (42.10%) having a pituitary adenoma as the final diagnosis.

Chart 3 shows the various etiologies which gave rise to optic atrophy in our patients.

Hypertensive retinopathy

Of the 13 patients, with a clinical impression of accelerated hypertensive retinopathy being the cause of disc edema; 12 patients indeed had a final diagnosis of malignant hypertension with one patient having a corona radiata infarct. Of the 26 eyes of these 13 patients, 20 eyes (76.92%) had a vision of 6/12 or better at presentation with ten patients being

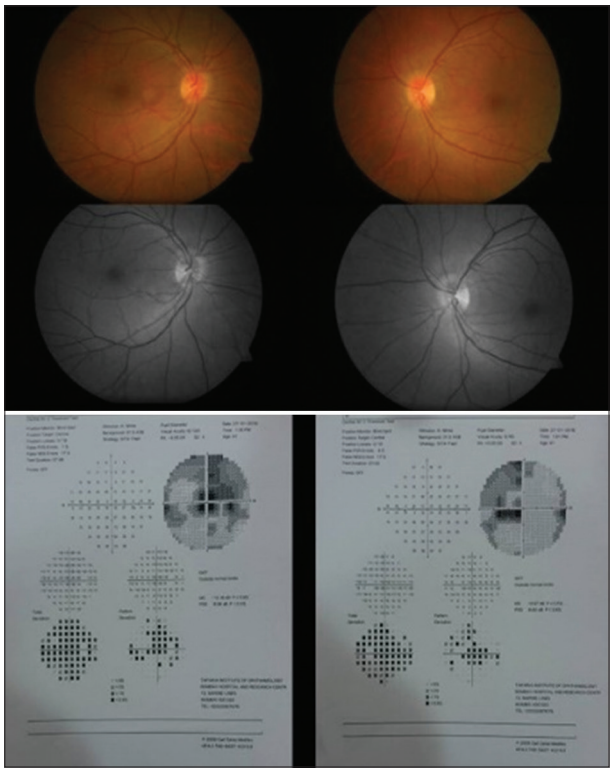

Figure 2: Fundus picture of both eyes of a 41-year-old gentleman showing bilateral mild disc hyperemia with mild edema. Visual field of the same 41-year-old gentleman showing bilateral predominantly central scotomas with central visual field affection

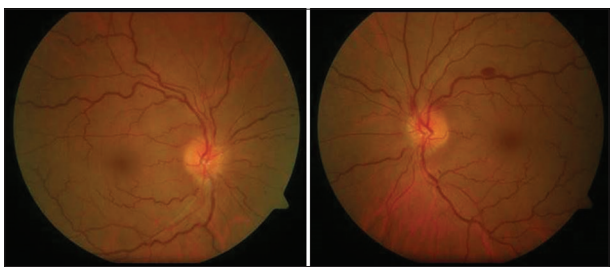

Figure 3: Fundus picture of both eyes of a 35-year-old gentleman showing moderate disc edema right eye more than left eye

known hypertensive, that is, three patients had no past history of hypertension and presented with accelerated malignant hypertension. All these three patients were young hypertensive being 24 years of age.

Optic neuropathy

Seven patients (4.11%) had toxic optic neuropathy as their final diagnosis. Six of these seven patients were on anti-tuberculosis treatment with ethambutol and one patient was using anti-leprosy drugs. Six of the 14 eyes (42.85%) of these seven patients had a presenting visual acuity of counting finger 3 m or lesser. The optic nerve findings included mild

