

## Case Report

# Adult-onset bilateral optic neuritis: a rare presentation

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## ABSTRACT

A rare case of adult-onset bilateral optic neuritis without associated autoimmune or infectious disorders has been reported in this study. A 19-year-old male described sudden bilateral diminution of vision (sequential) with headache with no other remarkable history. Ophthalmological findings revealed asymmetrically reduced visual acuity on the initial visit. Fundus examination showed nerve fibre layer oedema (more in right eye). Radiological examination was suggestive of bilateral, asymmetrical optic neuritis. Visual evoked potential tests suggested asymmetrical and sequential P100 delay. No auto-immune/infectious aetiology was found. The condition was diagnosed as an atypical presentation of optic neuritis with sequential bilateral involvement. Patient was treated with intravenous methyl prednisolone followed by tapering oral doses owing to the bilaterality of the involvement. A careful follow-up is being performed. The condition involving bilateral inflammation of the optic nerves can have hazardous outcome and hence prompt, careful and comprehensive diagnostic and treatment approach is necessitated.

**Keywords:** Optic neuritis, Visual acuity, Visual evoked potentials

## INTRODUCTION

Optic neuritis (ON) is described as inflammation of the optic nerve, which is mostly idiopathic. It is also suggested to be associated with demyelinating lesions, autoimmune disorders, infectious and inflammatory conditions. Diagnosis of ON is based on the clinical examination. MRI (magnetic resonance imaging) is suggested to evaluate the risk of multiple sclerosis as clinically silent white matter lesions may be evident. Visual evoked potentials show delayed latencies in the involved eye (long after acute attack, acute stage shows absent waveforms).

The classic presentation with visual loss, periocular pain, dyschromatopsia and unilaterality define typical form of optic neuritis. The natural course of most unilateral acute optic neuritis is described as sudden onset of visual loss associated with pain on eye movements, which reaches its maximum deficit over 1-7 days. On the other hand,

simultaneous or sequential bilateral ON is an atypical feature. Bilateral optic neuritis is usually thought to affect children and is often associated with infectious demyelination.<sup>1</sup>

Adult onset optic neuritis is typically unilateral and is commonly linked to multiple sclerosis. In adults, simultaneous or sequential bilateral acute optic neuritis has been considered rare, particularly in individuals without known systemic inflammatory or autoimmune disorders. The study aims to report such a rare case of adult-onset bilateral optic neuritis without associated autoimmune or infectious disorders; early diagnosis and appropriate management in which, can contribute to optimal outcomes.

## CASE REPORT

A 19 year-old male patient had sudden diminution of vision in both the eyes (loss of vision in right eye was

reported to be preceded by left) along with headache. No history of trauma was present. No muscle weakness or symptoms of paralysis. Past history was not remarkable with no episode of illness with fever and rashes or other significant disease. Clinical examination revealed normal neurological examination. Ophthalmological examination revealed visual acuity of 6/60 and 6/24 in right and left eyes respectively on the day of presentation. Afferent pupillary defect (APD) was present in both eyes (right>left). Fundus examination showed blurring of disc margin as well as nerve fibre layer oedema, which was more pronounced in right eye compared to left eye. Colour vision was abnormal in both eyes.

Visual field examination was performed on same day by Humphrey Field Analyser which showed diffuse depression with central and paracentral scotoma in both eyes.

### Radiological findings

MRI of brain and orbit with contrast was performed. The right optic nerve showed subtle thickening with mild enhancement at orbital apex while significant enhancement at right optic chiasma was found. The left optic nerve showed diffuse thickening and enhancement up to left optic nerve head. Features were suggestive of bilateral, asymmetrical optic neuritis with no evidence of intracranial SOL (space occupying lesion).

