

A Case of Partial Horner's Syndrome caused by Herpes Zoster Ophthalmicus

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Horner's syndrome may rarely accompany herpes zoster ophthalmicus (HZO). A 78-year-old woman suffered from HZO accompanied ipsilateral ptosis and miosis. Before skin eruption, she was diagnosed as a paratrigeminal syndrome of Raeder. She was treated with intravenous acyclovir and prednisone for 7 days. Ptosis and miosis was not completely improved after 5 months of follow-up.

Key Words: Horner's syndrome, Herpes zoster ophthalmicus

INTRODUCTION

Varicella-zoster virus (VZV) is an exclusively human herpesvirus that causes chickenpox (varicella), becomes latent in cranial nerve and dorsal root ganglia, and frequently reactivates decades later to produce shingles (zoster) and postherpetic neuralgia. In immunocompetent elderly persons or immunocompromised patients, VZV may produce disease of the central nervous system (CNS)^{1,2}

Herpes zoster (HZ) can cause various ophthalmologic complications including ophthalmoplegia^{3,4} but rarely accompany Horner's syndrome.

CASE REPORT

A 78-year-old woman noted a left hemicranial headache for 10 days. The nature of headache was insidious onset and intermittent burning or

throbbing. One day before admission, she presented an acute onset of ptosis and conjunctival injection on left eye. Neurologic examination revealed a left ptosis and anisocoria (right 4 mm, left 2 mm), but direct and indirect light reflexes were intact (Fig.). Other neurologic examinations were unremarkable. She had hypertension for 25 years with regular medication. Other past, family, and social history were unremarkable.

Brain MRI showed multiple small and patchy high signal intensities in both cerebellum, basal ganglia, and cerebral white matters, suggesting



Figure. Left Horner's syndrome caused by herpes zoster ophthalmicus.

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small ischemia or infarctions. But MR angiogram was normal. Other various serum and urine tests were normal. And EKG, echocardiogram, carotid sonogram, transcranial doppler, and chest x-ray were unremarkable. But lumbar puncture and 4-vessel angiogram were not done due to patient refusal for test.

Before skin eruption, she was diagnosed as a paratrigeminal syndrome of Raeder. And starch iodine test on face was intact. But 3 days after admission, she showed painful grouped papulovesicles on left scalp, forehead, eyelid, and nose. She treated with acyclovir and prednisone for 7 days. After treatment, vesicles were improved slowly but partial Horner's syndrome was not improved for more than 5 months. Postherpetic neuralgia was absent.

DISCUSSION

Following primary infection, VZV becomes latent in cells of the dorsal root ganglia. Reactivation of endogenous latent virus produces HZ. The virus can reactivate after injury or trauma to the spine or nerve roots or in response to waning cell-mediated immunity to VZV caused by age or immunosuppression related to HIV infection, cancer, cytotoxic drugs, or systemic illness.⁵

The mechanism of reactivation of VZV that results in HZ is unknown. Presumably, the virus infects the dorsal root ganglia during chickenpox, where it remains latent until reactivated. Histopathologic examination of representative dorsal root ganglia during active HZ demonstrates hemorrhage, edema, and lymphocytic infiltration.⁶

While HZ most commonly affects the sensory nerves of the thoracic dermatomes, the cranial nerves are the second most commonly affected distribution. Acute complications of VZV infections are encephalitis, meningitis, contralateral hemiplegia, myelitis, Guillain-Barré syndrome, and facial paresis, etc., and delayed one is postherpetic neuralgia.⁷ Neurologic disease as a complication of HZ results from two mechanisms: direct invasion by unchecked replicating virus and delayed inflammatory response to the resulting antigenic load.⁸ The delay between primary infection and onset of hemiparesis is explained by the time taken for antibody-antigen complexes to

form and to induce thrombosis in the affected artery.⁹

In a community-based study, herpes zoster ophthalmicus (HZO) was diagnosed in 10% of all cases of HZ. However, these patients were significantly older with a greater male:female ratio than the rest of the population affected.¹⁰

The ocular complications of HZO, which usually occur during or after the cutaneous eruption, appear in approximately 50% of cases of HZO (range, 20~72%). Extraocular muscle involvement occurs much less frequently.³

The mechanism by which the ophthalmologic complications of HZ are not clear. Pathologic correlation has been limited because most patients with HZO do not die of the disease. But histopathologic studies of VZV infection suggest mechanisms of disease including features of viral replication, secondary inflammation, and vascular occlusion. The damage worked by zoster is caused by both chronic inflammation and vascular ischemia in response to direct viral invasion of a multitude of tissues. The cardinal pathologic features of reactivating HZ are inflammation and hemorrhagic necrosis of the ganglion and corresponding sensory nerve, often associated with neuritis, localized leptomeningitis, unilateral segmental myelitis, and degeneration of related motor and sensory roots.¹¹ In addition to traveling along the trigeminal sensory nerves, the VZV may travel via the affected sensory nerve roots to the brain stem or spinal cord to cause necrosis in the corresponding sensory nuclei. Virus probably spreads directly from the trigeminal ganglion to adjacent blood vessels and the CNS, thereby gaining access the arterial walls.¹² Therefore the ocular complications of HZO are related to direct viral invasion combined with host inflammatory, immune, and vascular reactions.^{13,14}

Horner's syndrome consists of unilateral enophthalmos, ptosis, miosis, and loss of sweating over the ipsilateral half of the face or forehead. It is caused by ipsilateral involvement of the sympathetic pathways in the carotid plexus, the cervical sympathetic chain, the upper thoracic cord, or the brain stem.¹⁵

The patient usually experiences a sharp, burning discomfort in a dermatomal distribution for 2 to 5 days before the onset of rash. A localized redness with red macules that become vesicles

develops in the same dermatomal pattern.¹⁶

Initially my patient did not present any skin lesion except partial Horner's syndrome. To be distinguished is a paratrigeminal syndrome of Raeder, which consists of pain like that of tic douloureux in the distribution of the ophthalmic and maxillary divisions of the fifth nerve, in association with ocular sympathetic paralysis (ptosis and miosis) but with preservation of sweating.⁷ My patient showed normal response to starch iodine test on face. But I did not try to pupil response test due to vesicles developed in the skin made diagnosis of HZO.

My patient presented a partial Horner's syndrome after HZO. It may be due to sympathetic pathway involvement as ischemia because the patient was old, had hypertension, acute onset of symptoms, and MRI revealed multiple lacunar infarction. Assal et al⁸ reported a case of internal ophthalmoplegia due to HZO, which occurred before cutaneous eruptions and any evidence of intraocular inflammation. Bak et al⁴ also reported a case of ophthalmoplegia caused by HZO. Internal ophthalmoplegia include a variety of conditions involving parasympathetic fibers to pupil's sphincter, from the brain stem to the iris. Lavin et al.¹⁹ suggests the mechanisms of ocular palsies of HZO includes direct neuropathogenic effect of the virus, demyelination process, compression due to generalized orbital inflammation by perineuritis and perivasculitis or vasculitis. The only published neuropathological report showed inflammation and profound demyelination of the ocular motor nerves within the cavernous sinus. But the ocular complications of HZO are also related to direct viral invasion combined with host inflammatory, immune, and vascular reactions as vasculitis.¹⁴

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