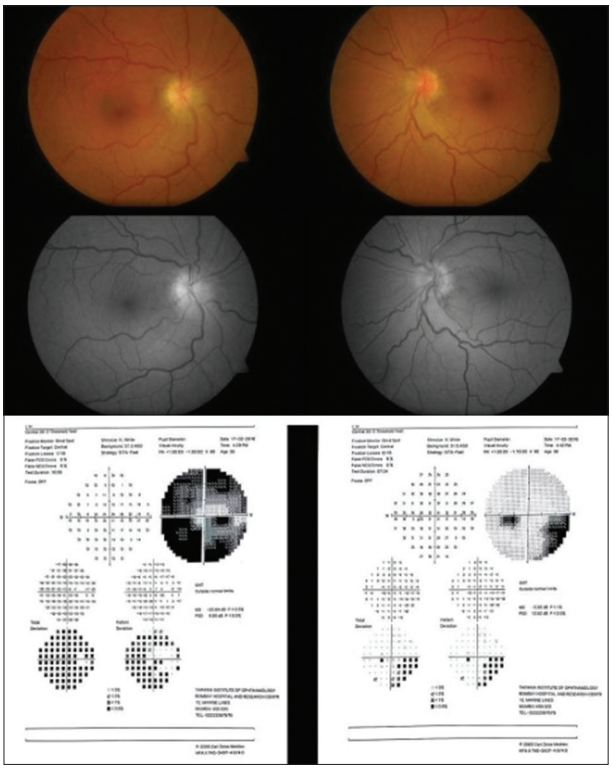

Figure 4: Fundus picture of both eyes of a 36-year-old lady showing moderate disc edema in both eyes with the early secondary optic atrophy in the right eye. Visual field of the same 36-year-old lady showed bilateral affection with an advanced field defect in one eye and enlarged blind spot and a scotoma in the left eye

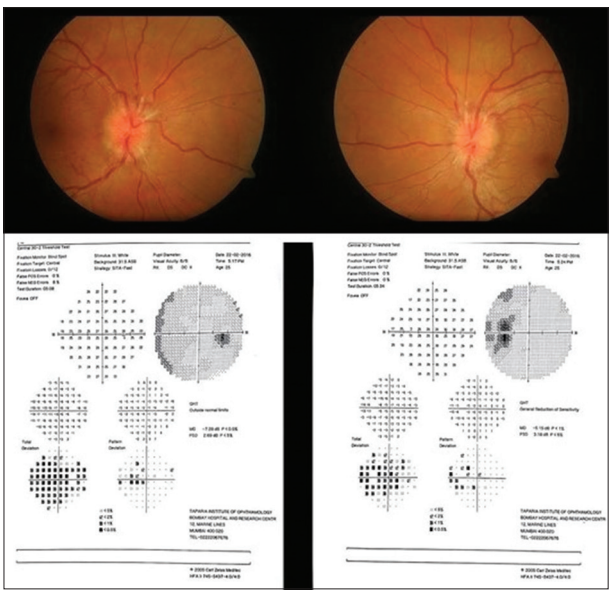

Figure 5: Fundus picture of both eyes of a 25-year-old gentleman showing bilateral moderate to severe disc edema. Visual field analysis of the same 25-year-old gentleman showed enlarged blind spot in the left eye

