

A Young Boy with Painful Eye Swelling and Facial Rash

MD. SHAHRIAR MAHBUB,¹ ABDULLAH AL FARUQ,¹ MD. MAHMUDUR RAHMAN SIDDIQUI,¹ MOHAMMAD ASHIK IMRAN KHAN,² HAM NAZMUL AHASAN,³ MD. BILLAL ALAM,⁴ MD. TITU MIAH,⁵ RATAN DAS GUPTA⁵

Abstract

Oculomotor palsy is a recognized uncommon complication in herpes zoster ophthalmicus. A case of herpes zoster ophthalmicus with ophthalmoplegia is reported. A 15 year old boy presented with ptosis, chemosis, proptosis, ophthalmoplegia involving IVth and VIth cranial nerve following vesicular eruption on his right forehead. He was treated with oral acyclovir and oral steroids. He made a partial recovery with improvement of ptosis, proptosis but with residual VIth nerve palsy.

Keyword: Herpes zoster ophthalmicus, ocular herpes zoster, ophthalmoplegia, Bangladesh

Introduction

Herpes zoster is a common disease caused by varicella zoster virus (VZV). Cranial nerves are the second most common site of involvement in herpes zoster.¹ Herpes zoster ophthalmicus (HZO) occurs when the ophthalmic division of the trigeminal nerve is affected by reactivation of the virus. Ocular involvement is observed 10% of all herpes zoster cases and ophthalmoplegia is reported in 10-20% patients.² Multiple oculomotor cranial nerve palsy is an uncommon presentation in HZO. Although recovery is expected in most cases, a minority of patient may be left with permanent visual loss.

Case Report

A 15 year old previously healthy boy presented with two days history of painful blisters on his right forehead, fever and swelling of both eyes. The blisters started on the right side of the forehead without crossing the midline. Subsequently he developed fever, painful swelling of right eye first then left eye and double vision. Examination revealed vesicular eruption along the distribution of the ophthalmic division of the trigeminal nerve on the right side of the forehead, proptosis of the right eye, chemosis, ptosis, (Fig. 1) reduced visual acuity, ophthalmoplegia involving right IVth and VIth cranial nerve. Corneal sensation was also diminished but funduscopy showed a normal disc.

Based on these findings, a clinical diagnosis of cavernous sinus syndrome was considered. His blood count showed raised ESR with neutrophilic leukocytosis. A magnetic resonance imaging (MRI) of the brain did not show any significant changes associated with inflammation either in

the region of cavernous sinus or in the orbital apex. An MRV was also normal.

The presence of vesicular eruption associated with ocular and extraocular involvement and imaging findings led to the diagnosis of herpes zoster ophthalmicus. The patient was treated with oral acyclovir, topical antibiotics (ciprofloxacin) and topical steroids (0.1% dexamethasone). After 2 weeks of treatments cutaneous eruption, ptosis, proptosis and visual acuity improved but the right VIth nerve palsy persisted (Fig. 2 & 3). Fundoscopic follow-up examination was also normal. He was discharged with a tapering dose of prednisolone and advised for regular out-patient follow-up.



Fig.-1: The patient with ptosis, proptosis and chemosis in the right eye



Fig.-2: The patient with residual right VIth nerve palsy after 2 weeks of presentation

1. FCPS (Medicine) course student, Department of Medicine, Dhaka Medical College
2. Post-graduate trainee, Department of Medicine, Dhaka Medical College
3. Professor, Department of Medicine, Dhaka Medical College
4. Associate Professor, Department of Medicine, Dhaka Medical College
5. Assistant Professor, Department of Medicine, Dhaka Medical College

Correspondence: Dr. Md. Shahriar Mahbub, Department of Medicine, Dhaka Medical College, Dhaka, Bangladesh. E-mail: shahriar04@yahoo.com



Fig.-3 Healed vesicular eruption in the distribution of ophthalmic division of the trigeminal nerve

Discussion

Herpes zoster ophthalmicus typically involves the ophthalmic division of the trigeminal nerve. Ocular involvement is found in 10-20% of all herpes zoster infection.² The skin eruptions do not cross the midline. The trigeminal nerve divides into supraorbital, lacrimal and nasociliary branches. Eruptions in the tip, side or root of the nose indicates nasociliary nerve involvement and known as Hutchison's sign. It is an indicator of ocular involvement and is a strong predictor of sight-threatening disease.³

The ocular manifestation of HZO may involve eyelid, conjunctiva, cornea, sclera, uvea, retina, choroid and cranial nerves. The presentation includes blepharconjunctivitis, secondary bacterial infection with staphylococcus aureus, subconjunctival hemorrhage, episcleritis or scleritis, epithelial or interstitial keratitis, uveitis, chorioretinitis, acute retinal necrosis, optic neuritis and oculomotor palsies.⁴

