ABSTRACT

Objectives: The evaluation of asymmetric sensorineural hearing loss (SNHL) typically includes an audiogram and gadolinium enhanced MRI to inspect for retrocochlear pathology, most commonly a vestibular schwannoma. Previous reports have identified nonneoplastic lesions mimicking vestibular schwannoma. To our knowledge, this is the first case reported of an isolated cochlear neuritis secondary to varicella reactivation, which clinically and radiographically mimicked the presentation of a vestibular schwannoma.

Study Design: Case Report

Methods: A case is presented of a patient with progressive unilateral SNHL and tinnitus with internal auditory canal (IAC) enhancement on MRI secondary to isolated cochlear neuritis from varicella reactivation. MRI images are presented showing resolution of IAC enhancement following varicella treatment.

Discussion: For an otolaryngologist, varicella reactivation is most commonly seen in the form of Ramsay Hunt syndrome or shingles of head and neck dermatomes. Ramsay Hunt syndrome is known to produce abnormal MRI enhancement within the IAC from the local facial and vestibulocochlear neuritis; however, its characteristic clinical signs aid the diagnosis. Our case is unique in that the patient’s only manifestation of his varicella infection was unilateral SNHL manifesting as an MRI lesion indistinguishable from a vestibular schwannoma.

Conclusions: This case outlines the importance of maintaining a broad differential diagnosis in the evaluation of unilateral SNHL. MRI remains an important tool in vestibulocochlear neuritis diagnosis; however, its limited specificity must be recognized.

CASE PRESENTATION

The patient was a 41-year-old male with a history of AIDS who presented for evaluation of asymmetric SNHL. He reported progressive left-sided hearing loss for the past four months with intermittent tinnitus. The patient had no evidence of facial weakness or auricular vesicles on the affected side. An audiogram confirmed profound left-sided SNHL as well as mild right-sided SNHL. An MRI showed abnormal enhancement within the left IAC concerning for a vestibular schwannoma (Fig 1). Upon closer examination, there was subtle biformal leptomeningeal enhancement which raised suspicion for a viral or opportunistic etiology given the patient’s immune status (most recent CD4 cell count was 4). A lumbar puncture was positive for varicella zoster via polymerase chain reaction (PCR) testing. The patient was treated with a three-week course of intravenous acyclovir. A post-treatment MRI showed resolution of both the IAC and bifrontal enhancement, further confirming the infectious etiology (Fig 3). A repeat audiogram showed unchanged, profound left-sided SNHL. At no point in the treatment course did the patient develop facial weakness or auricular vesicles.

INTRODUCTION

Evaluation of hearing loss typically includes an audiogram and, if asymmetric sensorineural, an auditory brainstem response (ABR) study or gadolinium enhanced MRI to inspect for retrocochlear pathology. Approximately 2.7-10.2% of patients with sudden sensorineural hearing loss (SNHL) are found to have an internal auditory canal (IAC) lesion on MRI, most commonly a vestibular schwannoma. Vestibular schwannomas appear as areas of enhancement on postcontrast T1 weighted images. Ramsay Hunt syndrome, secondary to varicella reactivation, is known to cause abnormal MRI enhancement in the IAC due to inflammation of the facial nerve. Latent varicella virus has been isolated in the spinal ganglion and reactivation here may have a potential cause of hearing loss.

IMAGING

• Vestibular schwannoma is by far the most common enhancing IAC lesion; however, an isolated neuritis can have a similar appearance.

• Viral reactivation was the likely cause of this patient’s hearing loss given his positive CSF PCR for varicella, immunocompromised state, and resolution of IAC enhancement following acyclovir treatment.

• Approximately 70% of patients with Ramsay Hunt syndrome show enhancement of both the facial and vestibulocochlear nerves on MRI secondary to inflammation.

• Latent varicella virus has been isolated in the spiral ganglion and reactivation here may have been a source of hearing loss.

• Several cases of Ramsay Hunt syndrome presenting with isolated hearing loss have been described; however, these patients developed subsequent facial paralysis and auricular vesicles.

• Specialized imaging sequences, such as FIESTA and CISS, are nonenhancing techniques which limit sensitivity to inflammation and are useful in diagnosing neoplastic lesions.

DISCUSSION

• Vestibular schwannoma can be a potential cause of unilateral hearing loss, particularly in immunocompromised patients.

• Abnormal MRI enhancement in the IAC can have various causes including neoplastic, infectious, and inflammatory.

• FIESTA/CISS imaging sequences help distinguish inflammatory lesions from neoplastic lesions.

CONCLUSIONS

• Varicella reactivation can be a potential cause of unilateral hearing loss, particularly in immunocompromised patients.

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REFERENCES

Adam Goodale, MD
University of Cincinnati - Department of Otolaryngology, Head and Neck Surgery
Email: adam.goodale@uc.edu

Isolated cochlear neuritis from varicella reactivation mimicking a vestibular schwannoma

Adam D. Goodale, MD1; Judging Gulab, MD2,3; Rebecca S. Cornelius, MD2; Carl J. Echtenbaum, MD2; Ravi N. Sany, MD1,4

1Department of Otolaryngology, Head and Neck Surgery, University of Cincinnati Medical Center
2Department of Neurosurgery, University of Cincinnati Neuroscience Institute
3Department of Infectious Diseases, University of Cincinnati Medical Center
4Department of Infectious Diseases, University of Cincinnati Medical Center

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