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EFFECTIVE TREATMENT OF AUDIOVESTIBULAR INVOLVEMENT IN COGAN'S SYNDROME WITH BIOLOGICS AND HYPERBARIC OXYGEN THERAPY

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ABSTRACT

Introduction: Cogan's syndrome is a rare autoimmune disease typically affecting the ocular and audiovestibular system. Inner ear involvement is associated with a poor prognosis, as despite therapy, over 50% of patients develop profound deafness. The standard treatment remains the use of high dose glucocorticosteroids and immunosuppressants. Studies from recent years have reported significantly higher rates of improvement and recovery among patients treated with biologic disease modifying antirheumatic drugs (bDMARDs), particularly infliximab.

Case description: We report the case of a 39-year-old Caucasian female who experienced bilateral interstitial keratitis, sudden hearing loss, labyrinthine symptoms and arthralgia. Based on the clinical picture, Cogan's syndrome was diagnosed. Initial treatment included high doses of glucocorticosteroids which did not result in a satisfactory clinical response. Due to the high risk of permanent deafness, it was decided to administer infliximab. Simultaneously, subcutaneous methotrexate was introduced, and the patient was referred for hyperbaric oxygen therapy. Follow-up hearing tests conducted four weeks later revealed a significant improvement. As a result of the treatment, hearing normalized and labyrinthine and ophthalmological as well as systemic symptoms disappeared.

Conclusion: Considering the unfavourable prognosis in Cogan's syndrome with vestibulocochlear involvement, it seems reasonable to quickly escalate treatment if there is no sufficient response to glucocorticosteroids within two weeks. Infliximab has the most documented effectiveness among DMARDs. Combining pharmacotherapy with HBOT can significantly improves treatment outcomes.

KEYWORDS

Cogan's syndrome, hearing loss, infliximab, hyperbaric oxygen therapy (HBOT)

LEARNING POINTS

- *Improved prognosis:* This case highlights a potential therapeutic strategy that may improve outcomes in patients with Cogan's syndrome and reduce the risk of disability.
- *Disease reminder:* Knowledge of this rare disease is essential so that the internist, in cooperation with the otolaryngologist and ophthalmologist, can quickly make the appropriate diagnosis and implement intensive treatment.

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INTRODUCTION

Cogan's syndrome is a rare autoimmune vasculitis that predominantly affects young Caucasian adults. Typically, the disease is characterized by non-syphilitic interstitial keratitis and audiovestibular symptoms such as sensorineural hearing loss, tinnitus, dizziness and acute episodes of vertigo[1]. Interstitial keratitis, manifests with ocular pain, redness, photophobia and blurred vision, usually occurs in the initial stages of the disease, progresses rapidly and then resolves, commonly with corneal scarring[2]. Ocular manifestations of Cogan's syndrome tend to respond well to steroid therapy alone (84% of cases according to Arnaud et al.[3]) and disease modifying antirheumatic drugs (DMARDs). The management of cochleovestibular involvement remains the therapeutic challenge, as progressive sensory hearing loss causes deafness in 1-3 months[2]. Standard first-line treatment involves the use of high-dose glucocorticosteroids[4]. In the steroid-resistant or steroid-dependent patients, treatment with conventional immunosuppressants is attempted. However, their efficacy is not satisfactory and in the 43-52% of cases deafness develops due to fibrosis and osteogenesis in the organ of Corti[5]. Recent case reports/series have shown promising results with the use of biologic DMARDs, especially infliximab[6]. Due to the rarity of Cogan's syndrome and limited studies, there is currently no consensus on the therapeutic management of audiovestibular involvement. The aim of this article is to present a case of steroid-resistant patient whose second-line treatment resulted in the restoration of normal hearing and disappearance of systemic symptoms and labyrinthine disorders.

