

Case Report

Orbital apex syndrome and meningoencephalitis: a rare complication of herpes zoster

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Abstract: Orbital apex syndrome is a rare manifestation of Herpes Zoster (HZ). HZ Meningoencephalitis are also rare in immunocompetent persons. We report a rare case that was considered to be HZ meningoencephalitis with orbital apex syndrome in an immunocompetent female. The patient initially manifested with HZ skin lesions and lose of her left sight, diplopia, ptosis, followed by headache, neck pain, and fever, dizziness. Cerebrospinal fluid analysis showed elevation of lymphocytes and protein. A MRI abnormality was remarkable for the presence of a left cerebellum, occipital lobe as well as dura lesion. Head computed tomography and Magnetic resonance venography was normal. Corticosteroid therapy and antiviral therapy was effective to decrease the headache and skin pain. Symptoms were markedly improved after corticosteroid therapy. Three months later, we called the patient to follow up. Her meningoencephalitis symptom recovered. Her follow up brain MRI was normal. But left blindness and external ophthalmoplegia was persistent. This case suggested HZ could affect central nervous system and peripheral nervous system at the same time.

Keywords: Herpes zoster, meningoencephalitis, orbital apex syndrome

Introduction

Herpes zoster ophthalmicus (HZO) refers to involvement of the ophthalmic division of the trigeminal nerve from reactivation of latent varicella zoster virus (VZV) harbored in the trigeminal sensory ganglion. It is characterized by an acute dermatomal eruption that evolves through papular, vesiculobullous, pustular, and crusting stages over days to 3 weeks. The zoster rash is often accompanied by periocular pain and neurosensory disturbances in the first trigeminal division.

Ocular manifestations are observed in 20%-70% of patients with HZO, with involvement of every ocular structure. Postherpetic neuralgia is the most common neurologic sequela, but other neurologic complications, including cranial nerve palsies, stroke, myelitis, meningoencephalitis, and polyneuropathy, have also been reported [1, 2]. Ocular motor cranial nerve palsies are reported in 5%-31% of patients, but the occurrence of complete unilateral ophthalmoplegia, defined here as concurrent unilateral

impairment of ocular ductions in all directions, is rarer. Here we report a rare case of Herpes Zoster meningoencephalitis with orbital apex syndrome.

Case report

A 65-year-old woman presented with a history of localized zoster on the left forehead, and visited a local clinic treated as Herpes Zoster. Six days after the outbreak of the rash, she developed lose of her left sight, ptosis. 4 days later, she had headache, neck pain, and fever, dizziness, followed by confusion. Since brain MRI showed a brain and dura lesion, she was admitted to our hospital. She had no recent travel, animal exposure, corticosteroid intake, chemotherapy, or radiation therapy.

Physical examination showed a blood pressure of 125/72 mmHg, pulse rate of 84/min, respiratory rate of 20/min, temperature of 36.2°C, nuchal rigidity, and an erythematous bandlike vesiculopapular rash over the left periocular area and forehead.

HZ meningoencephalitis with OAZ



Figure 1. The patient showed severe ptosis and a partial limitation of ocular motility.

