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The Clinical Picture

Odynophagia, peripheral facial nerve paralysis, mucocutaneous lesions

FIGURE 1. Vesicular eruption on the left concha and external auditory meatus.

FIGURE 2. Multiple vesicles and pustules on the left side of the tongue and soft palate.

A 54-YEAR-OLD WOMAN presented with a 7-day history of odynophagia, pharyngeal swelling, and painful skin lesions on her left ear. She had been on antiretroviral therapy for human immunodeficiency virus infection but had not been fully compliant with the treatment.

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On examination, she had painful erythematous vesicles and pustules on the left auricle and in the external auditory canal (**FIGURE 1**), as well as small vesicles and circumscribed erosions on the left anterior two-thirds of her tongue (**FIGURE 2**) and left palate. Facial sensory function was normal; however, she had lagophthalmos, a flattened nasolabial fold, ptosis of the oral commissure, and a loss of the forehead wrinkles on the left side of her face—all signs of peripheral facial nerve paralysis.

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Q: Which is the most likely diagnosis?

- ☐ Ramsay Hunt syndrome
- ☐ Herpes simplex
- ☐ Contact dermatitis
- ☐ Malignant external otitis
- ☐ Erysipelas

A: This patient had Ramsay Hunt syndrome, also known as herpes zoster oticus. It is a rare complication of herpes zoster in which the reactivation of latent varicella-zoster virus infection in the geniculate ganglion causes the triad of ipsilateral facial paralysis, ear pain, and vesicles in the auditory canal and auricle. Taste perception, hearing (eg, tinnitus, hyperacusis), and lacrimation can be affected.[1]

Ramsay Hunt syndrome is generally considered a polycranial neuropathy of cranial nerves VII (facial) and VIII (acoustic). In some cases other cranial neuropathies may be present and may involve cranial nerves V (trigeminal), IX (glossopharyngeal), and X (vagus). Vestibular disturbances such as vertigo are also often reported. It is more severe in patients with immune deficiency. Because the classic symptoms are not always present at the onset, the syndrome can be misdiagnosed.

DIAGNOSIS

Once the vesicular rash caused by herpes zoster has appeared, the diagnosis is usually readily apparent. The other main disease to consider in the differential diagnosis is herpes simplex. Herpes zoster infection is characterized by a painful sensory prodrome, dermatomal distribution, and lack of a history of a similar rash. However, if the patient has had a similar vesicular rash in the same location, then recurrent zosteriform herpes simplex should be considered. A noninfectious cause to consider is contact dermatitis. However, contact dermatitis usually produces intense itch rather than pain.

If the clinical presentation is uncertain, then viral culture, direct immunofluorescence testing, and a polymerase chain reaction assay is indicated to confirm the diagnosis. Polymerase chain reaction testing is the most sensitive test.[3]

TREATMENT

The rapid start of antiviral therapy is particularly critical in immunocompromised patients,[4] even if the vesicles have been present for 72 hours. Immunocompromised patients with Ramsay Hunt syndrome and other forms of complicated herpes zoster infection should be hospitalized for intravenous acyclovir therapy.

Corticosteroids and oral acyclovir (10 mg/kg three times daily for 7 days) are commonly used in Ramsay Hunt syndrome. In a recent review,[5] combination therapy with a corticosteroid and intravenous acyclovir did not show a benefit over corticosteroids alone in promoting resolution of facial neuropathy after 6 months.[5] However, randomized clinical trials are needed to evaluate both therapies.

Although antiviral therapy reduces pain associated with acute neuritis, pain syndromes associated with herpes zoster can still be severe. Nonsteroidal anti-inflammatory drugs and acetaminophen are useful for mild pain, either alone or in combination with a weak opioid analgesic (eg, tramadol, codeine). For moderate to severe pain that disturbs sleep, a stronger opioid analgesic (eg, oxycodone, morphine) may be necessary.[6]

Vestibular suppressants may be helpful if vestibular symptoms are severe. Temporary relief of otalgia may be achieved by applying a local anesthetic to the trigger point, if in the external auditory canal. Carbamazepine may be helpful, especially in cases of idiopathic geniculate neuralgia.[7]

OTHER CONSIDERATIONS

Once drug therapy is started, the patient should be seen at 2 weeks, 6 weeks, and 3 months to monitor the evolution of nerve paralysis.[8]

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