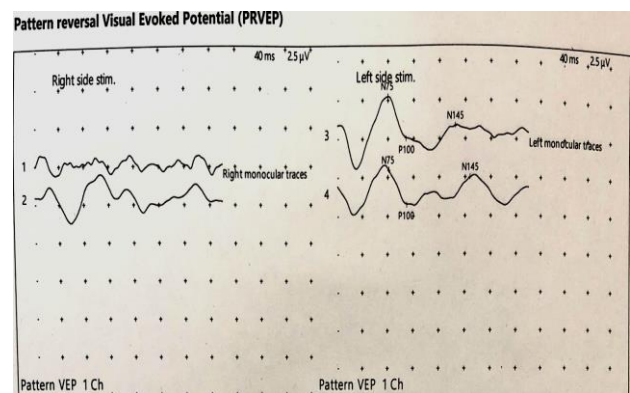
Since it was an atypical presentation with bilateral involvement of optic nerves, arising the suspicion of secondary involvement of optic nerves, presence of aetiology was sought for.<sup>2-5</sup> Investigations (baseline chest X-ray, serology for infectious aetiology, blood culture and CSF examination), however did not indicate infectious or autoimmune aetiology.

### Visual evoked potential findings

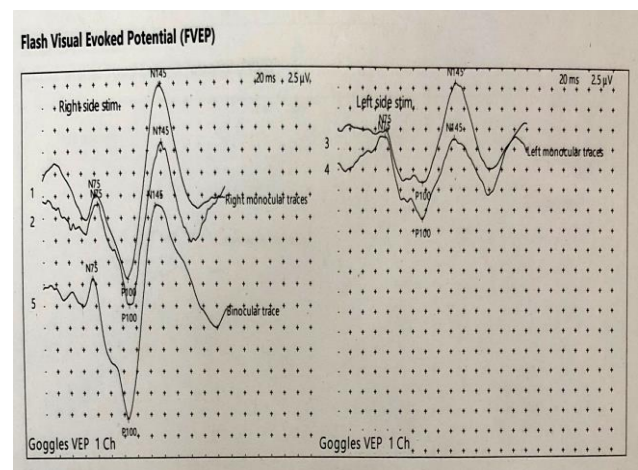
Visual evoked potential (VEP) findings were obtained by both pattern reversal (Figure 1A and B) and flash stimuli. Pattern reversal VEP was not recordable in right eye with reduced amplitudes in left eye at initial presentation (Figure 2). Flash VEP demonstrated similar findings. After 10 days of initial visit, Pattern reversal VEP (PRVEP) revealed delayed P100 latency in both the eyes (greater delay in the right eye). Flash VEP exhibited recordable and reproducible VEP with similar P100 latency delay in right (140 ms) as well as left eye (134 ms) (>30% deviation from normative lab values) (Figure 3). Interocular latency difference (right eye-left eye) was greater. Interocular amplitude ratio was also found to be reduced (left/right eye). Latency delay was more in right eye (latency delay is a common finding after the acute stage of ON has passed) while amplitudes were found to be more affected in left eye (which are lost in acute stages of ON).



**Figure 1: Patient with adult-onset bilateral optic neuritis during VEP recording procedure.**



**Figure 2: PRVEP record of the patient at the initial visit with no recordable VEP in right eye while recordable VEP with reduced amplitudes in the left (flash VEP record exhibited similar findings).**



**Figure 3: Flash VEP to goggle stimulation in the patient at tenth day after initial visit showing delayed P100 latencies in both the eyes (P100 in right eye: 140 ms and left eye: 134 ms)**

VEP records suggest the presence of bilateral optic neuritis with asymmetry, which is in concordance with his clinical presentation (diminished vision in right eye preceding the left) and also with the radiological findings.

Patient was started on IV (intravenous) methyl prednisolone 1 g for 3 days followed by oral prednisolone 1 mg/kg per day for 11 days and tapered in next one week as per ONTT (optic nerve treatment trial). Visual acuity of the patient improved dramatically to 6/9 in both the eyes on 5<sup>th</sup> day and after completing full course it improved to 6/6 in both eyes. Color vision improvement was partial. Presently, patient is on monthly follow-up with no deterioration of vision.