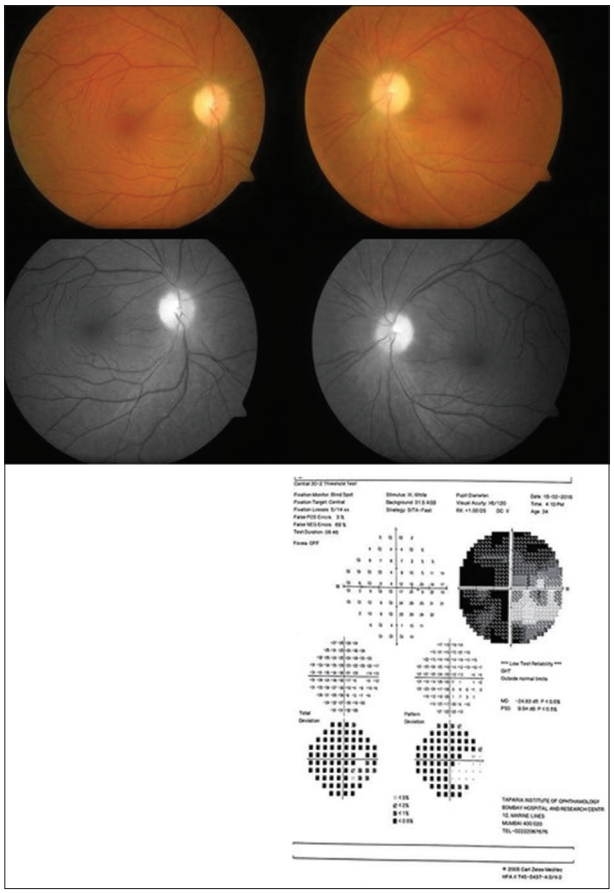

Figure 6: Fundus picture of both eyes of a 33-year-old gentleman showing bilateral chronic mild disc edema with secondary optic atrophy. Visual field of the same 33-year-old gentleman which was possible only in the right eye showed a residual inferior island of vision

hyperemia in two eyes; mild disc swelling in three eyes; moderate edema in five eyes; and optic nerve pallor in four eyes. Of the ten eyes of five patients who were requested to have a visual field analysis, a central/centrocecal scotoma was noted in five eyes (50%) and an advanced field defect including the central field was noted in five eyes (50%). The single patient classified as hereditary optic neuropathy had ethambutol induced Leber's Hereditary Optic Neuropathy (LHON).

Representative cases

Case 1 [Figure 1]

A 19-year-old lady presented with right occipitofrontal headache and vertigo.

Right Eye – 6/9, N6 with normal color vision

Left Eye – Counting finger 2 m, N36, and inability to identify demo plate of Ishihara Chart.

Clinical impression was ON (Left Eye>Right Eye)

MRI brain confirmed demyelination and further diagnosis of MS.

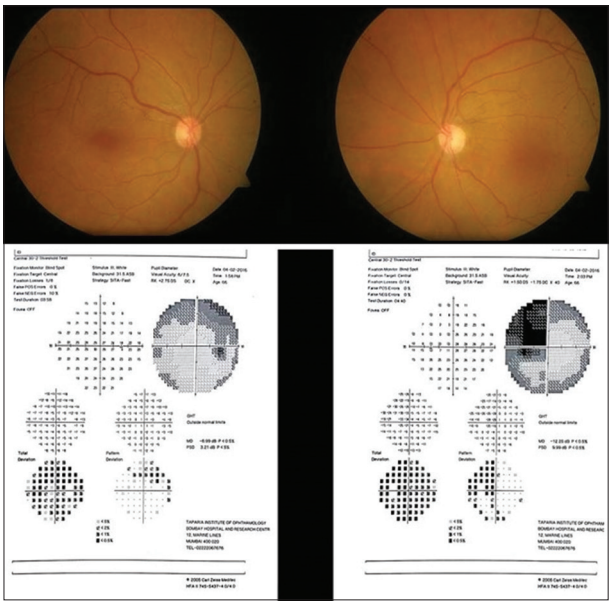

Figure 7: Fundus picture of both eyes of a 65-year-old lady showing mild temporal optic nerve pallor in the right eye with partial primary optic atrophy in the left eye. Visual field of the same 65-year-old lady showed a quadrantanopia in the left eye with a superior field defect in the right eye

