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CLINICAL VIGNETTE

On the Hunt for Ramsay Hunt

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A 48-year-old otherwise healthy male presents with symptoms of left ear pressure and pain along the left face, and low-grade fever for two days. Vitals: BP 137/77, Pulse 76, Temp 37 °C, Resp 12, Ht 5' 4.5", Wt 129 lbs. 6 oz, BMI 21.86 kg/m². Physical exam findings include normal facial symmetry, without rash. HEENT notable for edematous left external ear canal with erythematous and abnormal left TM, which was bulging and erythematous. Normal nose, mouth and throat. Neck: mild anterior cervical adenopathy, otherwise supple with midline trachea. He was diagnosed with acute non-suppurative left otitis media and prescribed amoxicillin 500mg three times daily for 10 days, ibuprofen 600mg every six hours as needed, and neomycin-polymixin-hydrocortisone 1% otic solution into left ear three times a day. The patient called back the next day stating that the ibuprofen was not helping to relieve the pain in the left ear, neck and throat. He requested for a different pain reliever, and was prescribed nabumetone 750mg once daily for pain relief.

Patient returned the next day for follow up of worsening symptoms. The left ear pain and pressure had been gradually worsening since initial evaluation two days ago, despite taking prescribed antibiotics as directed, ear drops and second pain reliever. Repeat vitals were unremarkable and physical exam findings included normal facial symmetry, without rash and normal appearing left external ear canal with normal TM and mild anterior cervical adenopathy. Patient was diagnosed with otalgia of the left ear and possible influenza infection. He was advised to finish the course of amoxicillin and oral prednisone taper was added for pain control. Of note POCT rapid influenza A/B tests were both negative.

The following day, patient presented to the local ER for evaluation of worsening pain despite taking antibiotics, pain reliever, and steroid. He reported a loss of appetite and inability to eat. When he tried to drink water and swallow, he had to spit it back up again. He also developed a hoarseness of his voice.

Physical exam findings in the ER: included normal vital signs. HEENT included: markedly dry mucous membranes, and left EAC and TM with erythema, retraction and swelling. Gastro-intestinal: soft, nontender, non-distended and neuro exam was free of focal deficits. Mental status was normal. He was admitted on a Friday night for dehydration and further evaluation of his inability to swallow. GI consultation was obtained and EGD on HD#3 showed ulceration in the cricopharynx, and a few small ulcers in the esophagus measuring 2-3mm as well as gastritis. On HD #4 patient developed some

hearing loss on the left side, as well as a rash on the left ear and left cheek area. He reported new facial asymmetry. ENT was consulted and flexible fiberoptic laryngoscopy showed two ulcers approximately 1mm in diameter along the lingual surface of the epiglottis, which was also somewhat inflamed. Examination of the larynx showed the left true vocal fold to be immobile and in the paramedian position, with fully mobile right true vocal fold. He was diagnosed with Ramsay Hunt Syndrome with multiple neuropathies of the cranial nerves V, VII, VIII, and X. He was started on valacyclovir and high dose steroids. His symptoms stabilized and he was discharged home on HD#8. He was advised that Ramsay Hunt Syndrome may result in permanent neurologic deficit as a result of inflammation of several cranial nerves.

Patient returned to ER six days after hospital discharge because of continued severe pain with swallowing and inability to eat. He reported weight loss of 17 pounds since onset of symptoms. He was unable to swallow his medications, felt dehydrated, and reported the muscles in his throat were not working. He was admitted for dehydration, malnutrition, and pain control.

Discussion

Herpes Zoster, shingles, results from the reactivation of latent varicella-zoster virus that gains access to sensory ganglia during initial varicella infection. It is characterized by a painful, unilateral vesicular eruption, which is limited to a specific dermatomal distribution. Complications can occur with shingles, such as postherpetic neuralgia, herpes zoster ophthalmicus with involvement of the ophthalmic division of the fifth cranial nerve, acute retinal necrosis, and Ramsay Hunt syndrome or herpes zoster oticus.¹ J. Ramsay Hunt described the various clinical presentations of facial paralysis and rash. He also reported other frequently associated symptoms such as tinnitus, hearing loss, nausea, vomiting, vertigo, and nystagmus, and explained that these eighth nerve features by the close proximity of the geniculate ganglion to the vestibulocochlear nerve within the bony facial canal.² Hyperacusis (dysacusis) occurs in some patients, and decreased tearing.³ Recognition of the general somatosensory function of the facial nerve and defining of the geniculate zone of the ear, with this syndrome which bears his name. Ramsay Hunt Syndrome is caused by the varicella zoster virus, and the strict definition of the syndrome are ipsilateral facial paralysis, and erythematous vesicular eruption on the ear or in the mouth.² Cranial nerves V, IX, and X are often affected. Compared to Bell's palsy (facial paralysis without rash), patients with Ramsay Hunt syndrome have more

severe paralysis at onset and are less likely to recover completely.² It is less common than Bell's Palsy, with new case reports of Ramsay Hunt occurring every 53 minutes as compared to every 10 minutes for a new case of Bell's palsy.³

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