

# Extraocular Myositis Preceding Herpes Zoster Ophthalmicus

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## CASE PRESENTATION

### Initial Presentation and History

A 38-year-old woman presented to her primary care physician with a 1-day history of left-sided facial pain. The pain was believed to be acute left-sided maxillary sinusitis, and she was prescribed antibiotics. Four days later, the patient presented to the emergency department with left eye swelling, redness, double vision, and a dull achy pain on eye movement. She complained of blurred vision with increased tearing from the left eye associated with a generalized left-sided headache. She also reported a deep achy pain in the left eye that worsened with eye movements. Upon questioning, she denied any recent history of sick contacts or trauma to the face or eye. The patient's past medical history was unremarkable. She was not taking either prescribed or over-the-counter medications (other than the antibiotics prescribed 4 days earlier) and denied smoking, alcohol abuse, and recreational drug use. Her review of systems was remarkable for generalized malaise for 5 to 6 days, subjective fevers, and decreased appetite. No upper respiratory symptoms or nasal discharge was observed.

### Physical Examination

On examination, the patient appeared to be in moderate distress due to left eye pain and left-sided headache described as dull and constant. Her vital signs were within normal limits. Ear, nose, throat, and neck examination revealed no abnormality. The sinuses were nontender and no cervical lymphadenopathy was noted. Eye examination showed visual acuity of 20/20 in the right eye and 20/40 in the left eye. On slit lamp examination of the left eye, the conjunctiva was congested with minimal chemosis, and the anterior chamber was positive for 1 to 2 cells. Indirect funduscopic examination was normal. Left eye adduction was

limited by pain and diplopia. A tender, erythematous, vesicular, noncrusting eruption was noted on the peri-orbital skin and eyelids of the left eye and on the left side of the forehead, spreading down over the bridge of her nose. The rash had well-demarcated contours and did not cross the facial midline. On examination, the right eye was normal.

### Laboratory and Imaging Studies

Laboratory studies, including complete blood count and comprehensive metabolic panel, were ordered. The results of these studies were within normal limits, with the exception of an elevated erythrocyte sedimentation rate of 60 mm/h (normal, 0–20 mm/h). The patient was admitted to rule out preseptal orbital cellulitis.

Computed tomography scanning of the orbits showed edematous left extraocular muscles (**Figure 1** and **Figure 2**). The cavernous sinus did not demonstrate thrombosis, and the paranasal sinuses were normal. On hospital day 2, the vesicular elements spread over the nasal bridge reaching down to the tip of the nose. This clinical picture was consistent with herpes zoster ophthalmicus (HZO) complicated by extraocular myositis. Viral cultures of specimens obtained from the skin vesicles were subsequently found to be positive for varicella zoster virus (VZV).

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**Figure 1.** Coronal computed tomography image of the orbits demonstrating edematous left extraocular muscles (*thin arrows*) and a normal optic nerve (*thick arrow*).



**Figure 2.** Transverse computed tomography image showing inflamed extraocular muscle (*arrow*) and swollen eyelid (*arrowhead*).

### Treatment and Follow-up

Intravenous acyclovir and a low dose of oral prednisone were started as recommended by the ophthalmologist. Oral prednisone was started at 60 mg daily to reduce the severity of pain. Over the next few days, the patient's eye complaints improved dramatically, with marked reductions in pain and swelling. On the fifth day of hospitalization, the patient was discharged on valacyclovir 1000 mg orally twice daily for 10 days along with a 5-day tapering dose of methylprednisolone. At follow-up 2 weeks after discharge, the rash and ocular symptoms had resolved and the eye examination was normal.

### DISCUSSION

HZO is caused by VZV infection of the ophthalmic branch of the trigeminal nerve. The condition produces a vesicular rash confined to the dermatome supplied by the ophthalmic branch. VZV, a member of the Herpesviridae family, has double-stranded DNA and is characterized by an extremely short reproductive cycle (a few hours). After primary infection, VZV establishes latent infection in sensory nerve root ganglia.<sup>1</sup>

### Pathophysiology

The primary infection with VZV results in varicella (chickenpox), which is characterized by a generalized vesicular eruption. Viral latency is established during acute VZV infection. The virus takes up asymptomatic

permanent residence in the sensory dorsal root ganglia. Viral DNA can be detected by in situ hybridization in the dorsal root ganglia of adults many years after VZV infection.<sup>2</sup> Shingles develops when the VZV within the sensory ganglion is reactivated, which typically produces a vesicular rash within a well-defined dermatomal distribution. If the VZV residing in the trigeminal nerve becomes reactivated, the inflammatory reaction involves the eye, resulting in HZO. Of the 3 branches of the trigeminal nerve, the ophthalmic branch is affected twice as frequently as the other branches.<sup>3</sup> Exogenous exposure to the virus may occasionally be responsible for HZO.<sup>4</sup>