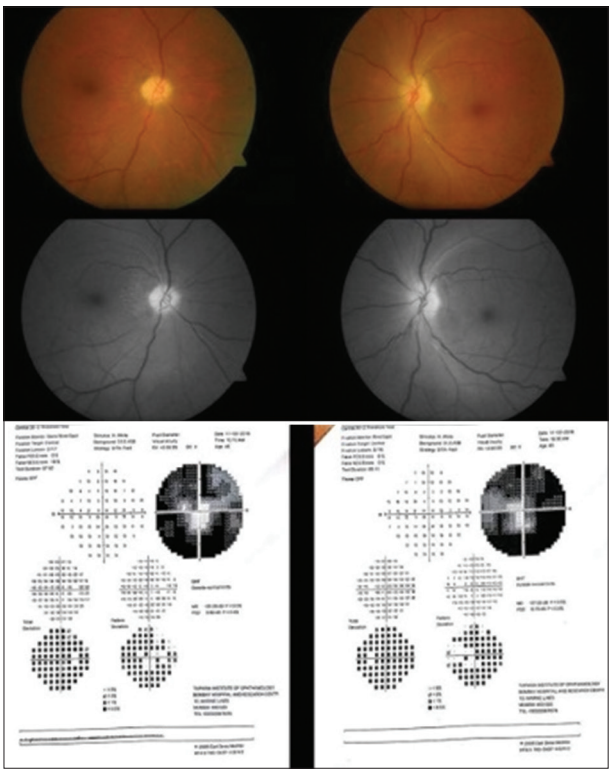

Figure 8: Fundus picture of both eyes of a 45-year-old gentleman showing mild disc edema with pallor in both eyes left eye more than right eye. Visual field of the same 45-year-old gentleman showing bilateral advanced field loss with a central sparing

Case 2 [Figure 2]

A 41-year-old gentleman presented with sudden bilateral painless loss of vision.

Known to be suffering from Koch's Lymphadenitis on treatment

Right Eye – Counting finger 3 m, N36

Left Eye – 6/60, N36

Cannot identify demo plate on Ishihara Charts.

Clinical impression was optic neuropathy (Left Eye>Right Eye) related to use of ethambutol – toxic optic neuropathy.

MRI brain and orbits were normal.

Case 3 [Figure 3]

A 35-year-old gentleman with bilateral sub-dural hemorrhage.

Right Eye – 6/9, N6 with normal color vision

Left Eye – 6/6, N6 with normal color vision.

Clinical impression was moderate papilledema left more than right eye.

Case 4 [Figure 4]

A 36-year-old lady on treatment as BIH presented with decrease of vision right eye since 1 year and left eye since 2 months.

Right Eye – 6/9, N6, very hazy

Left Eye – 6/6, N6

Color vision normal in both eyes.

Clinical impression was moderate papilledema in both eyes with the right eye secondary optic atrophy in a case of BIH.

Case 5 [Figure 5]

A 25-year-old gentleman with sudden onset squinting and double vision.

Right Eye – 6/6, N6

Left Eye – 6/6, N6

Normal Color Vision.

Clinical impression was moderate-to-severe papilledema in both eyes

MRI brain showed thalamic SOL.

Case 6 [Figure 6]

A 33-year-old gentleman, known case of superior sagittal sinus thrombosis, posted for optic nerve sheath fenestration.

Right Eye – Counting finger 1 m

Left Eye - Hand movements close to face.

Clinical impression was bilateral chronic papilledema with secondary optic atrophy.

Case 7 [Figure 7]

A 65-year-old lady operated for pituitary adenoma 8 years back presented with headache which was gradually progressive over past few months.

Right Eye – 6/6, N6

Left Eye – 6/18 with inability to identify plates 2, 3 on Ishihara Chart.

Clinical impression was partial primary optic atrophy in both eyes left eye more than right eye.

Case 8 [Figure 8]

A 45-year-old gentleman presented with sudden bilateral painless loss of vision first in the right eye and then in the left eye over 1 week

Known to be suffering from Hansen's Disease on treatment

Right Eye – 6/18, N10

Left Eye – 6/18, N12

Normal color vision in both eyes.

Clinical impression was optic neuropathy (Left Eye >Right Eye) in a known case of Hansen's disease – Unlikely drug related and more likely ischemic.

DISCUSSION

Optic disc edema manifests as swelling of the unmyelinated nerve fibers. The edema results from impaired axoplasmic flow from any cause, including increased intracranial pressure, local mechanical compression, ischemia, and inflammation. Optic disc and retinal vascular changes can also accompany optic disc edema.

Regardless of cause, the following clinical features of optic disc edema may be observed

1. Elevation of the nerve head with variable filling in of the physiologic cup; retinal vessels may appear to drape over the elevated disc margin
2. Blurring of the disc margins
3. Peripapillary NFL opacification; the NFL becomes grayish white and opalescent with feathered margins, obscuring portions of the retinal vessels that course within this level of the retina
4. Hyperemia and dilation of the disc surface capillary net
5. Retinal venous dilation and tortuosity
6. Peripapillary hemorrhages, exudates, or cotton wool spots
7. Retinal or choroidal folds or macular edema.

