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Cogan's Syndrome: A Case Report

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INTRODUCTION

Cogan's syndrome (CS) is an immune-mediated disease characterized by ocular inflammation and audiovestibular dysfunction with or without vasculitis or other systemic manifestations (1). Audiovestibular disease that is difficult to treat can lead to the loss of hearing (2). By a timely recognition and initiation of glucocorticoid therapy at the onset of the disease, poor outcomes, especially complete hearing loss, could sometimes be prevented (2). Certain patients may require long-term corticosteroid therapy because of recurrent hearing loss during attempts to taper the prednisone dose (3). We report Cogan's syndrome in a 14 year-old-girl with marked deterioration of hearing power following tapering of corticosteroid therapy.

CASE REPORT

A 14 -year-old Kuwaiti girl was admitted with vertigo and a bilateral decrease of hearing. Two months earlier, she visited an ophthalmologist for a red left eye, a diagnosis of mild uveitis was made, and she improved on corticosteroid eye drops. Clinical examination revealed bilateral sensorineural hearing loss and there were no other cranial nerve or central nervous system deficits. She did not show any stigmata of autoimmune or collagen disease. Pure tone audiogram showed bilateral mild high frequency sloping sensorineural hearing loss. Tympanometry demonstrated a curve of the "A" type bilaterally with absence of both ipsilateral and contralateral reflexes. Auditory brain stem response showed ill defined waveforms recorded down to 90 dBnHL indicating severe affection of auditory nerves. Magnetic resonance imaging (MRI) of the brain showed normal brain stem, cerebellum, and cerebellopontine angles with no evidence of demyelination. Laboratory investigations showed an ESR of 65 mm/Hr. and a total leukocyte count of $15x10^9$/L. with a neutrophilic predominance. Rheumatoid factor, antinuclear antibodies, and antineutrophil cytoplasmic antibodies (P&C-ANCA) were negative.

Keywords: Cogan's syndrome, Deafness, Eye inflammation, Vasculitis

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Anti-cochlea antibodies (anti-p68, anti-p58, anti-p38, and anti- p30) were also negative. MRI of the chest and abdomen showed no evidence of vascular abnormalities. Oral prednisone (60 mg/day) was started and, with the marked improvement in hearing power, the dose was tapered after 8 weeks to a maintenance level of 10 mg/day. Four months after the initial admission, and while on maintenance corticosteroids, she presented with severe recurrence of hearing loss, polyarthralgia, and a left lower motor neuron facial palsy. Pure tone audiogram showed severe sensorineural hearing loss (Figure 1). Pulse i.v. methyl prednisone therapy (1 gm/day) was given for 3 days and was followed by oral prednisone 60 mg /day. Clinical follow up showed partial improvement in both facial palsy and hearing acuity (Figure 1).

DISCUSSION

CS was first described by Dr. David G Cogan in 1945 as non-syphilitic interstitial keratitis with vestibuloauditory symptoms (4). CS as a rare entity, with fewer than 250 reported cases in the literature, is mostly described in young adult Caucasian patients of both sexes (1). Although the mechanisms responsible for the eye and inner ear disease in CS are unknown, an autoimmune pathogenesis is suggested by finding autoantibodies to inner ear proteins or epithelial cells (5,6). While interstitial keratitis is the classical form of ocular involvement; iritis or uveitis, scleritis or episcleritis, and conjunctivitis have been also described (1). In absence of interstitial keratitis the term atypical CS is used (1). The most characteristic cardiovascular manifestation reported in association with CS is vasculitis of the aorta leading to aortic aneurysm and/or aortic valve regurgitation (1). Other systemic manifestations include fever, arthralgia (or arthritis), lymphadenopathy, splenomegaly, skin rash, and peripheral or central nervous system involvement (1,7). Review of 222 published cases revealed peripheral and central nervous system affection in 7% of typical CS cases and in 22% of atypical CS with significantly higher neurologic involvement in atypical CS ($p=0.01$) (1). In another review of 60 cases of CS, peripheral neuropathy was found in 10 patients (6%) (7). In a review of 32 patients by Grasland et al., facial palsy was found in only one case (1). CS is very rarely reported in Arabic and Middle Eastern countries. In one study of the characteristics of uveitis referrals to a tertiary eye care center in Iran, CS was detected in one out of 544 cases (8). Our case report has some peculiar criteria as the patient is a young 14-year-old girl of Arabic ethnicity, who got unilateral lower motor neuron facial palsy among the features of CS.

