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Case Report

From Darkness to Light: A Rare Case of Visual Recovery from Absent Light Perception to 20/20 in a Young Girl with Bilateral Optic Neuritis

Indranil Saha[1*] and Manu Taneja[2]

[1]Vitreo-Retina Sitapur Eye Hospital Sitapur, Uttar Pradesh, India

[2]Postgraduate Sitapur Eye Hospital Sitapur, Uttar Pradesh, India

[*]**Corresponding Author:** Indranil Saha, Consultant, Vitreo-Retina Sitapur Eye Hospital Sitapur, Uttar Pradesh, India.

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Abstract

Optic Neuritis is an ophthalmic emergency, characterized by sudden loss of vision, which needs immediate attention. Although steroids are the main stay of treatment, stringent diagnostic measures including an extensive serology workup and neuroimaging need to be done as to ascertain the etiology. This case report describes a young girl with bilateral severe optic neuritis, with denied perception of light in both eyes, and no systemic association. Immediate administration of methylprednisolone is the norm in such cases, and as per Optic Neuritis Treatment Trial, it is usually given for 3 days followed by oral prednisolone. In this patient, intravenous 1gram methylprednisolone was administered for 5 days, and then followed by oral prednisolone. The patient showed an excellent response with a complete recovery of vision to 20/20, which is an extremely rare phenomenon.

Keywords: Optic neuritis; Methyl prednisolone; Optic neuritis treatment trial.

Introduction

Optic neuritis (ON) is an acute inflammatory demyelinating disorder affecting the optic nerve. Usually, an event of isolated Optic neuritis has the following characteristics such as, being unilateral, more common in females and associated with painful visual loss without systemic or other neurological symptoms. The visual loss can range in severity from mild (20/20) to very severe (denied perception of light) [1,2]. The etiology for ON can be multiple, such as demyelinating disorders or infections, inflammation, genetic disorders, and toxic reasons. Idiopathic optic neuritis are those cases where the responsible etiology may not be known. When a patient presents with an initial episode of ON, patients should undergo further tests. For proper assessment of the patient, a battery of investigations needs to be done such as, routine blood work, contrast enhanced magnetic resonance imaging of the brain and orbit, cerebrospinal fluid tests, and visual evoked potentials. Optic neuritis is an

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inflammatory condition affecting the optic nerve causing severe loss of vision. Acute optic neuritis presents quite commonly, in clinical practice, requiring an algorithmic assessment to guide management and restore vision. The optic nerve is the most accessible part of the central nervous system, so optic neuritis also represents an important paradigm to help decipher mechanisms of damage and recovery in the central nervous system [3]. Pain on eye movement, which otherwise is an important sign in patients with optic neuritis can be absent in the 8% of patients whose inflammatory focus lies in the intracranial portion of the optic nerve and thus proximal to its mobile portion.

Exciting developments have occurred over the past decade in understanding of optic neuritis pathophysiology, and these developments have been translated into treatment trials. In its typical form, optic neuritis presents as an inflammatory demyelinating disorder of the optic nerve, which can be associated with multiple sclerosis. Atypical forms of optic neuritis can occur, either in association with other inflammatory disorders such as Neuromyelitis optica (Devic's disease) or in isolation.

Case report

24-year-old female presented to the emergency with loss of vision in both eyes for 10 days. Patient was apparently well till 2 months ago when the patient complained of frequent headaches which lasted for a few hours. There were no aggravating factors noticed. The patient had undergone a detailed systemic examination and a CT scan brain, but all tests were reported normal. 2 weeks prior to presenting to us the patient noticed blurring of vision in the right eye and 2 days later the patient felt the same in the left eye. There was rapid progressive diminution of vision in both eyes. On examination the patient denied perception of light in both the eyes. The intraocular pressures were normal in both eyes. Bilateral non reacting mid dilated pupils were noted. Fundus examination revealed severe bilateral disc edema with pallor. The systemic condition of the patient was evaluated to rule out any inflammatory or infectious etiology, all of which came normal the erythrocyte sedimentation rate (ESR) was mildly raised to 40. Contrast enhanced MRI of brain and orbit revealed bilateral optic nerve enhancement. Rest of the examination was unremarkable. The imaging included a screening of the spine, which was within normal limits. The patient was started on intravenous 1gram methylprednisolone once daily for 5 days following which the patient was administered 1mg/kilogram body weight oral prednisolone for 10 days and then gradually tapered. By the 3rd day, the patient's vision started improving, and the pupils regained normal reactions. The disc edema started reducing. By the 14th day of treatment, the patient was having 20/20 as best corrected visual acuity in both eyes. The fundus was unremarkable with normal looking optic nerves. Although mild pallor was noted in the left optic nerve (Figure 1). Her vision and clinical condition remained stable for 6 months. A repeat contrast enhanced MRI brain and orbit was done at 6 months, which was unremarkable.

Discussion

Optic neuritis is an inflammatory disease of the optic nerve that typically affects young adults. Its second only to glaucoma as the most common acquired optic nerve disorder in people younger than the age of

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50 years. Patients with optic neuritis complain an ocular discomfort associated with eye movements, which is believed to occur due to the close proximity of the optic nerve sheath to the superior and inferior recti sheath [4]. However, the ONTT trial and certain studies from Africa documented ocular pain/discomfort in only 7.8% patients. This patient complained of a persisting headache which had been aggravating prior to vision loss.

