Cogan's Syndrome: Auditory and Medical Management

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Abstract

Cogan's syndrome is a rare autoimmune disease characterized by the presence of interstitial keratitis and audiovestibular symptoms. The audiovestibular symptoms include fluctuating sensory hearing impairment, tinnitus, vertigo, and reduced vestibular response. Immediate diagnosis and medical intervention provides optimum auditory recovery. Frequent audiologic assessments are necessary to monitor the disease activity and to aid in the therapeutic levels of steroidal medications. Amplification is often required on a temporary or permanent basis. Two case studies are presented to illustrate the audiologist's role in the identification and management of patients with Cogan's syndrome.

Key Words: Cogan's syndrome, interstitial keratitis, autoimmune disease, vasculitis, hearing loss, amplification, corticosteroid therapy

Cogan's syndrome (Cogan, 1945) is considered a nonsyphilitic, autoimmune disease characterized by the presence of interstitial keratitis in conjunction with vertigo, tinnitus, and hearing loss (Smith, 1970; Peeters et al, 1986; Barna and Hughes, 1988; Hughes et al, 1988; Jahrsdoerfer et al, 1988). Other symptoms may include vasculitis and general systemic complaints such as fever, weight loss, and fatigue. Histopathologic evidence indicates infiltration of lymphocytes and plasma cells in the cornea and the spiral ligament (Hughes et al, 1983; McDonald et al, 1985; Barna and Hughes, 1988).

Cogan's syndrome affects both men and women with onset ranging from 20 to 50 years of age (median age 25 years). The interstitial keratitis component presents clinically as photophobia, decreased visual acuity, ocular pain and redness (Peeters et al, 1986), and to a lesser degree, conjunctivitis, uveitis, and scleritis have also been observed (Hughes et al, 1983). Other possible laboratory findings may include medium and small vessel vasculitis and nonspecific inflammatory signs such as leukocytosis, thrombocytosis, anemia, and elevated erythrocyte sedimentation rate. Differential diagnosis involves ruling out other causes of interstitial keratitis (e.g., congenital syphilis), other forms of autoimmune inner ear disease, audiovestibular symptoms with eye involvement (e.g., viral labyrinthitis), or systemic diseases with eye, and ear complaints (e.g., relapsing polychondritis).

AUDITORY OR VESTIBULAR DEFICITS

The audiovestibular component presents clinically with a fluctuating hearing loss, vertigo, nystagmus, nausea, and vomiting (Bachynski and Wise, 1984; Morgan et al, 1984; Jahrsdoerfer et al, 1988). The hearing loss is sensory in nature and may be either unilateral or bilateral. Auditory sensitivity fluctuates between the range of normal limits and a severe hearing auditory deficit. Interestingly, pure-tone thresholds may remain stable from one evaluation to the next, yet word recognition ability may show significant deterioration or improvement. Typically, an initial evaluation
shows sensory hearing loss with abnormal electronystagmography.

The clinical course may begin with either ocular or audiovestibular complaints, but both are usually present within 1 month of onset. If untreated, severe to profound auditory deficits generally occur within 3 months (Vollertsen et al., 1986). Immediate recognition and intervention with high-dose corticosteroids (1.0–1.5 mg/kg/day) is essential to provide optimum auditory recovery. Studies suggest the administration of corticosteroids must be initiated within 2 weeks of initial presentation of hearing loss for adequate auditory restoration (McDonald et al., 1985; Laffin et al., 1987; Barna and Hughes, 1988).

**CASE REPORTS**

The following case reports describe the auditory and clinical findings of two young adult females who presented with Cogan's syndrome. Both patients demonstrated typical interstitial keratitis and associated fluctuating auditory deficits of varying degree and amelioration. Both patients received corticosteroids in the management of their disease and audiometric work-ups that demonstrated improvements in hearing sensitivity as a result. Although amplification was required during therapy for both patients, only one patient required long-term auditory rehabilitative management.

**Case 1**

CR is a 25-year-old Hispanic female who initially presented with left ear tinnitus. One week later she experienced sudden nausea, vomiting, and an ataxic gait. By the second week, there was evidence of significant left ear auditory deficits. The complaint of photophobia and "foggy vision" prompted the suspicion of Cogan's syndrome. Figure 1 illustrates the results of the first baseline audiologic evaluation. The patient was immediately placed on high-dose corticosteroids (Prednisone, 60 mg daily) while differential diagnosis was completed. Formal evaluation involved complete rheumatologic, ophthalmologic, and otolaryngologic examinations. Laboratory findings included nonspecific inflammatory signs of thrombocytosis, leukocytosis, elevated erythrocyte sedimentation rate, and hyperglobulinemia.

Initially, hearing sensitivity and visual acuity showed a rapid and dramatic improvement to the high-dose anti-inflammatory drug regimen. Figure 1 shows the audiometric improvements in left ear scores. Vestibular abnormali-

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**Figure 1** Audiometric records of patient CR. Serial audiograms cover an 8-month period showing fluctuation of pure-tone sensitivity.