Eye disease usually responds to topical corticosteroids and systemic corticosteroids are reserved for unresponsive cases or those with posterior uveitis (7). In contrast to vestibuloauditory affection, poor long term ophthalmologic outcomes due to Cogan syndrome are unusual (7). Deafness is the most common serious outcome of CS (9). Systemic corticosteroid therapy is warranted as soon as possible after the onset of hearing loss (9,10). An initial dose of 1 mg/kg (0.5-2) per day of prednisone is usually recommended (1). If an improvement is observed (objectively and subjectively), corticosteroids are gradually tapered over 2–6 months (9). However, reductions in the corticosteroid dose are contingent upon stable auditory acuity and vestibular function (11). Certain patients may require long-term corticosteroid therapy because of recurrent hearing loss during attempts to taper the prednisone dose (3). The use of corticosteroid-sparing immunosuppressive therapy should be considered in patients for whom excessive

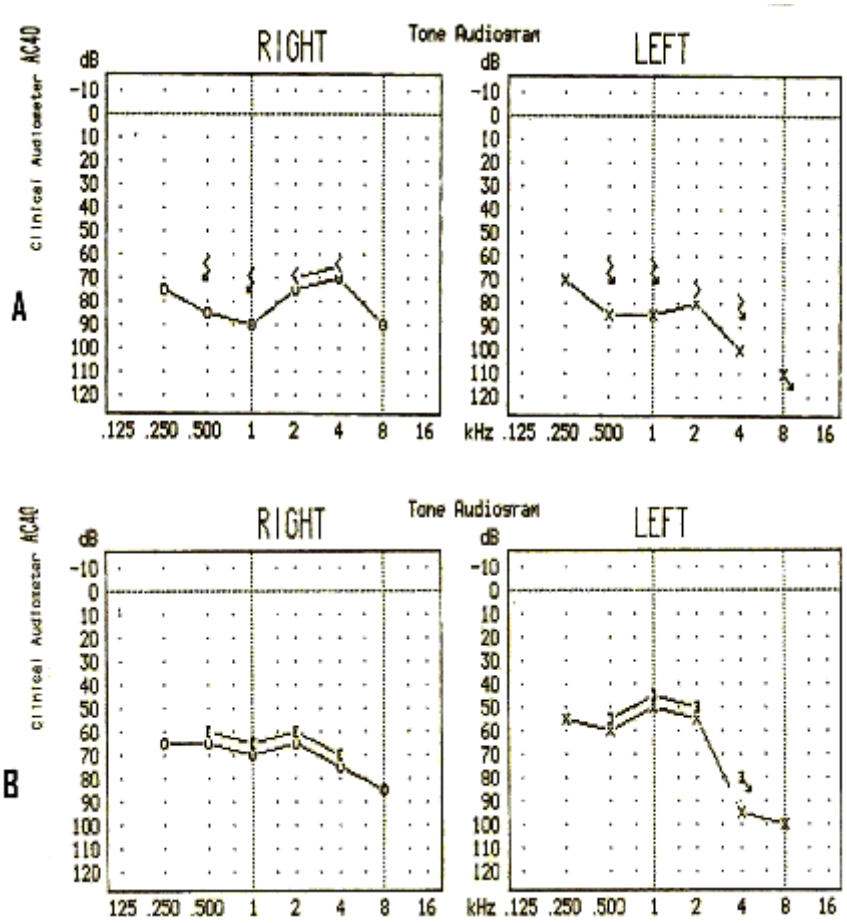

Figure 1. Pure tone audiogram: (A) at time of relapse (B) after treatment with a higher steroid dose

corticosteroid doses are either required to control hearing loss or result in toxicity (3). Azathioprine, cyclophosphamide, cyclosporine, methotrexate, and tumor necrosis factor alpha blockers have been used in this respect (1,12). Some patients fail to respond to such therapy, thereby resulting in severe hearing loss. Cochlear implants have been shown to improve auditory function in such patients (1). Cogan's syndrome is a rare serious disorder that threatens hearing and sight and occasionally may threaten life. It is important to recognize the clinical features of the syndrome since early institution of corticosteroid therapy may prevent complete hearing loss. The decision to taper corticosteroids should be taken carefully and on the basis of sustained improvement in hearing power.

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No conflict of interest is declared.

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