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Post-Varicella Retrobulbar Optic Neuritis with Encephalitis in Immunocompetent Child: A Case Report

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Abstract

Optic neuritis as a complication of varicella infection is rarely encountered. We hereby report a case of a 3-year-old immunocompetent boy who presented with sudden bilateral vision loss with recurrent generalised seizures and myoclonus four weeks after eruption of lesions of chickenpox. Ophthalmologic examination revealed only light perception in both the eyes with normal fundus examination, but prolonged latency in both eyes on visual-evoked potential (VEP), suggestive of bilateral retrobulbar optic neuritis. Preceding clinical history of chickenpox, findings of VEP and concomitant neurological involvement supported the diagnosis of post-varicella retrobulbar optic neuritis. After receiving a short course of intravenous methylprednisolone followed by tapering dose of oral steroid along with anti-epileptics, the visual acuity normalised bilaterally six months later.

Keywords

Acyclovir, Chickenpox, Encephalitis, Optic Neuritis

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1. Introduction

Retrobulbar optic neuritis, being a rare complication of varicella infection in an immunocompetent child, is reported here along with comparison to already published cases of its kind.

2. Case History

A 3 year old boy, presented to our institute with the history of one episode of generalized tonic clonic convulsion 9 days back, managed outside with antiepileptic drugs. On the next day, his parents noticed that the boy was neither able to perceive her mother nor the objects. He used to reach toys in wrong direction when presented in front of him. He also was not able to locate the source of light in the room. His plain CT head done before coming to us did not reveal any abnormality. Four weeks back, he had history of chickenpox with typical vesicular rashes on trunk, back and limbs along with fever, which recovered after being treated with oral acyclovir for 7 days. Developmental history did not reveal any abnormality except that he was not vaccinated for varicella. On detailed ophthalmologic evaluation, he was not able to perceive hand movements, though he could perceive bright light; pupils were normal in size, very sluggishly reacting to light, without relative afferent papillary defect, with normal fundoscopic examination. The rest of neurologic examination was normal.

Routine blood chemistry was within normal limits except for raised serum lactate level of 37.2 mg/dL (normal: 4.5-20mg/dL). Serology for varicella zoster virus was negative, along with negative results for HIV, Toxoplasmaand cytomegalovirus infections.

VEP revealed latency of 121.88 ms in right eye and 110 ms

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in left eye with amplitude of 11.74 and 10.32 microvolt in right and left eyes respectively; suggestive of bilateral retrobulbar neuritis (interpretation based on age-matched normative data of our laboratory).

His MRI brain with contrast including orbital cuts revealed high signal intensity in bilateral parieto-occipital and temporal cortex in T2-weighted images without diffusion restriction. Both optic nerves revealed non-specific changes in signal intensity, with interruption in CSF sleeve around left optic nerve. However, no contrast enhancement was seen in optic nerves. His cerebrospinal fluid (CSF) examination revealed normal values of sugar(55 mg/dL), protein(40.70 mg/dL), cell count (4/cmm, 100% lymphocytes), lactate, adenosine deaminase; with negative gram stain, ziehl-neelsen stain, fungal smear, cryptococcal antigen, routine culture, bacterial antigen panel for meningitis, herpes simplex virus 1 & 2 real time NASBA. CSF varicella IgM was negative; however, IgG was positive, corroborating well with history of chickenpox 4 weeks back. Sleep electroencephalography (EEG) (triclofos-induced) was normal.

During hospital stay, he suddenly started having spontaneous as well as action generalised myoclonic jerks.As he was already being managed with sodium valproate, phenytoin and clobazam; their doses were adjusted accordingly. EEG repeated on the next day of development of myoclonic jerks did not reveal cortical correlate of myoclonus. His lumbar puncture was also repeated to look for progression of underlying disease, which showed normal CSF composition [protein (36 mg/dL), sugar (73 mg/dL), cells (2/cmm), chloride, lactate dehydrogenase and lactate]. In view of raised serum lactate with no clues in CSF or EEG and non-specific features in MRI brain, mitochondrial genetic profile was also sent to rule out progressive myoclonic epilepsy with underlying mitochondrial etiology as a reason for continuous myoclonic jerks in this child, which was negative. Finally myoclonus was presumed to be of post-viral etiology after ruling out other causes.

He was given injectable methylprednisolone according to his weight for 5 days and discharged on oral steroid with tapering dosage as well as on sodium valproate, clobazam and phenytoin sodium as myoclonic jerks were not being controlled with monotherapy.

Bilateral vision loss was not improved much during hospital course, but he was closely followed on outdoor basis. To our surprise, his vision gradually improved over 6 months and became absolutely normal in both the eyes and myoclonus also recovered completely. This patient has been in our follow up for one year since discharge and neither recurrence of optic neuritis nor any other neurological disorder has been found till now. Follow-up MRI brain 6 months after discharge was normal.

3. Discussion

Varicella is a communicable disease caused by *Varicella zoster virus*. The diagnosis is purely clinical by the occurrence of typical skin eruptions. Common complications of varicella include secondary bacterial infection, varicella pneumonia, and neurologic manifestations. Apart from post-varicella encephalitis, other less common neurological complications include cerebellar ataxia, Guillain-Barre syndrome, transverse myelitis and facial nerve palsy.

