Original Research Paper

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Ophthalomology

CLINICAL PROFILE OF ATYPICAL OPTIC NEURITIS – CASE SERIES.

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KEYWORDS :

INTRODUCTION:

Optic neuritis (ON) is a group of conditions that involve inflammation of the optic nerve. Patients present with partial to complete vision loss within a few days of onset and also have dyschromatopsia with pain on ocular movements at presentation. Various etiologies can be responsible for ON. Presentation of ON is broadly grouped into typical ON and atypical ON**.** A typical ON follows a specific clinical pattern and is usually related to a demyelinating lesion that can be isolated or associated with multiple sclerosis (MS). A clinical pattern of ON is considered as atypical ON if it differs from this presentation and has a long list of differentials and includes neuromyelitis optica spectrum disorder, infectious causes, and autoimmune causes. Diagnostic procedures are CBC/ESR/CRP/CSF analysis, anti AQP4 antibodies, anti-MOG antibodies, antinuclear double-stranded DNA & Anti neutrophil cytoplasmic antibodies, CE MRI of brain with orbits and whole spine, OCT of the peripapillary retinal nerve fiber layer (pRNFL) and ONH and VEP [visually evoked potential].

The Optic Neuritis Treatment Trial (ONTT) was a randomized, controlled clinical trial designed to evaluate the efficacy and safety of different corticosteroid regimens in the treatment of acute optic neuritis (ON). The study compared three treatment arms: oral placebo, intravenous methylprednisolone (IV-MP) 250 mg every 6 hours for 3 days followed by oral prednisone at a dose of 1 mg/kg for 11 days, and oral prednisone alone at 1 mg/kg daily for 14 days. The trial aimed to determine the most effective approach in improving visual outcomes and minimizing recurrence in patients with acute ON. IV steroid-treated patients recovered faster within the first 4-6 weeks post onset. However at 6 months to 10 years out, there was no statistical difference in final visual outcome between IV-steroid treated patients and placebo. Oral steroids in conventional doses alone are contraindicated for acute ON because of a higher rate of recurrences of ON. Patients who do not respond to initial IV steroid therapy (e.g., NMO or MOG) may benefit from early plasma exchange (PLEX) or IVIG. Azathioprine/mycophenolate mofetil [long-term immune suppression /prevent recurrence]. RITUXIMAB [anti CD 20] – decreases relapse and increases stabilization of disability in NMO.

AIMS AND OBJECTIVES:

The purpose of this study is to study the clinical presentations of atypical optic neuritis among patients who came to a tertiary health care centre i.e., the government general hospital, Guntur.

MATERIALS AND METHOD:

Patients presenting with clinical features of atypical optic neuritis were studied over a period of 8 months in the government general hospital, Guntur. All patients were evaluated with detailed history, clinical examination, required investigations, and regular follow-up of prognosis were done. Informed and written consent from patients were taken. Data regarding patients' age, sex, and presenting features will be recorded in patients who are presenting in the government general hospital, Guntur.

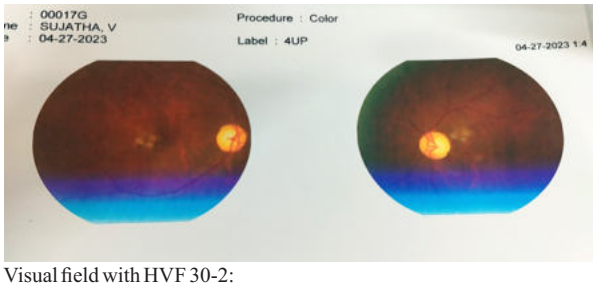
CASE 1:

A 34-year-old female patient presented with c/o diminution of vision in both eyes [Left>Right] for 3 months which is gradual and progressive in nature, associated with headache. H/O diminution of vision in LE for 2 months followed by which she had diminution of vision in RE for 1 month.

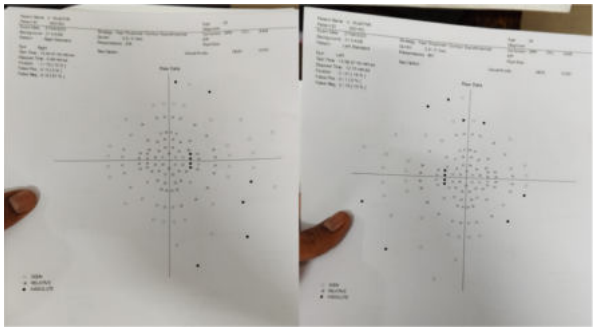
No h/o fever, vomiting, seizure, or any other systemic illness.