## DISCUSSION

Based on its clinical features, ON can be classified as atypical or typical. Careful ophthalmic, neurologic and systemic examinations are required to distinguish between typical and atypical ON to make a proper treatment plan. Typical and atypical presentations of optic neuritis have been distinguished on the basis of various features such as age, gender, progression of symptoms, unilaterality or bilaterality, degree of visual acuity at the time of presentation, normal or swollen optic disc, severity of the ocular findings, presence or absence of RAPD (relative afferent pupillary defect), visual field defects, visual recovery, history of MS (multiple sclerosis) or ON, presence or absence of family history and extent of the risk of MS. Our case of a young male presenting with bilateral sequential loss of vision is an atypical presentation with respect to bilaterality, gender, presence of swollen optic disc and with no associated risk of MS. Patients with atypical optic neuritis are reported to be at lower risk of developing MS and are suggested to be extensively evaluated for other causes of optic neuropathy. In our case no such associated infectious aetiologies were found. This form of presentation is extremely rare in adults. More commonly, bilateral optic neuritis is manifested in children with associated infectious or autoimmune aetiology. Atypical cases are thought to be having post-infectious, post-vaccination or other aetiologies. These associations are however, more common in children. Based on ophthalmologic findings and the location of involvement, ON can present with retro bulbar neuritis with normal optic disc appearance; papillitis with swollen disc; perineuritis involving optic nerve sheath while optic disc may or may not be swollen and neuroretinitis with optic disc oedema.<sup>2-4,6,7</sup> Our patient had optic disc oedema bilaterally. Electrophysiological reports by visual evoked potentials clearly indicated the features of ON at the acute stages of ON with absent waveforms and reduced amplitudes and the features of sequential involvement of the eyes at the subsequent visits with delayed latencies (a greater delay in the initially affected eye was evident). A delay in the P100 of the visual evoked response (VER) is the

electrophysiologic manifestation of slowed conduction in the optic nerve as a result of axonal demyelination.<sup>8</sup>

Hence, regarding differential diagnosis of optic neuritis, where clinical features and fundusoscopic examinations help differentiating the typical and atypical optic neuritis, MRIs and VEPs provide confirmation of diagnosis as structural and functional impairment of optic nerves. Our patient was started on intravenous prednisolone followed by oral which was according to the ONTT recommendations which suggests intravenous steroid therapy in bilateral involvement, unocular patients and in those desiring intervention.<sup>2,3,9,10</sup>

Visual recovery in atypical presentation is usually not spontaneous and deterioration of vision has been suggested after steroid discontinuation in these cases. Typical forms, however, are said to have spontaneous visual recoveries.<sup>2,5</sup>

Visual evoked potentials with P100 latency measures can be another objective means of visual outcome prediction.<sup>11</sup> Serial studies of VEP have shown that the shortening of VEP latency proceeds for up to 2 years and possibly for longer in younger patients. P100 latencies remain prolonged even after visual acuity improvements and hence also employed for finding the evidences of previous asymptomatic episodes of optic neuritis. As compared to patients with associated MS, it was found that these patients (without MS) might have less severe electrophysiological deficit initially but the latter demonstrate faster recovery.<sup>12</sup> This case with no features of associated MS or infectious aetiologies with bilateral sequential involvement of both the optic nerves was started on steroids and was found to demonstrate visual recovery. The follow-up of the patient for his ophthalmological and electrophysiological findings will be continued. Steroid responsiveness and visual functions thereafter will reveal his long-term visual outcome.

## CONCLUSION

Optic neuritis in typical or atypical form demands attentive and careful diagnostic approach as the condition can have perilous visual outcomes. A comprehensive diagnostic work-up including clinical features, ophthalmological findings, electrophysiological tests and imaging techniques (MRI) is valuable in identifying structural and functional involvement of the optic nerves and in revealing the form of presentation. Risk of development of serious neurological disease in typical form while associated infectious etiologies in atypical presentation necessitates appropriate diagnostic and prognostic perspectives.

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