Optic neuritis as a complication of varicella infection is rare especially in an immunocompetent child, and may occur with other neurological complications. Ocular manifestations may manifest during the infection course or after varicella infection. The pathogenesis behind central nervous system (CNS) involvement is presumed to be direct viral invasion, immune-mediated, or both [1,2]. Molecular mimicry between virus antigens and neural proteins in genetically selected individuals may play a role in immune-mediated mechanism [2]. Both the delayed onset of optic neuritis post-varicella and complete recovery of visual function as in our case suggests a reversible process substantiating an immunologic mechanism. Moreover, CSF IgG positivity for varicella virus also supported the diagnosis. Etiological link between the varicella virus and optic neuritis can be established indirectly by the history of chickenpox and the exclusion of other common etiologies, because histopathological diagnosis of direct specimen from optic nerve is not possible. However, there is still a possibility of direct invasion by the virus into the optic nerve in case of acute varicella infection; hence symptoms can worsen with steroids.

Till date, only 12 cases of post-varicella optic neuritis have been reported as per our search,including the one who initially manifested as having retinitis [3](Table-1). In contrast to the patients with optic neuritis and resultant functional recovery, children with chickenpox-associated neuroretinitis showed a poor outcome [4-6]. The youngest patient of post-varicella optic neuritis was 3 year old, and the shortest and the longest intervals between onset of chickenpox and that of optic neuritis were 2 and 38 days respectively as per the available data [6,9]. Recurrence of optic neuritis in the case of post-varicella has not been reported yet. Definite period for neurological follow-up after discharge has not been defined; however, we recommend following the patient at least 6 months after the visual recovery to detect any recurrence of optic neuritis or other post-varicella neurological complications.

The use of steroids in the treatment of optic neuritis is still debatableas usually the recovery is rapid and spontaneous [2].

No randomized controlled studies are available as the entity is rare. However, steroids can quicken the recovery if used early in absence of the contraindications [1, 2]. As encephalitis is presumed to be due to an auto-immune damage of the white matter, the use of corticosteroids with antiviral drugs is justifiable if optic neuritis co-occurs with encephalitis, as in our case, who received steroids and had a good functional outcome [8].

4. Differential Diagnosis

As compared to the optic neuritis in adult patients, pediatric population has different characteristics namely bilateral involvement, papillitis, headache, post infectious or post immunization etiology rather than a demyelinating one like multiple sclerosis (MS) or neuromyelitisoptica (NMO). However, if there is past history of neurologic symptoms, possibility of childhood MS should be considered in the differential diagnosis. Similarly, if it is associated with other neurologic features, it may suggest acute disseminated encephalomyelitis (ADEM) as a differential diagnosis.

Evaluation of such cases should include detailed history including any recent infection or vaccination, brain imaging to rule out demyelinating etiology, markers for vasculitis, work up for sarcoidosis, anti-NMO antibodies, as well as evaluation to rule out infections specifically involving optic nerves like tuberculosis, cryptococcus, lymes disease etc. Analysis of CSF can be of great help in solving the question of etiology for optic neuritis and in ruling out an infective etiology in case if steroids are being planned as a part of treatment.

Seizures in association with sudden bilateral vision loss in a child with the background of recent varicella infection makes parainfectious etiology more likely, although other secondary causes for seizures including toxic, metabolic, other infections as mentioned above should be ruled out.

Recommendations

Children with active or recent past infection of chickenpox, presenting with sudden vision impairment or any other ocular symptoms, irrespective of their immunologic status, should be urgently evaluated to rule out optic neuritis, retinitis, and their consequent complications such as retinal detachment and macular involvement. Early, immediate and vigorous management including acyclovir and steroids (when appropriate and in absence of contraindications) can potentially preserve residual visual function. Close long term follow-up of the patient for visual function recovery and development of any other delayed ophthalmic or neurological complications is indispensable for the complete patient management.

Table 1. Existing patient data of ocular complications of varicella infection.

Ref.	Authors	Publication year	Age (yr)	Gender	Interval in days between onset of chickenpox and ocular Symptoms	Primary Manifestation	Further Complications
	Chavernac	1908	11	Male	Gradual	Optic neuritis	Bilateral
14	Paton	1917	14	Male	9	Optic neuritis	--
	Ratner	1925	4	Male	NA	Optic neuritis	Bilateral
9	Hatch	1949	6	NA	14	Optic neuritis	--
4	Copenhaver	1966	3	NA	2	Optic neuritis	Bilateral with unilateral macular lesion
10	Walsh & Hoyt	1969	7	NA	14	Optic neuritis	Bilateral
11	Bruguier et al	1981	8	Female	NA	Optic neuritis	--
7	Selbst et al	1983	10	Female	38	Optic neuritis	Bilateral
13	Purvin et al	1988	14	Female	7	Optic neuritis	Bilateral
5	Capone & Meredith	1992	2	NA	3	Retinitis	--
12	Lee & Charles	2000	4	Male	42	Retinitis	Retinal detachment
6	McKinnon et al	2002	9	NA	NA	Retinitis	--
8	Roelandt et al	2005	3	Male	7	Optic neuritis	--
15	Stergiou et al	2007	6	Female	7	Optic neuritis	--
3	*Tappeiner et al*	2010	3	Male	3	Retinitis	Optic neuritis / papillitis
	Own case	2013	3	Male	28	Optic neuritis	Bilateral retrobulbar neuritis

(NA- Not available)

(Courtesy: Tappeiner C, Aebi C, Garweg JG. Retinitis and optic neuritis in a child with chickenpox. Pediatr Infect Dis. 2010; 29: 1150-1152 [5])

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