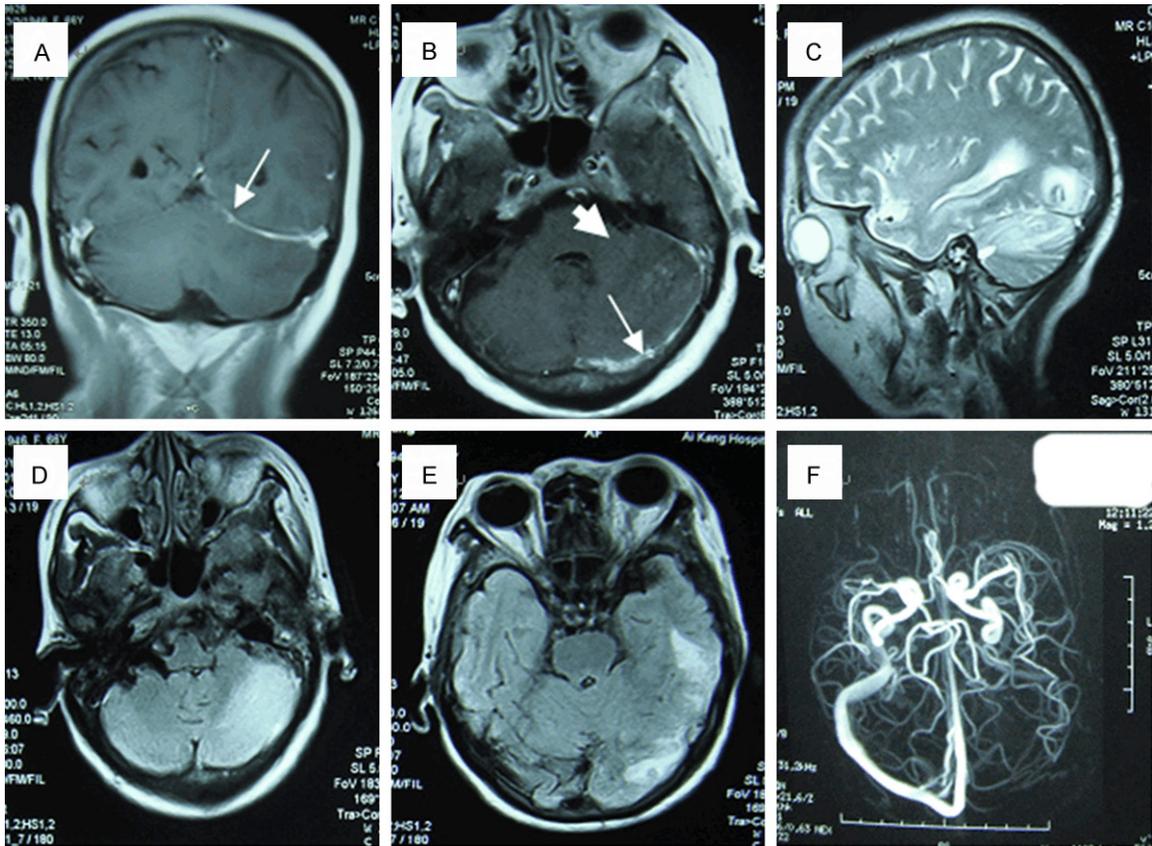


Figure 2. Cerebral MRI findings. A and B: Enhanced MRI showed cerebral dura mater (arrows) and left occipital lobe (arrow head) enhancement. C-E: Noncontrast MRI T2 weighted images showing Long T2 lesion in left occipital, temporal lobe and cerebellar hemisphere. F: MRV is normal.

Neurological examination revealed drowsiness, severe ptosis and a complete limitation of ocular motility in the left eye (**Figure 1**). A slit lamp biomicroscopic examination revealed corneal edema and signs of keratouveitis in the left eye and pigmentation on the lens capsule. Pupil sizes were 3 mm on the right and 3.5 mm on the left, and pupil reflexes disappeared on left eye. Sensations over the left half of the forehead and left cornea were impaired. Fundus examination revealed normal discs and macula in both eyes although soft exudates were present in the inferior retina of the left eye. Tendon

reflexes were normal, no pathological reflexes were noted. The finger-to-nose test was a little poorly performed on the left side. She had no ataxic gait. No abnormalities were seen in the autonomic nervous system. On examination, neck rigidity was present. But there was no Kernig's sign. A clinical diagnosis of herpes zoster meningoencephalitis complicated with orbital apex syndrome was made.

Results of the following were normal: serum electrolytes, renal and hepatic profiles, urinalysis, electrocardiography, chest roentgenography. Complete blood cell count was normal

HZ meningoencephalitis with OAZ

(3,570 cells/uL), which consisted of 27.24% lymphocytes, 68.34% neutrophils, 3.14% monocytes, and 1.14% eosinophils. Lumbar tap showed a yellow CSF. Cerebrospinal fluid analysis revealed 85 white blood cells per microliter with 86% lymphocytes. Cerebrospinal fluid glucose was 62 mg/dL (reference range, 45-60 mg/dL), which was 57% of the serum glucose of 108.18 mg/dL (normal, >50%), and protein was elevated at 114 mg/dL (reference range, 15-45 mg/dL). Results of CSF bacterial, mycobacterial, and fungal cultures were negative. Acquired immunodeficiency syndrome virus, Hepatitis B virus, Hepatitis C virus, syphilis tests were negative.

