ABSTRACT

Objective: Profound sensorineural hearing loss (SNHL) is a potentially serious and irreversible complication of viral labyrinthitis. We present our case report to highlight the challenges in early diagnosis of unilateral deafness in a young child. We would advocate for early, comprehensive audiologic testing and subsequent temporal bone imaging in children presenting with vestibular symptoms in the setting of an upper respiratory infection (URI).

Study Design: Case report with review of medical records and current literature.

Results: A 5-year-old female presented with unilateral profound hearing loss 3 years following a URI associated with disquilibrium. Documented ear examination and head computed tomography (CT) were normal and the differential diagnosis was post-viral cerebellitis versus labyrinthitis. After the patient presented to us with subjective worsening hearing, an audiogram demonstrated profound, unilateral SNHL and a temporal bone CT demonstrated extensive, unilateral labyrinthitis ossificans. She underwent unilateral hearing aid placement for single-sided deafness.

Conclusion: We propose that in children presenting with vestibular symptoms in the setting of a URI, serial audiograms and imaging studies should be added to the diagnostic algorithm.

INTRODUCTION

Labyrinthitis ossificans (LO) is a pathologic heterotopic bone formation within the otic capsule. Ossification occurs in response to inflammatory or destructive changes in the inner ear from the following infection, most commonly bacterial meningitis, temporal bone trauma, autoimmune disease, otosclerosis, leukemia, other malignancy, or vascular occlusion. (1,2) Presenting symptoms of LO may include vertigo, hearing loss, tinnitus, hearing loss, otitis media, otalgia, visual disturbances, nausea and vomiting, fever, neck pain or stiffness, and current or recent URI.

Labyrinthitis ossificans is most commonly caused by bacterial invasion, which occurs via one of 3 routes: hematogenous spread from cochlear vasculature, otitis media pathogens passing through the round window membrane, or, most commonly, meningogenic spread from the subarachnoid space. Hearing loss may be seen as early as 48 hours after infection in meningitis, with ossification visualized intraoperatively as early as 21 days post-infection and demonstrated on hi-resolution CT (HRCT) within one year. (3)

Children presenting with acute disquilibrium provide a diagnostic dilemma, largely due to the patient’s inability to provide a precise description of symptoms. A detailed history from caregivers including onset, recent infection, history, injury or potential toxin exposure, progression, and associated symptoms such as vomiting, or headache should be investigated. Comprehensive ophthalmologic, otologic, neurologic, and vestibular examinations are warranted prior to proceeding with imaging. (4)

The differential diagnosis for pediatric disquilibrium encompasses a diversity of conditions including: infectious and autoimmune disorders of the cerebellum, meninges, and labyrinth, alcohol, drug, or toxin exposure, head trauma, intracranial masses or lesions, atypical seizure disorder, migraine phenomena, psychogenic dissequilibrium, and genetic ataxia disorders.

CASE PRESENTATION

A 5-year-old female presented to our practice for evaluation of a newly diagnosed, profound, unilateral hearing loss. The child was healthy at birth, passed her newborn ALGO hearing screening, and had no known risk factors for hearing loss. Her speech and language development had been normal.

Her parents recalled a febrile URI with nausea and vomiting 3 years prior. One week into the viral illness, the child, then 2 years old, was improving and had defervesced, then became irritable and severely ataxic. She was afebrile upon presentation to a local Emergency Department (ED) with evaluations documenting ataxia with otherwise normal exam. The patient reportedly could not maintain her balance for more than 3 seconds without becoming unstable. A head CT was marred by motion artifact but interpreted as normal. Neurologist’s diagnosis was post viral cerebellitis versus labyrinthitis. After observation for 2 days, she was discharged to home. No treatment, follow-up testing, or imaging was recommended and the ataxia gradually resolved.

The parents began to suspect unilateral hearing loss in the patient’s 5th year of life and a hearing screening was abnormal unilaterally. A pure tone audiogram (see Figure 1) confirmed profound SNHL across all frequencies on the right, so referral to Pediatric Otolaryngology was made. An HRCT of the temporal bone (see Figure 2) demonstrated a normal left inner ear and hypertensive basal and middle turns of the right cochlea with a heterotensive vestibular apparatus. Diagnosis of LO was reviewed with the family and they were counseled regarding the permanent nature of the hearing loss. Discussion ensued about amplification and the family ultimately chose to proceed with an osseointegrated temporal implant.

DISCUSSION

In the case of our patient, imaging at the time of disquilibrium was normal and there was no obvious source of supplicative labyrinthitis. Therefore, viral labyrinthitis was the most responsible cause of her LO. We suspect this was from a hematologic source, given the normal ear exam and lack of systemic symptoms. Literature review indicates that labyrinthitis ossificans from a tympanic source is outnumbered by meningeal causes. (5,6) It is not entirely clear why, in our patient, unilateral disease resulted from a presumably systemic illness.

Most causes of acquired sensorineural hearing loss in childhood can be associated with vestibular complaints. Bacterial labyrinthitis of any type accounts for one in three cases of all acquired hearing loss. (7) Viral labyrinthitis carries fewer morbidities and mortalities than bacterial, but is much more common. We believe that a hearing evaluation for any child presenting with vestibular complaints is warranted, though there is no data on this in the literature.

Unilateral sensorineural hearing loss of childhood is often investigated with high resolution computed tomography (HRCT) of the temporal bone. The risks of CT have recently been highlighted in several papers, with greater life-time cancer risk in the pediatric population. HRCT of the temporal bone can be performed with a lowered dose protocol and preferential angle to minimize the radiation to the child, and specifically away from the ocular lenses. Careful patient education and pre-procedure conversation with parents regarding the risks of CT should be a part of the decision making process in the workup of pediatric hearing loss.

CONCLUSIONS

Profound sensorineural hearing loss is a potentially serious and irreversible complication of viral labyrinthitis. We present our case report to highlight the challenges in early diagnosis of unilateral deafness in a young child. We would advocate for early, comprehensive audiologic testing and, when indicated, temporal imaging in children presenting with vestibular symptoms.

REFERENCES