### PURE TONE AUDIOMETRY

#### RIGHT EAR

<table>
<thead>
<tr>
<th>FREQUENCY (Hz)</th>
<th>250</th>
<th>500</th>
<th>1000</th>
<th>2000</th>
<th>4000</th>
<th>6000</th>
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<td>dB</td>
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#### LEFT EAR

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<th>FREQUENCY (Hz)</th>
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### SPEECH AUDIOMETRY

(PB word recognition scores)

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<tr>
<th>HL in dB</th>
<th>0</th>
<th>50</th>
<th>80</th>
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<tr>
<td>%</td>
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<td>50</td>
<td>80</td>
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ties were somewhat slower to improve however, and a residual ataxic gait remained. After 1 month of treatment, a cautious steroid taper was begun. Three months into the steroid taper, the patient experienced a significant flare-up with new right ear pain, tinnitus, and further hearing loss that is presented in Figure 1. Amplification was then initiated to support communication demands. Steroid treatments were immediately increased to previous levels. Ocular steroids and atropine were begun to address ocular inflammation. After development of upper extremity weakness, immunosuppressants were further increased by the addition of IV-pulse methylprednisolone and a cytotoxic agent—azathioprine.

With steroid administration, CR had a subsequent improvement of all symptomatology and although a mild hearing loss remained, amplification was no longer required. CR remained audiologically stable throughout the second steroid taper as demonstrated in Figure 1. Audiologic management continues routinely and although a mild decrease in pure-tone sensitivity has been observed above 3000 Hz in the right ear, speech recognition has remained stable with no subjective change in right ear sensitivity. Currently, the patient does not use amplification but recognizes the potential need given possible fluctuations in decreased auditory sensitivity.

Case 2

LC, a 27-year-old white female, initially presented clinically with ocular symptoms of erythema, pain, and photophobia. This was interpreted as an allergic reaction and discontinuation of contact lens use was recommended. In the following 2 weeks, the diagnosis of interstitial keratitis was made and treated locally. Following, she experienced sudden onset of vertigo, nausea, vomiting, and left ear sensory hearing loss. Initial baseline audiometrics are seen in Figure 2. Symptoms worsened over the next 6 weeks and a moderate-to-severe hearing loss was recorded bilaterally. The eventual diagnosis of Cogan’s syndrome finally initiated corticosteroid therapy. Although left ear symptoms improved with a subsequent dramatic improvement in word recognition ability, right ear abnormality continued to deteriorate. Figure 2 reflects the decrease in right ear sensitivity with essentially stable left ear results.

As the disease entered a remission state, hearing sensitivity began to stabilize and steroid treatments were tapered off to cessation. Figure 3 shows the results of testing performed
during remission of the disease. Of particular interest are the unexplained fluctuations of speech recognition scores with little change in pure-tone thresholds. This phenomenon reflects the often unrecognized existing differences between the physiologic complexity of speech processing and the comparative rudimentary requirements of pure-tone measurement.

One year following remission, symptomaticology recurred with associated sudden left ear decreased auditory sensitivity (see Fig. 3). The reinstitution of steroid treatments was successful in re-establishing the previous baseline audiometric in the left ear, however, little overall auditory recovery was observed. Immunosuppressive therapy has been discontinued and no further recovery is expected. The most recent audiometric evaluation is shown in Figure 3. LC was initially given an FM system as she pursued speech reading classes, psychological counseling, and coping strategies through various resources. She presently wears binaural amplification for daily communicative needs. LC is now accustomed to monitoring her auditory acuity and speech recognition abilities and returns for periodic re-evaluation and hearing aid monitoring. This case clearly demonstrates the need for longitudinal audiologic management in Cogan’s syndrome.

**COMMENTS**

Although Cogan’s syndrome is rare, it should be considered a possibility in patients presenting with audiovestibular symptoms similar to Meniere’s disease. This suspicion should be heightened by the concurrent presence of ocular symptoms. As demonstrated in the two case reports, prompt corticosteroid treatment should be initiated to prevent permanent severe sensory hearing impairment while formal diagnostic evaluation is conducted. It becomes clear that audiologic documentation of fluctuating hearing loss is a primary criterion in the diagnosis and management of Cogan’s syndrome. Frequently, treatment decisions are based on audiologic evaluation, and therefore, open dialogue between collaborators is necessary during the course of the disease process.

The optimum use of amplification in the rehabilitative management of Cogan’s syndrome is challenging. Due to the fluctuating auditory nature of the syndrome, a versatile hearing aid and thorough patient counseling is recommended. Amplification may be necessary at any time and therefore continuous audiologic monitoring is a requirement. Hearing sensitivity...
may fluctuate such that very different styles and models of hearing aids may be required. The patient must be aware of the flexibility and limitations of each hearing aid in order to utilize appropriate fitting. Because auditory deficits may be the first indication of disease exacerbation, long-term audiologic follow-up is necessary to monitor the disease activity. Finally, these case reports unquestionably demonstrate the required working relationship between patient, physician, and audiologist in the diagnosis and medical and audiologic management of Cogan's syndrome.

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REFERENCES