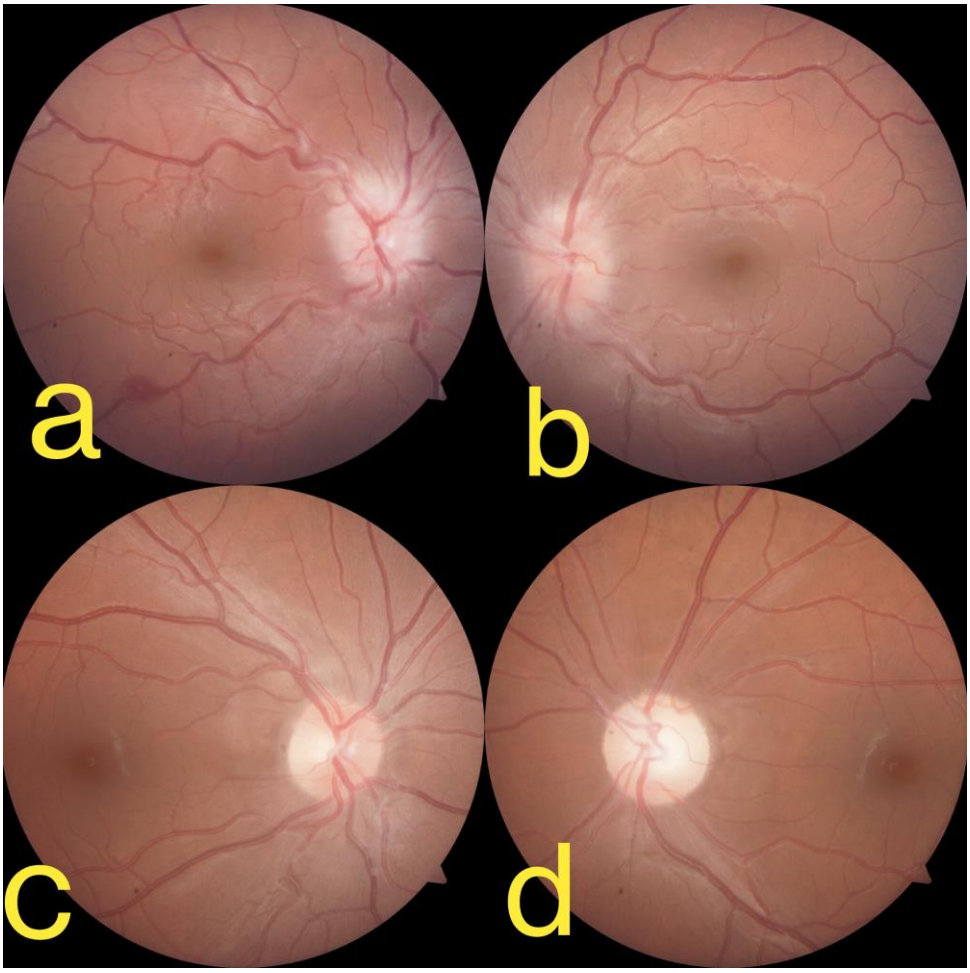

Figure 1: (a) and (b) Fundus images of right and left eyes at presentation. Both the optic nerves appear grossly edematous, (c) and (d) Fundus images showing resolution of disc edema in both the optic nerves. Pallor can be noted bilaterally. Significant pallor noted in the Left Optic nerve.

Simultaneous bilateral optic neuritis like in our patient is a very rare phenomenon and has been reported in only 0.4% cases [5]. Immediate high dose corticosteroids have been well established in the treatment of acute optic neuritis. Mahashweta Dutt et. al [6] advocated that early initiation of high dose corticosteroids has favorable outcomes in cases of optic neuritis and also preserved the retinal ganglion cells in relapsing/ remitting cases of optic neuritis. The Optic Neuritis Treatment Trial (ONTT) outlined the treatment protocol in cases of acute optic neuritis as intravenous 1gram methylprednisolone given once daily for three days followed by oral prednisolone (1mg/kg/day) for 11 days. In our patient, due to the severe inflammation affecting both the nerves and complete visual loss, 1 gram methylprednisolone was given once daily

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for 5 days (more than the recommended duration), followed by 10 days of oral 1mg/kg body weight oral prednisolone which was gradually tapered. The patient showed remarkable progress and within 2 weeks of treatment, having improved from no perception of light to 20/20 vision in both eyes. Hence, it postulates an extended regime of 5 days, once daily, 1gm intravenous Methylprednisolone compared to the 3-day regimen as per the ONTT trial, in severe cases of Optic Neuritis.

In the ONTT, the adverse effects of treatment reported insomnia, mood changes, gastritis, facial flushing and weight gain. These were attributed to low levels of Ascorbic acid [7]. This patient was prophylactically put on oral 500mg Ascorbic acid once daily for a month and didn't report any of the mentioned adverse effects. Optic neuropathies can also occur due to certain bacterial (Toxoplasmosis, Tuberculosis, Syphilis, Lyme's disease) or viral infections (Hepatitis A virus, Herpes, or enteroviruses). In these cases, too, there is a progressive visual loss and severe optic disc edema [8]. The patient went through extensive serology and was found negative for any infections. The patient in this report, presented with bilateral no perception of light with severe disc edema affecting both optic nerves. Neuroimaging was positive for only bilateral optic nerve enhancement. The patient was treated with intravenous 1gram Methylprednisolone for 5days followed by 1mg/kg body weight oral prednisolone for isolated bilateral optic neuritis. The vision recovered to bilateral 20/20, such recovery is extremely novel with not much mention in literature to the best of our knowledge.

Consent

Written informed consent was obtained from the patient to publish this case

Conflicts of interest

Nil

Financial disclosure

Nil

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