

Fluctuating Hearing Loss Responding to Corticosteroids: A Case of Cogan's Syndrome Concomitant with Crohn's Disease

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Abstract

Hearing loss has a broad differential diagnosis. Hearing impairment responding to corticosteroids is suggestive of an underlying autoimmune cause. We report a rare case of Cogan's syndrome in a 28-year-old woman with no previous personal medical history. She had experienced two years of fluctuating hearing loss and bilateral keratitis that would respond to high-dose prednisone prescribed by her otorhinolaryngologist. We performed a complete work-up and ruled out most common causes of autoimmune inner ear diseases. Patient was diagnosed with Cogan's syndrome based on clinical findings and prescribed corticosteroid-sparing immunosuppressive medication. During follow-up, she developed worsening fatigue, new onset anemia, and melena. Endoscopic and pathologic findings were diagnostic of Crohn's disease. This case emphasizes the high prevalence of concomitant autoimmune diseases.

Keywords: Autoimmune disease; Cogan's syndrome; Crohn's disease; Hearing loss; Inner ear

Case Report

A 28-year-old woman was referred to our internal medicine clinic by her otolaryngologist to pursue investigations for recurrent disabling episodes of hearing loss that would respond to systemic corticosteroids. The patient had no particular personal medical history and had a family history of sarcoidosis and Crohn's disease in first-degree cousins. Two years before referral, she experienced an acute episode of left ear sensorineural hearing loss in the high frequencies (>2000 Hz) with tinnitus and severe vertigo without a precipitating event. Previous initial investigations at the time revealed mild systemic inflammation with a C-reactive protein at 14 µg/ml (normal <4), with normal CBC and blood biochemistry, and negative viral and bacterial serology. A head CT-scan was performed and was interpreted as normal. She received a diagnosis of probable sudden sensorineural hearing loss (SSNHL) and was prescribed systemic prednisone empirically.

Her vestibuloauditory symptoms improved significantly with corticosteroids initially. However, with tapering down doses of prednisone, she relapsed with substantial decreased hearing acuity. Over the course of the next two years, she had monthly recurring episodes that necessitated new courses of high-dose prednisone of 1 mg/kg daily for up to 2 weeks. Finally, her symptoms became refractory to corticosteroids and serial pure-tone audiograms revealed irreversible severe unilateral sensorineural hearing loss in the low to mid frequencies (250-2000 Hz) (Figure 1).

Upon further questioning on possible previously missed symptoms, patient mentioned to us that concomitantly to her left inner ear dysfunction, she developed bilateral painful red eyes with photophobia. She noted that these ocular symptoms seemed to disappear while taking prednisone. Ophthalmic examination off medication demonstrated a non-specific bilateral interstitial keratitis.

We suspected from patient's history, audiologic, and ophthalmic findings the possibility of Cogan's syndrome. Remaining questionnaire did not provide further clues. Physical examination was unremarkable. ANA titers were mildly positive at 1/160. Other autoimmune antibody markers were negative. The rest of the laboratory investigations did not provide an alternative explanation to her presentation. MRI of the brain and internal auditory canals did not reveal white matter anomalies such as hypo- or hyperintensities, pathological gadolinium enhancing lesions, or structural anomalies. Diagnosis of Cogan's syndrome was made and azathioprine, a corticosteroid-sparing immunosuppressive medication, was started.

On follow-up at our clinic, patient mentioned worsening fatigue, abdominal cramps, and melena. CBC showed a new anemia with Hb of 104 g/L (normal range 120-160). Upper and lower GI endoscopic evaluation revealed severe gastritis and mild terminal ileitis. Biopsies were performed and pathological examination showed chronic