A PUBMED search on non-glaucomatous optic nerve disorders brings up a number of articles for study. Osaguona and Okeugbemen in a retrospective review of the medical records of all adult patients aged 16 years and above with non-glaucomatous optic atrophy at the eye clinic of the University of Benin Teaching Hospital over a 4-year period showed that 104 patients had non-glaucomatous optic atrophy with a male: female ratio of 1.3:1 and a majority (75%) of the patients were in the age range of 31–70 years.[2] Chorioretinal disease (23.1%), trauma (13.5%), toxic-nutritional (7.7%), and compressive lesions (4.8%) were the most common among the known etiologies.[2] Another pilot study conducted in the eye clinic, University College Hospital, Ibadan by Ogun and Adediran showed that 46.8% of new cases presenting to the neuro-ophthalmology unit, had non-glaucomatous optic neuropathy (NGON), in which the precise etiology of optic neuropathy was never diagnosed.[3] Findings from the Beijing Eye Study reflect an association between intracranial abnormalities and/or systemic diseases and non-glaucomatous optic nerve damage and may

alert the ophthalmologist to consider non-glaucomatous optic nerve damage, even if relatively subtle, as a factor potentially associated with an increased risk for death.[4] Articles specifically from our country include a previous case series by Raju and Khadder, where optic atrophy was a presentation in 18% of patients with intracranial SOLs.[5] Causes of optic neuropathy in India also include traumatic optic neuropathy which was responsible for 7 (7%) cases of optic atrophy as shown in another case series from India by Chaddah *et al*.[6]

Of all the optic nerve related disorders, looking purely at optic atrophy as a clinical sign, we know that it results from disease processes that cause irreversible damage to the ganglion cells and axons in the anterior visual pathway. The diagnosis of optic atrophy is based on the findings of disc pallor, with associated changes in the integrity of the retinal nerve fiber layer and retinal vessels, and visual dysfunction (vision and/or visual field loss) attributable to the optic nerve.[7] The etiology of optic atrophy is broad and may be a presentation of life-threatening processes.[8] In a previous case series from India, optic atrophy was a presentation in 18% of patients with intracranial SOLs.[5] In our series, it was noted that pituitary adenoma was the commonest cause of optic atrophy; with 16 of the 38 patients with optic atrophy (42.10%) having a pituitary adenoma as the final diagnosis. Other causes of optic neuropathy include traumatic optic neuropathy. In another previous case series from India, traumatic optic neuropathy was responsible for 7 (7%) cases of optic atrophy.[6] Optic atrophy in trauma may result either from indirect injury to the optic nerve due to transmitted shear forces to the nerve in closed head injury such as blunt trauma, or from direct injury to the optic nerve in penetrating injuries.[9] The most common form of injury to the optic nerve is by indirect injury with an incidence of 0.5–5% of all closed head injuries.[10,11] Common causes of traumatic optic neuropathy include road traffic accidents and falls; other less common causes include assaults, stab wounds, gunshots, and trivial injuries.[9] The patient with traumatic optic neuropathy commonly presents with a history of visual loss following head trauma and may have other injuries such as fracture to the base of the skull and multiple facial fractures with associated ocular motor nerve palsies and optic atrophy usually results 8 weeks after injury.[12] It is worth noting that of the 170 patients in the current series only three patients had trauma as a cause of optic neuropathy. This is likely to be due to a bias introduced by the fact that this was a clinic-based study and, hence, patients with traumatic optic neuropathy who first required an intensive care setting due to multiple face and skull fractures and are likely to have been left out of this study. Other known toxic-nutritional causes include anti-tuberculous medications such as ethambutol and isoniazid, methanol, tobacco, amiodarone, deficiencies of Vitamin B12 or folate, lead, quinine, and tamoxifen among many others.[5,13,14] Our single case of ethambutol-induced LHON highlights the important phenomenon of conversion in LHON. Broadly, as highlighted by our study, NGON is only a clinical impression and not a diagnosis; as it results from various etiologies;[6] with some cases of like those of optic neuropathy, being amenable to treatment and a good visual outcome.[15,16]

