Case of Disseminated Varicella Zoster in Patient with AIDS

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Background
Varicella zoster virus (VZV) infection has two principle manifestations: primary disease, causing “chickenpox” and reactivation of latent disease, known as herpes zoster (HZ) or “shingles”. The most common manifestation of HZ is a dermalatal rash with acute neuritis; however, disease can be complicated by ophthalmic, otic and in about 3% of cases, neurologic involvement. Complicated disease is more common in immunocompromised patients. Here, we discuss a case of disseminated varicella zoster principally manifesting as Ramsay Hunt Syndrome (RHS) in a patient with Acquired Immunodeficiency Syndrome (AIDS).

Case Report

Initial Presentation
EM is a 57 year old man with AIDS chronically non-adherent to antiretroviral therapy who presented with two months of headache and subjective fever that was followed by right sided otalgia and facial drop two days prior to admission. On further questioning, he noted onset of a diffuse rash several weeks prior that was improving.

Evaluation
Exam: Edematous right pinna with purulent drainage in the conchal bowl and upper and lower right facial palsy. Diffuse erythematous follicular based papules, some with scarring and crusting over the entire body surface.

MRI: abnormal enhancement of geniculate ganglion and right facial nerve;

Diagnosis: His cranial nerve findings and geniculate ganglion enhancement were consistent with RHS, and diffuse rash suggested disseminated zoster. Flow cytometry was performed on CSF, which was normal, excluding CNS lymphoma as a cause for his symptoms, abnormal imaging and CSF pleocytosis.

In addition to identification of RHS, EM had evidence of otitis externa, likely a bacterial superinfection of initial rash.

Hospital Course
EM was initiated on IV antibiotics for his otitis externa and treated initially with IV acyclovir and corticosteroids for RHS. Steroids were discontinued after several days due to challenging glycemic control. He was discharged to home on culture directed antibiotics for otitis externa and valacyclovir to complete three week course for disseminated zoster.

Discussion
Ramsay Hunt Syndrome: The Basics
- Epidemiology: In one large case series, accounted for 12% of facial nerve paralysis1
- Presentation: Classic triad of otalgia, facial nerve palsy and vesicular eruption of the auricle and auditory canal2; see Fig. 1 for additional cutaneous and mucocutaneous manifestations
- Pathophysiology: Caused by the reactivation of varicella zoster virus in the geniculate ganglion and often affects nerves in close proximity, such as CN VIII, by mechanisms of both VZV neuritis and inflammatory edema3, commonly causing vertigo, tinnitus and hearing loss2 (Fig. 2)
- Diagnosis: Clinical alone; LP has no role in diagnosis, though CSF is abnormal in about 60% of patients with RHS5
- Treatment: Combination antiviral and steroid therapy; despite lack of evidence for their use in this syndrome when systematic reviews were undertaken9,8, Eye cares are an important adjunctive therapy.
- Prognosis: Recovery tends to be less favorable than Bell’s Palsy3

Herpes Zoster and HIV Infection
- Age-adjusted relative risk of HZ in HIV patients was 16.9 in 1992 study10
- Recurrent infections occur in 10-27% of HIV infected patients, compared to 1-4% of immunocompetent patients1
- Those with lower CD4 count are at higher risk and antiretroviral therapy appears to be protective3
- Complicated disease, particularly ocular and neurologic, is more common (see Table 2)

Take Home Points
- Ramsay Hunt Syndrome, a rare manifestation of HZ, is characterized by otalgia, facial nerve palsy and vesicular eruption of the ear
- HZ can be complicated by neurologic involvement; these cases are more common in immunocompromised patients
- Patients with neurologic HZ disease should be monitored very closely and treated with antivirals given the potentially devastating consequences; eye cares are also important

References
7. You know I hate this! Yes, it’s true.