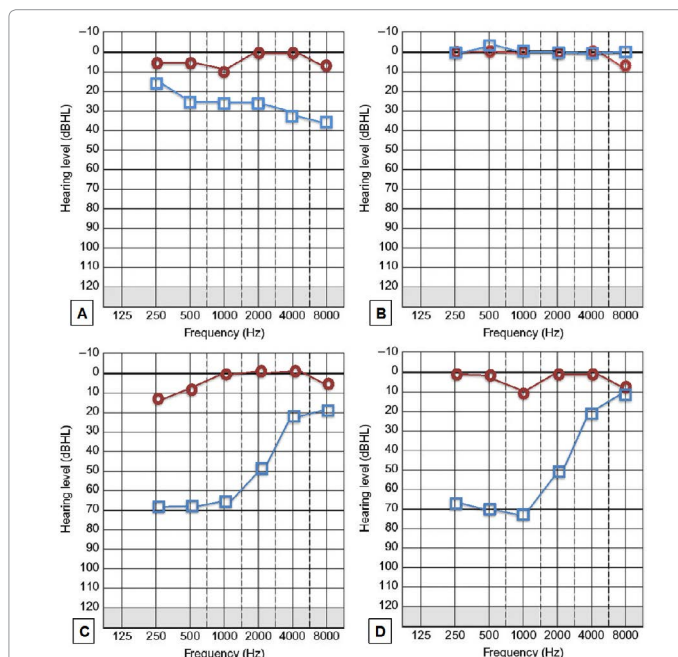


Figure 1: Pure-tone audiometry: (A) before treatment on January 2011, (B) after treatment on June 2011, (C) during a relapse on November 2012, and (D) despite retreatment on October 2013. Blue lines - left ear, red lines -right ear.

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inflammation with presence of granulomas and plasmacytomas diagnostic of Crohn's disease.

Discussion

Internal medicine physicians, especially when faced to young patients, rarely question symptoms affecting the ears. Many systemic conditions, such as systemic lupus erythematosus, Behçet's disease, polyarteritis nodosa, and granulomatosis with polyangiitis, can cause vestibuloauditory symptoms. Hearing loss responding to corticosteroids should evoke in clinicians the possibility of primary and secondary autoimmune inner ear diseases (AIED), which are rare entities; they account for less than 1% of hearing impairment cases [1].

Cogan's syndrome (CS), the prototype of secondary AIED, is a rare disorder afflicting young adults and is characterized by non-specific interstitial keratitis, recurrent vestibuloauditory symptoms reminiscent of Ménière's disease, and progressive sensorineural hearing loss. Most cases evolve to unilateral or, more frequently, bilateral deafness within two years [2]. Recent evidence suggests that CS pathogenesis involves autoantibodies against corneal and inner ear proteins, CD148 and connexin 26, which have been detected in the serum of affected patients [3].

This case also highlights the importance of exploring for the possibility of co-occurring autoimmune diseases. Of the 250 cases of CS that have been reported in the literature, about 30% developed an associated inflammatory disease such as aortitis, rheumatoid arthritis, sarcoidosis, or, such as in the case of our patient, Crohn's disease [2-4]. Therefore, patients with CS require a complete extensive evaluation at diagnosis and during follow-up in order to identify potential co-occurring disorders and complications, including frequent laboratory studies such as CBC, urinalysis, creatinine, electrolytes,

liver transaminases, C-reactive protein and erythrocyte sedimentation rate, as well as thoracic and cardiac imaging (CT-scan, PET-scan, and echocardiogram) in order to rule-out aortitis and aortic insufficiency.

There is no gold standard treatment for patients with CS as there has been no randomized controlled study comparing performance of immunosuppressive medications. Prednisone is typically prescribed initially at a dose of 1 mg/kg daily as first line treatment. Corticosteroid-sparing immunosuppressive medications are often introduced in order to limit corticosteroid side effects and toxicity. Azathioprine, cyclophosphamide, cyclosporine A, methotrexate, rituximab, and anti-TNF alpha drugs such as infliximab and etanercept have all been used in case reports with varying degrees of success in patients with CS [3].

Identification of patients who have corticosteroid-responding hearing loss is essential since 15%-30% of them will have a secondary AIED such as CS [1]. These patients may therefore benefit from autoimmune and inflammatory disease screening and surveillance with a thorough clinical evaluation and laboratory studies including antibody panel.

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