Etiological groupings or etiological cause of the optic neuropathy needs to be determined based on documentary evidence from case notes that identify a clear etiology of optic nerve dysfunction from clinical examination, ancillary investigations, or neuroimaging.[16]

One of the biggest limitations of this study is that it is in a hospital-based setting and, hence, is unlikely to be completely representative of the etiology of non-glaucomatous optic nerve head disorders in a community-based setting.

This is obvious by the fact that despite seeing many patients with temporal optic nerve pallor in our clinics, we were not able to make a stand-alone etiological diagnosis of "Nutritional Optic Atrophy" with the other causes overshadowing it in the hospital-based neuro-ophthalmic practice.

Nonetheless, our study highlights a variety of non-glaucomatous optic nerve head disorders are encountered in clinical neuro-ophthalmic practice. Some of the rare cases in which there was interdisciplinary co-ordination among various departments included a 32-year-old lady who had a neurological collapse with seizures in the bathroom followed by loss of vision to as low as finger counting 2 m in the left eye with a fundus picture of optic neuropathy which was diagnosed to be gas geyser syndrome which is an important preventable cause of disabling neurological events.[17] Another such patient was a 29 year gentleman with POEMS syndrome which is a paraneoplastic syndrome due to an underlying plasma cell neoplasm. The major criteria for the syndrome are polyradiculoneuropathy, clonal plasma cell disorder, sclerotic bone lesions, elevated vascular endothelial growth factor, and the presence of Castleman disease. Minor features include organomegaly, endocrinopathy, characteristic skin changes, papilledema, extravascular volume overload, and thrombocytosis. Diagnoses are often delayed, because the syndrome is rare and can be mistaken for other neurologic disorders, most commonly chronic inflammatory demyelinating polyradiculoneuropathy.[18]

Learning and recommendations from the study and previous review of literature

A patient with optic nerve disease should be thoroughly evaluated as it may be a treatable condition or a presentation of a life-threatening condition. With thorough history and examination, the etiology of optic nerve disease can be determined in most cases. The level of optic nerve function should be determined by visual acuity, color vision, brightness sensitivity testing, confrontation, and a formal perimetry. There is usually a relative afferent pupillary defect in unilateral disease or bilateral, asymmetric disease. The area of disc pallor (segmental or diffuse pallor), retinal nerve fiber layer loss, and the integrity of the retinal vessels should be assessed. A full neurologic examination should also be done to exclude other neurologic deficits. Laboratory investigation should be done as indicated from the clinical evaluation. In cases of unexplained

optic atrophy, neuroimaging with contrast of the orbit and brain with attention paid to the course of the optic nerve, chiasm, and retrochiasmal visual pathway should be done. Follow-up of the patient should be with visual acuity, color vision, quantitative visual field tests, and fundus photographs.

CONCLUSIONS

The two most common presenting complaints encountered in clinical neuro-ophthalmic practice are headache and progressive deterioration of vision. In Indian adults presenting to a specialized tertiary care hospital, non-glaucomatous optic nerve disorders are commoner in females with the mean age of presentation to clinic being 39 years. The most common optic nerve finding/pathological pattern seen by the neuro-ophthalmologist in the clinic is disc edema. Papilledema is the most common pathology seen in the optic nerve encountered followed by optic atrophy and ON. BIH is the most common etiology among all the various etiologies that can result in optic nerve head disorders in India. Most importantly, this study highlights etiopathogenesis of the optic nerve findings with color vision assessment and the visual field defects and how various etiologies give rise to different pathologies in the optic nerve which are seen as pathological patterns on examination of the optic nerve. A good understanding of these findings can help the neuro-ophthalmologist understand the etiopathogenesis of the optic nerve disorders and help the neurologist and neurosurgeon pinpoint the correct etiology resulting in better patient care.

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