On examination:

On examination, the best corrected visual acuity (BCVA) was 6/12 in the right eye (OD) and 6/36 in the left eye (OS). Colour vision was normal in the right eye, whereas the left eye was able to identify only primary colours, indicating a degree of colour vision deficiency. The anterior segment appeared normal in both eyes, with no abnormalities detected. Pupillary examination revealed a round, reactive pupil to light in the right eye, while the left eye demonstrated a grade 2 relative afferent pupillary defect (RAPD), suggestive of optic nerve dysfunction. The lenses were clear in both eyes. Fundus examination revealed clear media in both eyes (OD and OS). The optic discs in both eyes appeared mildly pale with distinct margins. The cup-to-disc ratio (CDR) was noted to be 0.6:1 bilaterally, with a healthy neuroretinal rim (NRR) and no signs of glaucomatous damage. The disc margins were clearly defined in both eyes. Retinal vessels appeared normal, and the foveal reflex (FR) was present in both eyes, indicating a preserved macular structure.



Visual field with HVF 30-2:



Right eye: Non-specific scattered depressions were noted in the pattern deviation plot

Left eye**:** Near normal fields noted on repeat fields of left eye [due to learning curve]

MRI brain showed E/O tortuous course of B/L pre-chiasmatic segments of optic nerve noted with subtle STIR hyperintensity on both sides with prominent perineural sheath, suspected IIH/DD- B/L Atrophic optic neuritis. Anti-MOG antibody was positive and anti AQP-4 antibody – negative

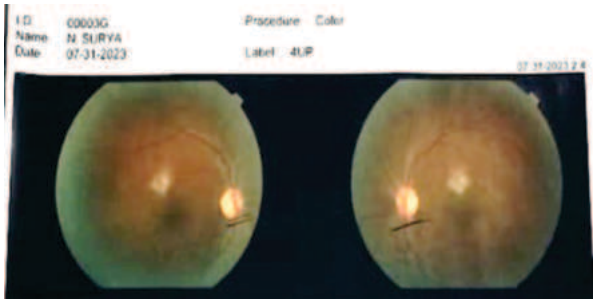
- Patient was admitted in neurology ward and got treated with IV pulse methylprednisolone [1gm] for 3 days followed by oral prednisolone 60mg OD. Injection RITUXIMAB was also administered.
- No visual improvement was noted following treatment.

CASE 2:

A 22-year-old male patient was referred from the neurology ward with H/o painful loss of vision in Both eyes for 1 month which is gradual and progressive K/C/O psychiatric disorder on anti-psychotic drugs [Risperidone] and seizure disorder on sodium valproate 500mg. History of chronic alcoholic consumption and smoking habit was present and no history of fever, vomiting, or any other systemic illness.

Clinical Examination:

On examination, the best corrected visual acuity (BCVA) in both eyes (OD and OS) was perception of light positive (PL+), indicating severely reduced vision. The anterior segment was normal in both eyes with no detectable abnormalities. Pupils were mid-dilated and reacted sluggishly to light bilaterally, suggesting possible optic nerve or severe retinal involvement. The lenses in both eyes were clear, with no evidence of cataract or opacities. Fundus examination revealed clear media in both eyes (OD and OS). Disc pallor was noted bilaterally, suggestive of optic nerve atrophy or previous optic neuropathy. Retinal vessels appeared normal in both eyes, and the foveal reflex (FR) was present bilaterally, indicating an intact macular architecture.



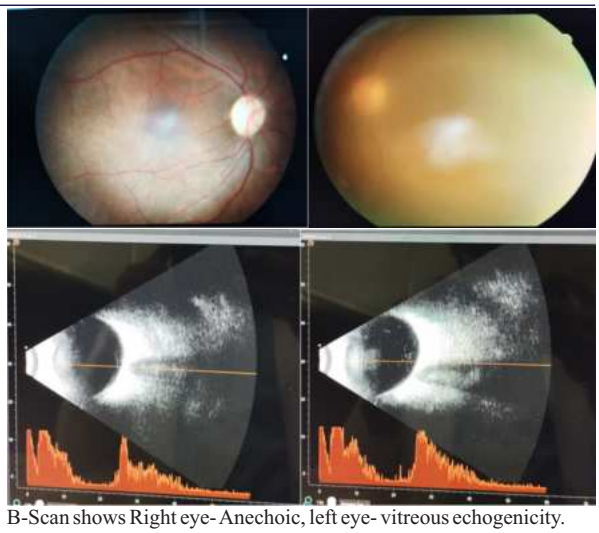
CE MRI brain with Both orbits– non-enhancing areas of hyperintense in brain stem and right cerebellar hemisphere - Demyelination/ encephalitis subtle FLAIR hyperintense areas in bilateral optic nerves– probably B/L optic neuritis. CE MRI whole spine shows non-enhancing subtle T2 hyperintense areas in the spinal cord from D8 to D11 probably demyelination. Anti-MOG antibody was positive and Anti AQP-4 antibody was negative Patient was admitted in neurology and got treated with IV pulse methylprednisolone [1gm] for 3 days followed by oral prednisolone 60mg OD. Vision improvement to hand movements was noted.

CASE 3:

A 53-year-old female patient was referred from the neurology ward with H/O loss of vision in BE for 15 days which is gradual and progressive and associated with retrobulbar pain. H/O RE cataract surgery done before 1 year. No h/o fever, vomiting, seizure, or any other systemic illness.