The exact mechanism by which the virus becomes reactivated is unknown. Virus reactivation causes severe ganglionitis and even hemorrhagic necrosis of the ganglion, which clinically manifests as dermatomal pain and hyperesthesia. Subsequently, the virus starts a 2- to 3-day course of anterograde axonal movement toward sensory nerve endings (cranial nerves III and IV). Skin eruptions appear when the virus reaches the skin after its slow migration via the nerve axons, which explains why pain and hyperesthesia precede the classical herpetic rash by several days.<sup>5</sup> The cell-mediated immunity specific for VZV acts to confine the spread of the virus within the ganglion during latent infection. This, in

turn, confines the replication of the virus within the cutaneous lesions.<sup>4</sup>

### Epidemiology

Approximately 90% of the population develop serologic markers of varicella infection by adolescence, with nearly 100% of the population having some evidence of the disease by age 60 years.<sup>1</sup> Typically, cell-mediated immunity to VZV in naturally immune individuals is boosted by reexposure to the virus from infected children. As more children are vaccinated against chickenpox, it is possible that adult immunity against VZV is reduced.<sup>4</sup> The incidence of herpes zoster in the general population has been reported to be as high as 3.2 per 1000 person-years. These numbers increase significantly with age, reaching 10.9 per 1000 person-years in populations over age 80 years.<sup>6</sup> Herpes zoster most often involves the thorax, neck, face, and lumbosacral area. HZO represents between 10% and 25% of all cases of herpes zoster, and there is risk of loss of vision in the affected eye in approximately half of HZO cases.

### Manifestations

In the prodromal stage of HZO, the patient may complain of fever, flu-like symptoms, and malaise lasting for 5 to 7 days before developing a rash. Pain and hyperesthesia localized to a defined dermatome are part of the prodrome. In up to 60% of patients, hyperesthesia can be severe and debilitating.<sup>3,7</sup> Within a few days, the skin overlying the dermatome shows signs of active involvement with the development of an erythematous maculopapular rash progressing to vesicles and pustules and finally, crusting and scarring. The rash typically evolves over a period of 4 to 14 days and does not cross the facial midline. The virus can be cultured from the lesions during the vesicular phase, making the patient infectious to individuals not previously exposed to the virus.<sup>1,4</sup> Spread of the rash to the tip of the nose signifies ocular involvement (known as Hutchinson's rule) and should warrant an ophthalmologic consultation.<sup>8</sup>

### Complications

Several complications, some of which can have a devastating effect on vision, are associated with HZO. They commonly include keratoconjunctivitis, scleritis, secondary glaucoma, optic neuritis, uveitis, chronic postherpetic neuralgia, secondary staphylococcal infection, and ophthalmoplegia caused by cranial nerve III, IV, or VI palsies; other rare but more serious complications include acute retinal necrosis and granulomatous angiitis of the central nervous system.<sup>8,9</sup> There are only

a few cases of extraocular myositis presenting as ophthalmoplegia reported in the literature.<sup>9–11</sup>

### Treatment

Treatment of HZO consists of administering antiviral drugs such as acyclovir (800 mg orally 5 times daily), valacyclovir (1000 mg orally 3 times daily), or famciclovir (500–750 mg orally 3 times daily) for 7 days; in severe cases, oral steroids may be added.<sup>12–15</sup> Initial treatment with intravenous acyclovir is recommended for several days before starting oral forms of antiviral drugs.<sup>8,12</sup> Treatment of HZO reduces the incidence and severity of most common ocular complications, such as dendritic keratitis, stromal keratitis, and uveitis.<sup>6,16</sup> Therapy is most effective if started within 72 hours of disease onset, but unfortunately this will not reliably prevent postherpetic neuralgia.<sup>17</sup> In patients over age 60 years, a 3-week course of acyclovir and high-dose oral corticosteroids is recommended. This combination may speed healing and improve quality of life and is superior to either drug alone.<sup>18</sup> Some data show that valacyclovir provides more prompt relief of herpes zoster-associated pain than acyclovir in patients older than 50 years.<sup>19</sup>

### CONCLUSION

HZO can present as unilateral myositis of the extraocular muscles with signs and symptoms of ophthalmoplegia, which are preceded by eruption of the classical skin rash. We recommend considering HZO in the differential diagnosis of patients presenting with unilateral ophthalmoplegia in association with ipsilateral facial pain.

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