A magnetic resonance imaging (MRI) of the brain was remarkable for the presence of a left cerebellum, occipital lobe as well as dura lesion. Head computed tomography (CT) is normal. Magnetic resonance venography (MRV) is normal too (**Figure 2**).

Intravenous Ganciclovir (500 mg every 12 hours) and intravenous ceftriaxone (2 g daily) were initiated. She did not develop seizures or focal neurological deficits. But she still felt severe headache. Then intravenous dexamethasone (10 mg per day) was added and the headache dramatically decreased. Oral administration of prednisolone at a dosage of 35 mg for two weeks, 20 mg for two weeks, 10 mg for two weeks.

Three months later, we called the patient to follow up. Her meningoencephalitis symptom recovered. Her follow up brain MRI was normal. But left blindness and external ophthalmoplegia was persistent.

Discussion

HZ, commonly called shingles, is the result of reactivation of VZV or the human herpesvirus 3 infection. HZ exhibits no seasonal pattern, confirming that the disease results from the reactivation of latent virus rather than new exposure to VZV. It has been predicted that the incidence of HZ will increase in the coming decades, due to the increasing age of the population and also as a possible consequence of childhood varicella vaccination [3].

The common ocular manifestations of HZ include blepharo-conjunctivitis, keratitis, and uveitis. HZO refers to involvement of the oph-

thalmic division of the trigeminal nerve from reactivation of latent VZV harbored in the trigeminal sensory ganglion. The incidence and severity of HZ increase with advancing age, especially in those older than 60 years [4, 5]. Although there are several case reports of internal or external ophthalmoplegia (partial or total), optic neuropathy in HZO is rare, with orbital apex syndrome being reported in about 20 cases [6-9].

Herpes zoster meningoencephalitis is also a rare complication of varicella-zoster virus infection [10]. Historically, central nervous system (CNS) complications of VZV have been mostly recognized in immunocompromised patients, specifically in association with HIV infection and/or malignancy, or in those undergoing immunosuppression for organ transplantation. However, with the aid of PCR-based assays, such CNS complications are being increasingly recognized in immunocompetent individuals [11].

There is no report of HZ with both orbital apex syndrome and meningoencephalitis to our knowledge. The severity of presentation in our case is keratitis, anterior uveitis, and total ophthalmoplegia along with optic nerve involvement. Meanwhile, she showed cerebellum, orbital lobe and dura lesion in MRI. Most curiously, she is an immunocompetent person. We report a case of HZO with its varied ocular manifestations in a single patient, which suggested HZ could affect CNS and peripheral nervous system at the same time. Meanwhile, it seems the lesion of CNS is more reversible than of PNS.

The rate of extraocular muscle palsy in herpes zoster ophthalmicus was reported to be from 5% to 14% in a study investigating the general complications of the condition. However, a study that investigated the ocular motor functions reported that the rate is as high as 31% [12].

The pathogenesis of cranial nerve palsy in HZ can be explained as follows. First, the direct cytopathic effect of virus is on the surrounding neural tissue. Second, an allergic response to the virus can happen in the CNS. Third, occlusive vasculitis may be induced by the virus. Fourth, another latent neuropathic virus can be activated by the varicella/zoster virus [13].

The National Guideline Clearinghouse™ (NGC) is a database of evidence-based clinical practice guidelines and related documents. It recommends systemic antiviral therapy as first-line treatment for all immunocompetent patients with HZ who are at least 50 years of age, have moderate or severe pain and rash, and have nontruncal involvement. The use of a systemic steroid may be effective to prevent occlusive vasculitis [14].

Conclusions

Orbital apex syndrome is a rare ophthalmic complication of herpes zoster infection. Therefore, an evaluation of extraocular muscle and visual function should be performed during the examination of Herpes Zoster meningoencephalitis patients in order to screen for ophthalmoplegia. HZ could affect CNS and PNS at the same time.

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Disclosure of conflict of interest

None.

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