Ramsay Hunt Syndrome

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Ramsay Hunt syndrome, or Zoster Oticus, is caused by reactivation of latent varicella zoster virus (VZV) in the geniculate ganglion of the 7th cranial nerve. The complex, proximal anatomy of the 7th and 8th cranial nerves results in the various manifestations of this rare clinical entity. As in zoster, Ramsay Hunt syndrome is more common in immunosuppressed and elderly hosts.

The manifestations of Ramsey Hunt syndrome include ipsilateral peripheral 7th nerve palsy with or without herpetiform rash and vestibulocochlear symptoms (3). The rash occurs in areas of afferent innervation of the geniculate ganglion: the palate, the anterior two-thirds of the tongue and the external ear (including the tympanic membrane).

The rash may also be absent (zoster sine herpete). Eighth cranial nerve ganglia may also be affected due to compression or vascular compromise and can lead to symptoms of dizziness, vertigo, ataxia, decreased hearing, tinnitus, nausea, vomiting or nystagmus. Tinnitus and sensorineural hearing loss may persist and become chronic. Zoster sine herpete is suspected when 7th nerve palsy is present with 8th nerve findings. Onset is usually more rapid and is usually more prolonged and severe than Bell’s palsy.

Diagnosis is confirmed with VZV PCR (1) of saliva or retrospectively with serology.

Classic Ramsay Hunt syndrome is a clinical diagnosis.

Early treatment is indicated to reduce chronic sequellae from Ramsay Hunt syndrome. Treatment includes both antivirals (acyclovir 800mg five times a day or famciclovir 500mg three times a day) and prednisone 60mg a day for five days with or without taper (2). Intravenous acyclovir can be used if oral antivirals are not tolerated. A longer course of steroids for severe or prolonged symptoms has not been studied but should be considered.

REFERENCES

