Hearing loss in Vogt-Koyanagi-Harada Syndrome Treated with Intratympanic Steroids

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Abstract

Educational Objective:
At the conclusion of this presentation, the participants should be able to describe Vogt-Koyanagi-Harada (VHK) Syndrome, know it as a rare cause of inner ear symptoms, and recognize the possible utility of intratympanic steroid injections for the treatment of sensorineural hearing loss (SNHL) in these patients.

Objectives:
• The case of a patient with VHK Syndrome and SNHL treated successfully with intratympanic steroid injections is described.
• The otologic manifestations of VHK Syndrome and treatment options reported in the literature are discussed.

Methods:
• Chart review, with discussion of the pertinent history, clinical course, and audiologic studies
• Literature review

Results:
The patient is a 68 year-old woman previously diagnosed with VHK Syndrome, presenting to the otologist with long-standing fluctuating vertigo, tinnitus, and bilateral sensorineural hearing loss. She experienced improvement in her symptoms after intratympanic steroid injections. This is the first report of a patient with VHK Syndrome treated successfully with intratympanic steroids.

Conclusion:
In patients with VHK syndrome who experience sudden or fluctuating SNHL, intratympanic steroids are an effective treatment option.

Case Presentation

A 68 ear old female presented with worsening hearing in her right ear. She had a history of Vogt-Koyanagi-Harada syndrome, with symptoms of chronic uveitis, vitiligo, and fluctuating bilateral sensorineural hearing loss and tinnitus. The patient had been previously treated for an acute hearing deterioration in her right ear with oral corticosteroids, which subsequently improved both her hearing and uveitis-related visual complaints. However, she also developed side effects related to underlying hypertension and diabetes mellitus, including an increase in blood pressure and blood glucose levels. She now presented with a new-onset decline in hearing and worsening tinnitus in the same ear.

Otolologic examination was normal bilaterally. The patient was noted to have patches of vitiligo on her face, arms, legs, and trunk. Audiometric testing showed moderate-to-severe SNHL in the right ear and moderate SNHL in the left ear at the speech frequencies (figure 2a). Speech discrimination scores were 80% in the right ear and 84% in the left ear.

Because the patient had previously experienced adverse effects to systemic corticosteroid administration, the decision was made to proceed with intratympanic dexamethasone injection. A total of 0.5 mL of dexamethasone 10mg/mL was injected into the right middle ear for chemical labyrinthotomy. The patient experienced transient dizziness, but no other temporary or long-term complications were observed. A repeat audiogram was performed four weeks following injection, showing a slight (10dB) improvement at 1 kHz. A second intratympanic injection was then performed. Subsequent audiometric exam performed two weeks after this second injection showed a 5-10 dB improvement across multiple frequencies (250-1000 Hz; see figure 2). Speech discrimination scores were not statistically improved (84% for right ear, and 88% for left ear). The patient also reported substantial improvement in her right ear tinnitus.

The patient has been followed clinically from the time of injection up until the present day, a period spanning 1.5 years. Serial audiometry over this time has shown small fluctuations (10-15db) in bilateral sensorineural hearing thresholds. These changes generally coincide with the variable severity of her visual and dermatologic manifestations. Her condition has not necessitated additional intratympanic injections.

Discussion

Vogt-Koyanagi-Harada (VHK) Syndrome is an autoimmune condition with ocular, neurologic, auditory, and dermatologic manifestations. The disorder targets organs containing melanocytes of neural crest origin. Patients most commonly affected are typically between ages 20-50 and of non-Caucasian ethnicity, suggesting a genetic component to the disorder.

Patients with VHK syndrome commonly present with symptoms of aseptic meningitis followed by bilateral uveitis. Prodromal meningeal symptoms occur in approximately half of cases and can last for weeks. Ocular symptoms manifest as decreased visual acuity and eye pain or irritation several days after the onset of meningeal symptoms. Eye exam may show optic nerve hyperemia, posterior choroid inflammation leading to retinal detachment, and inflammatory infiltrates within the iris, aqueous humor, and vitreous humor. Dermatologic changes present several weeks or months after the onset of ocular symptoms, and they may include poliosis, vitiligo, and alopecia. Dysacusis associated with VHK syndrome occurs with variable frequency ranging anywhere from 8-70%.1 Patients may experience hearing loss, tinnitus, and/or vertigo that typically coincides with the onset of ocular pathology.

Diagnosis of VHK syndrome is based upon the presence of a constellation of ocular and dermatologic findings initially outlined by the American Uveitis Society in 1978, and later modified in 2001.2 A thorough otologic evaluation includes fundoscopic examination and possibly fluorescein angiography, while assessment for suspected meningitis may necessitate lumbar puncture. The mainstay of evaluation for the auditory symptoms of VHK syndrome is the audiogram, although the nature and extent of hearing loss associated with VHK syndrome has not been well described. A recent series of patients with VHK syndrome revealed 5 of 24 individuals to have sensorineural hearing loss greater than that of age-matched control populations from previously published literature.3 However, these hearing losses had no clear pattern with respect to the cochlear frequency.

The most commonly utilized treatment for the ocular manifestations of VHK syndrome are systemic corticosteroids. Patients with sensorineural hearing loss associated with VHK syndrome are also typically treated with oral steroids. While no study has specifically evaluated their effectiveness in VHK syndrome, several studies have also indicated a benefit in smaller numbers of patients with autoimmune inner ear disease modest but significant improvements in hearing have previously been demonstrated in patients who received oral prednisone over a four week period.4

Most clinical evidence supporting the use of intratympanic steroids is in patients with sudden sensorineural hearing loss or Meniere’s disease.5-7 Several studies have also indicated a benefit in smaller numbers of patients with autoimmune inner ear disease, although no prior study has described their use specifically for treatment of VHK syndrome-related SNHL.8 Current indications for intratympanic steroids are when oral steroids fail to improve hearing, or when there is a contraindication to systemic steroid administration. The most effective type and dose of steroids used, technique of delivery, and number of injections has not reached consensus in the literature due to the retrospective nature of available data. Nevertheless, the results seen in our patient are promising and provide further evidence supporting the use of intratympanic steroids in VHK syndrome and other autoimmune inner ear disease, especially when there is a contraindication to use of systemic steroids.

Conclusions

VHK syndrome should be recognized as a potential cause of dysacusis and sensorineural hearing loss, particularly in patients with ocular and dermatologic symptoms. In patients with this disorder who experience otologic manifestations, intratympanic steroids are an option for those who do not tolerate systemic therapy.

References