CASE DESCRIPTION

A 39-year-old Caucasian female was admitted to our clinic due to hearing and vision disorders following sinusitis. The first symptoms appeared in the form of left eye pain, redness, photophobia and periodic vision disturbances, as well as episodes of vertigo. Two weeks later, the above-mentioned symptoms were accompanied by hearing loss in the left ear and tinnitus. An audiogram performed on April 13, 2023 revealed only a slight hearing loss in the high-frequency range in the left ear (up to 40 dB at 8000 Hz). On April 14, the audiogram showed a gradual bilateral deterioration of hearing *(Fig. 1)*, therefore prednisone at a dose of 60 mg per day was prescribed. Follow-up examinations conducted on April 20 revealed a mixed hearing loss in the left ear up to 110 dB with a reserve of approximately 25 dB and a sensorineural hearing loss in the right ear of 50 dB *(Fig. 2)*. On the same day, due to progressive hearing loss, she was admitted to the Department of Otolaryngology. Diagnostic work included a computed tomography (CT) scan of the head (which showed no significant abnormalities) and a slit-lamp ophthalmological examination (that revealed corneal epithelial edema and corneal haze in both eyes based on which interstitial keratitis was diagnosed). The patient received a 4-day course of intravenous dexamethasone 16 mg/day and was discharged home with the recommendation to use the same dose of oral dexamethasone and topical steroid to both eyes.

At the time of admission to our clinic on April 25, the patient reported persistent ophthalmological and cochlea-vestibular symptoms, as well as fatigue, arthralgia and abdominal pain. The following abnormalities were found during physical examination: steroid acne, tenderness on palpation in the epigastrium. Ophthalmological examination revealed corneal haze in both eyes. Syphilis, toxoplasmosis, Lyme disease and tuberculosis were excluded. Laboratory tests showed the absence of antinuclear and anti-

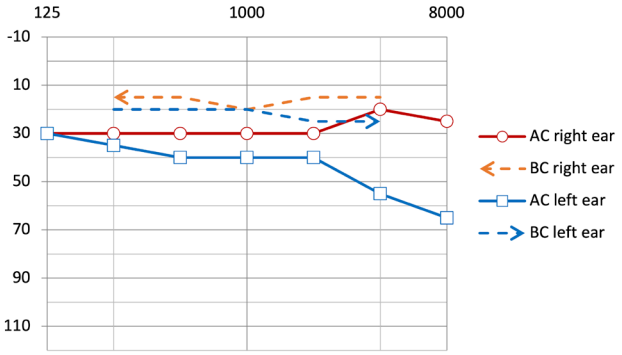


Figure 1. Audiogram performed on April 14, 2023. Abbreviations: AC, air conduction; BC, bone conduction.

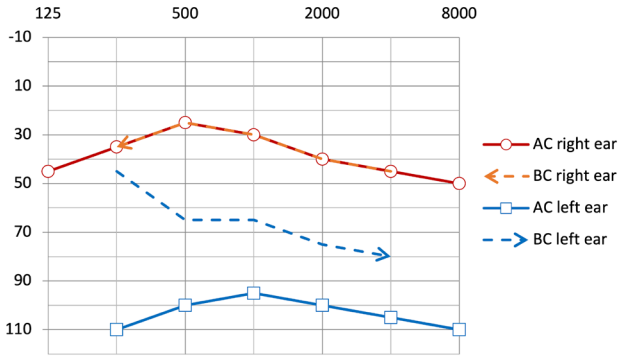


Figure 2. Audiogram performed on April 20, 2023. Abbreviations: AC, air conduction; BC, bone conduction.

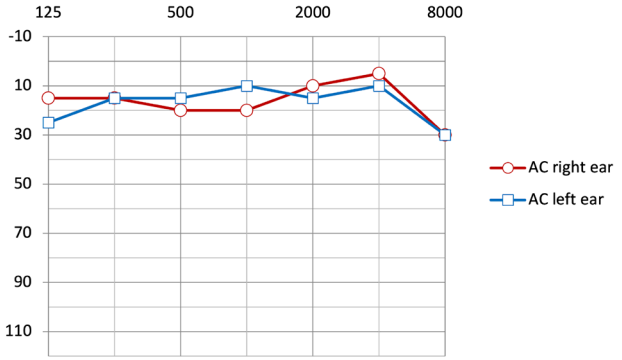


Figure 3. Audiogram performed on May 18, 2023. Abbreviations: AC, air conduction.