On Examination:

On examination, the best corrected visual acuity (BCVA) was perception of light (PL) positive in both eyes (OD and OS), indicating profound visual impairment. The anterior segment appeared normal in both eyes with no abnormalities detected. Pupillary reactions were sluggish to light bilaterally, suggestive of possible optic nerve or retinal pathology. The right eye (OD) was pseudophakic, indicating previous cataract surgery with intraocular lens implantation, while the left eye (OS) showed a greyish-white lens opacification consistent with a mature or hypermature cataract. Fundus examination of the right eye (OD) revealed clear media with pseudophakia, disc pallor suggestive of optic nerve atrophy, normal retinal vessels, and a present foveal reflex, indicating preserved macular structure. In contrast, the left eye (OS) had hazy media, making detailed assessment difficult. The optic disc, vessels, and macula could not be clearly visualized due to the media opacity.



B-Scan shows Right eye- Anechoic, left eye- vitreous echogenicity.

MRI brain showed bilateral optic neuritis and anti-MOG antibody was positive and anti AQP-4 antibody was negative. Patient was admitted to neurology and got treated with IV pulse methylprednisolone [1gm] for 3 days followed by oral prednisolone 60mg OD. Visual acuity after 1 month was improved to 6/60 [OD], CF 1M[OS]

DISCUSSION:

Optic neuritis (ON) is a group of conditions that involve inflammation of the optic nerve. Patients present with partial to complete vision loss within a few days of onset and also have dyschromatopsia with pain on ocular movements at presentation. Various etiologies can be responsible for ON. Presentation of ON is broadly grouped into typical ON and atypical ON. A typical ON follows a specific clinical pattern and is usually related to a demyelinating lesion which can be isolated or associated with multiple sclerosis (MS).A clinical pattern of ON is considered as atypical ON if it differs from this presentation and has a long list of differentials and includes NMOSD, infectious causes, and autoimmune causes.

The differential diagnosis for acute optic neuropathy includes several conditions such as typical and atypical optic neuritis, papilledema, Leber hereditary optic neuropathy, and ischemic optic neuropathy. A comprehensive diagnostic work-up is crucial and includes blood investigations like complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and cerebrospinal fluid (CSF) analysis. Specific antibody tests such as anti-aquaporin-4 (AQP4), anti-myelin oligodendrocyte glycoprotein (MOG), antinuclear double-stranded DNA antibodies, and anti-neutrophil cytoplasmic antibodies are essential to rule out autoimmune and inflammatory etiologies. Imaging studies such as contrast-enhanced magnetic resonance imaging (CE-MRI) of the brain, orbits, and whole spine are important to identify demyelination or structural causes. Optical coherence tomography (OCT) of the peripapillary retinal nerve fiber layer (pRNFL) and optic nerve head (ONH), along with visually evoked potential (VEP) testing, provide functional and structural assessment of the optic nerve.

Management of optic neuritis is guided by the findings of the Optic Neuritis Treatment Trial (ONTT), a randomized controlled clinical trial designed to evaluate the efficacy and safety of oral prednisone versus intravenous methylprednisolone followed by oral prednisone, in comparison to oral placebo. The three arms of the trial included (1) oral placebo, (2) intravenous methylprednisolone 250 mg every 6 hours for 3 days followed by oral prednisone 1 mg/kg for 11 days, and (3) oral prednisone 1 mg/kg alone for 14 days. The primary outcome measures were contrast sensitivity and visual field, while secondary outcomes included visual acuity and color vision, with patients followed up for a minimum of six months.

The outcomes of the ONTT revealed that patients treated with intravenous steroids showed a significantly faster visual recovery within the first 4 to 6 weeks post-onset. However, by six months to ten years post-treatment, there was no statistically significant difference in the final visual outcomes among the IV steroid group and the placebo group. Importantly, oral steroids alone, in conventional doses, were found to be contraindicated due to a higher recurrence rate of optic

neuritis episodes. Patients who do not respond to initial IV steroid therapy (e.g., NMO or MOG) may benefit from early plasma exchange (PLEX) or IVIG. Azathioprine/mycophenolate mofetil [long-term immune suppression /prevent recurrence. RITUXIMAB [anti-CD 20] – decrease relapse and increase stabilization of disability in NMO

CONCLUSION:

Three cases of optic neuritis affecting both eyes have been reported with varied presentations and visual acuity ranging from 6/60 to perception of light associated with abnormal pupillary reflex. Fundus findings showed a pale disc in one case and disc oedema with pallor in two cases. All are positive for Anti MOG-IgG and negative for Anti aquaporin-4-IgG. Patients were started on IV steroids followed by oral steroids and IV Rituximab in one case, under neurologist guidance. Visual improvement was noted in two of them.

Etiology of atypical ON varies widely and calls out for different treatment options. Various biomarkers have been identified to confirm and distinguish atypical optic neuritis from others in order to provide better outcomes.

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