Cranial nerve palsy is reported in 13% to 33% of cases of HZO. The most common cranial nerve affected is IIIrd nerve followed by VIth nerve and the IVth nerve.⁵ Combined cranial nerve palsy is also seen. Complete ophthalmoplegia is a very rare manifestation of HZO. Optic nerve involvement is even rarer and usually presents as orbital apex syndrome.⁶ The onset of ophthalmoplegia is usually 2 weeks after the skin eruptions but ranges from 2 days to 5 weeks and sometimes independent of other ocular manifestation.^{4,7} A minority of patients may only have the ocular features without developing any skin rash—an entity called zoster sine herpette.⁴

The VZV is responsible for the development of herpes zoster. The rash commonly occurs in the facial and mid-thoracic to

upper lumbar dermatomes. Cell-mediated immunity is responsible for keeping the virus in a suppressed state. Decline in cell-mediated immunity due to any reason leads to the reactivation of VZV. HZO occurs due to involvement of trigeminal ganglion and the VZV spreads along the ophthalmic and sometimes the maxillary division of the Vth nerve.

The pathophysiologic mechanism of ophthalmoplegia in HZO is unsettled. These include direct extension of the VZV from the Vth nerve to the IIIrd, IVth or VIth nerve either in the region of cavernous sinus or superior orbital fissure, inflammatory soft tissue edema causing cranial nerve palsy. One study suggested orbital inflammation produces high intraorbital pressure resulting in proptosis and ophthalmoplegia. Other possible explanations include secondary vasculitis, microinfarction, immune mediated demyelination, meningo-encephalitis.⁷⁻⁹

The clinical features and imaging findings of our patient are consistent with a diagnosis of HZO. Though HZO occurs more commonly in patients over 50 years it has been reported in younger patients as well.¹⁰ The typical vesicular eruption followed by ophthalmoplegia was likely to be complicated by secondary bacterial infection as evident by fever, raised inflammatory markers and neutrophilic leukocytosis. As the MRI did not show any involvement of the cavernous sinus or the superior orbital fissure, the ophthalmoplegia was likely caused by the mechanism previously explained.

Treatment of HZO consists of antiviral agents like acyclovir, valacyclovir or famciclovir. These drugs have been shown to be safe and effective in the treatment of active disease as well as preventing post-herpetic neuralgia. Ideally these should be started within 72 hours of onset of rash. Intravenous acyclovir is recommended for immunosuppressed patient, acute retinal necrosis or progressive outer retinal necrosis.^{3,11} Corticosteroids should be used in patients with moderate to severe pain, cranial nerve or central nervous system involvement.

The recovery from ophthalmoplegia may be delayed with a range between 2 months to 18 months. In a few patients, the condition may become permanent possibly due to ischemic vasculitis.¹² Our patient showed partial recovery with improvement of ptosis and proptosis but residual VIth nerve palsy.

Conclusion

Herpes zoster ophthalmicus associated with ophthalmoplegia is a rare presentation of VZV. The consulting physician needs to be aware of the complication of HZO and promptly start treatment without which permanent visual loss may ensue.

Conflict of Interest: None

References:

1. Im M, Kim BJ, Seo YJ, et al. Complete ophthalmoplegia after herpes zoster. *Clin Exp Dermatol* 2007; 32: 162–164
2. Liesegang TJ. Herpes zoster ophthalmicus. *Ophthalmology* 2008; 115: S3–S12
3. Sanjay S, Huang P, Lavanya R. Herpes zoster ophthalmicus. *Curr Treat Options Neurol* doi:10.1007/s11940-010-0098-1
4. Shaikh S, Ta CN. Evaluation and management of herpes zoster ophthalmicus. *Am Fam Physician* 2002; 66: 1723-30
5. Archambault P, Wise JS, Rosen J, et al. Herpes zoster ophthalmoplegia. Report of six cases. *J Clin Neuroophthalmol* 1988; 8: 185-193
6. Shirato S, Oshitari T, Hanawa K, et al. Magnetic Resonance Imaging in Case of Cortical Apex Syndrome Caused by Varicella Zoster Virus. *Open Ophthalmol J* 2008; 2: 109-111
7. Chang-Godinich A, Lee AG, Brazis PW, et al. Complete ophthalmoplegia after zoster ophthalmicus. *J Neuroophthalmol* 1997; 17: 262–5
8. Yildiz ÖK, Seđmen H, Bolayir E, et al. A Case of Herpes Zoster Ophthalmicus With Oculomotor Nerve Palsy. *J Neurol Sci Turk* 2009; 26: 500-504
9. Ying XH, Wagle AM, Tan L, et al. A rare case of herpes zoster ophthalmicus with complete ophthalmoplegia. *J Am Geriatr Soc.* 2008; 56: 2160-2
10. Sarabi K, Selim A, Khachemoune A. A young girl with blisters on her forehead. *J Fam Pract.* 2007; 56: 551-3
11. Vallejo-García JL, Vañó-Galván S, Rayward O, Moreno-Martin P. Painful eye with a facial rash. *Cleve Clin J Med* 2009; 76: 410-2
12. Dhingra S, Williams G, Pearson A. Severe, Permanent Orbital Disease in Herpes Zoster Ophthalmicus. *Orbit* 2008; 27: 325–327