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Ramsay Hunt Syndrome

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History of present illness: Patient is a 70-year-old female who presented to the emergency department for left-sided facial numbness and facial paralysis associated with left ear pain for 4 days. She had been unable to close her left eye, and had experienced drooling out of the left side of her mouth. On exam, she had a left-sided cranial nerve VII palsy, swelling of the left pinna and inner ear canal, with mild erythema and swelling of the left auricle. Notably, there were small vesicles found within the left external ear and posterior to left ear. Tympanic membranes were clear bilaterally.

Significant findings: Left-sided cranial nerve VII palsy with flattened forehead creases, inability to keep the left eye open, and drooping of the corner of mouth. Vesicular lesions were found in and posterior to the left ear in a unilateral, dermatomal distribution.

Discussion: Ramsay Hunt syndrome (RHS) is a clinical diagnosis characterized by otalgia, vesicular lesions of the auditory canal and auricle (herpes zoster oticus), and ipsilateral facial paralysis.\(^1\) It is associated with reactivation of the varicella zoster virus (VZV) in the geniculate ganglion.\(^2\) The risk is highest in the elderly, females, Caucasians, and those with a family history of shingles.\(^3\) RHS has an incidence of about 5 in 100,000 population in the US, and may be associated with up to 20% of cases of Bell’s palsy due to cranial nerve VII (lower motor neuron) dysfunction.\(^4,5\) The mainstay of RHS treatment includes oral antivirals (e.g. acyclovir); however, there is no strong evidence that it improves outcomes.\(^4\) RHS due to VZV reactivation has been associated with the development of vasculopathies that increase the risk for aneurysms, intracranial hemorrhage, and stroke.\(^6\) This highlights the importance of making the correct diagnosis in seemingly straightforward cases of unilateral cranial nerve VII palsy, as the diagnosis of RHS is an established risk factor for stroke and other serious outcomes associated with VZV vasculopathies.\(^7\)

Topics: Herpes zoster, shingles, Bell’s palsy, dermatology, neurology.

References: