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Varicella-Zoster Virus

Author: Wayne E Anderson, DO, Assistant Professor of Internal Medicine/Neurology, Western University of Health Sciences; Assistant Professor of Family Medicine, Touro University College of Osteopathic Medicine; Consulting Staff in Pain Management, Department of Neurology, California Pacific Medical Center; Consulting Staff in Neurology, Department of Neurology, California Pacific Medical Center
Contributor Information and Disclosures

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Introduction

Background

Varicella-zoster virus (VZV) is the cause of chickenpox and herpes zoster (also called shingles). Chickenpox follows initial exposure to the virus and is typically a relatively mild, self-limited childhood illness with a characteristic exanthem.

Approximately 1 per 4000 children develops VZV encephalitis, an acute neurologic disorder with potentially severe complications. In addition, immunocompromised children (eg, those receiving chemotherapy for leukemia or those with advanced HIV infection) can develop disseminated VZV infection, a potentially fatal complication.

After primary infection, VZV remains dormant in sensory nerve roots for life. Upon reactivation, the virus migrates down the sensory nerve to the skin, causing the characteristic painful dermatomal rash. After resolution, many individuals continue to experience pain in the distribution of the rash (postherpetic neuralgia). In addition, reactivation of VZV infection can cause a spectrum of atypical presentations, ranging from self-limited radicular pain without rash to spinal cord disease with weakness.

Pathophysiology

The host immunologic mechanisms suppress replication of the virus. Reactivation can occur if host immune mechanisms are compromised. This may be caused by medications, illness, malnutrition, or by the natural decline in immune function with aging. Upon reactivation, the virus migrates along sensory nerves and produces sensory loss, pain, and other neurologic complications. If motor nerve roots are also involved, weakness can develop in addition to sensory changes. Leptomeningeal involvement is rare but may develop when the ophthalmic branch of the trigeminal nerve is involved.

Frequency

United States

The rate of occurrence is about 5 persons per 1000 population. Immunosuppression increases this risk. The risk of postherpetic neuralgia increases with age. Approximately 50% of patients older than 60 years may have temporary or prolonged pain syndrome.

The frequency of VZV infection may decrease as the immunized children become adults.

International

VZV infection occurs with the same frequency in the United States and internationally.

Mortality/Morbidity

- Severe pain and insomnia are most bothersome to patients. About 95% of patients with zoster experience severe pain during the illness.
- Other presentations of zoster, including ocular (keratitis) and spinal cord (myelitis) presentations, may result in additional morbidity.
- Bacterial superinfection (impetiginization) of vesicular skin lesions can occur.

Race

The vesicular eruption of VZV infection may be more difficult to diagnose in patients with darker skin.

Sex

VZV infection occurs with equal frequency in males and females.

Age

- After primary infection, zoster can occur at any age. However, the risk of zoster increases with age.
- The risk of postherpetic neuralgia also increases with advancing age.

Clinical

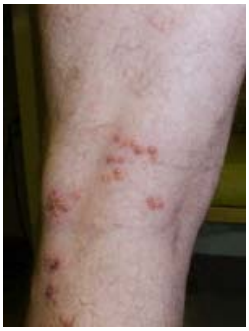
History

- Pain and paresthesia are typically the first symptoms. Until the characteristic vesicular rash erupts, diagnosis may be difficult.
- A prodromal period during which symptoms may vary is common. Pain occurs in 41% of patients, itching in 27%, and paresthesias in 12%.
- During the acute illness, 90% of patients experience pain, 20% describe helplessness and depression, and 12% experience flulike symptoms.

Physical

- Herpes zoster (shingles)
  - The most common presentation is the shingles vesicular rash, which most commonly affects a thoracic dermatome.
  - After a prodromal illness of pain and paresthesias, erythematous macules and papules develop

and progress to vesicles within 24 hours. The vesicles eventually crust and resolve.



Typical zoster in the vicinity of right popliteal fossa in a vertebral nerve L4 distribution.

- Pain and sensory loss are the usual symptoms, but motor weakness also occurs and is frequently missed on examination. Motor weakness results when the viral activity extends beyond the sensory root to involve the motor root. Cases of actual monoplegia due to varicella-zoster virus (VZV) brachial plexus neuritis have been reported.

- Zoster multiplex
  - Shingles may appear in multiple dermatomes, both contiguous and noncontiguous, on either side of the body.
  - They are more common in individuals who are immunocompromised.
  - Terminology depends on the number of involved dermatomes and on whether the condition is unilateral or bilateral. For example, zoster duplex unilateralis refers to the involvement of 2 unilateral dermatomes. Cases of zoster simultaneously occurring in 7 noncontiguous dermatomes have been reported.

- Zoster sine herpete
  - VZV infection may reactivate without causing cutaneous vesicles. These patients have severe dermatomal pain, possible motor weakness and possible hypesthesia, but no visible rash or vesicles.
  - Studies show that VZV infection may present as acute peripheral facial palsy in 8-25% of patients who have no cutaneous vesicles. This is more common in immunosuppressed patients who use acyclovir (or other agents) as zoster prophylaxis.[1]

- Myelitis
  - VZV infection may also cause central nervous system deficits.
  - Although deficits are more common in immunocompromised individuals, such presentations do occur in the general population.
  - In one report, the condition began as a typical shingles rash, but spinal cord involvement became apparent 3 weeks after the onset of the initial rash.
  - The manifestations are usually bilateral. The physical findings may progress.
  - The underlying pathology typically progresses for 3 or more weeks. Progression for 6 months in immunocompromised individuals has been reported.
  - With intravenous acyclovir treatment, most cases fully resolve. Recurrence is rare but has been reported.
  - Zoster encephalitis is also rare but is reported in otherwise healthy individuals. Due to the effectiveness of 2-dose vaccinations, fewer cases of VZV encephalitis occur,[2] yet most cases in vaccinated individuals are due to wild type from the vaccine strain.[3]

- Ramsay-Hunt syndrome
  - This syndrome occurs when the geniculate ganglion is involved.
  - The clinical presentation includes a peripheral facial palsy, pain in the ear and face, and vesicles in the external ear canal.
  - Additional auditory and vestibular symptoms may be present. The vesicles are not present in all cases.

- Keratitis (herpes ophthalmicus)
  - This is caused by reactivation of VZV infection in the ophthalmic division of the trigeminal nerve.

The presentation may include conjunctivitis or corneal ulcers. Complications include blindness.

- The vesicles do not have to be present.
- Rarely, in cases of herpes ophthalmicus, the virus migrates along the intracranial branches of the trigeminal nerve, causing thrombotic cerebrovasculopathy with severe headache and hemiplegia.

Causes

Immunosuppression increases the risk of both typical shingles and atypical presentations, such as myelitis, encephalitis, disseminated disease, and visceral involvement.

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