neutrophil cytoplasmic antibodies. No signs of large vessel inflammation were found in the CT angiogram of the aorta. Based on the overall clinical picture, we made the diagnosis of Cogan's syndrome. The treatment was changed from dexamethasone to prednisone 60 mg/day. Despite a total of 2 weeks of high-dose corticosteroid therapy, we did not notice any significant improvement. Due to the suspicion of steroid resistance, progressive hearing loss and high risk of irreversible deafness, after reviewing the available studies, we decided to administer infliximab at a dose of 5 mg/kg body weight. The patient received the first dose on April 28, 2023. We planned the next infusions in a 0-2-5 week cycle and then every 8 weeks. Simultaneously, methotrexate was administered as a subcutaneous injection at an initial dose of 20 mg per week, increased after 7 days to 25 mg per week. Additionally, owing to sudden sensorineural hearing loss, the patient qualified for hyperbaric oxygen therapy (HBOT). She underwent a total of twenty 60-minute sessions at a pressure of 2.5 ATA. The follow-up hearing test on May 18 showed significant improvement (curves for both ears in the range of 0-30 dB; *Fig. 3*). Further treatment included prednisone in gradually reduced doses (finally discontinued in September 2024), infliximab (a total of 10 infusions, the last in August 2024) and subcutaneous methotrexate at a dose of 25 mg/week. In May 2024, due to a considerable increase in alanine aminotransferase activity (ALT 8x uln), after excluding active viral hepatitis, methotrexate was discontinued, and infliximab was suspended for a month. Despite this, in September 2024 a re-increase in aminotransferase activity was observed, therefore it was decided to discontinue infliximab and introduce azathioprine at a dose of 1.5 mg/kg body weight as part of therapy to maintain remission. Check-ups performed after these modifications showed a significant improvement in liver enzyme levels. From September 2024 to the present, the patient has been treated with azathioprine 1.5 mg/kg body weight. There was never a recurrence of the symptoms during the therapy.

In the follow-up after 35 months, the patient assessed her health condition as very good and did not report any complaints. As a result of the treatment described above, she experienced restoration of normal hearing and her labyrinthine disorders, ocular symptoms, gastrointestinal disorders and arthralgia disappeared.

DISCUSSION

Available studies regarding Cogan's syndrome emphasize the unfavourable prognosis in the case of audiovestibular system involvement with a high risk of permanent profound deafness (over 50% according to Gluth et al.[7]). This justifies the need to implement rapid and intensive therapy. Systemic corticosteroids in high doses (1-2 mg/kg body weight/day) remain a standard of care. Intratympanic steroid injections allow for a higher concentration of steroid in perilymph with no systemic side effects, that is why they can be offered as adjunctive therapy[8]. If there is no symptomatic improvement within 2 weeks of initiating management with corticosteroids, patient can be considered steroid resistant[9]. A factor correlated with more frequent resistance to corticosteroids is the presence of dizziness, which is most likely due to more severe inner ear involvement in patients with concomitant vestibular symptoms[10]. In these patients, earlier initiation of DMARD therapy might be beneficial.

The standard treatment in the case of steroid-resistant, steroid-dependent and recurrent forms of the disease is the inclusion of conventional immunosuppressants, such as methotrexate, azathioprine or cyclophosphamide. However, on average only 50% of patients on conventional DMARDs achieve a vestibulocochlear response[11]. The frequency of this response is higher in the steroid responsive group (62.1% vs 45% in the steroid resistant group according to Marrero-Gonzalez et al.) and in patients treated with cyclophosphamide (60% with a mean improvement of -16.2 dB). In the last 20 years, bDMARDs have been increasingly introduced, mainly as third-line treatment. The most promising results come from studies on infliximab administration, showing an over 80% vestibulocochlear response at 6 months[12]. Stabilization or improvement in ocular and systemic symptoms was also noted during its use. Infliximab was most often administered at a dose of 3 mg/kg body weight, at intervals similar to those in other autoimmune diseases[6,11]. The appropriate duration of infliximab treatment has not yet been thoroughly studied but it appears to be more effective when started early in the disease. Data on other biologic therapies is more limited and controversial. Single case reports of adalimumab use have shown its ineffectiveness in terms of vestibulocochlear disorders. Current studies regarding tocilizumab indicate its potential use for treating symptoms of hearing loss however the number of case reports is extremely limited. HBOT is another treatment that may be used to improve outcomes in patients with sensorineural hearing loss. Particularly beneficial effects are observed when HBOT is performed within 2 weeks of symptoms onset and in patients with moderate to severe hearing loss[13]. In cases with severe sensorineural hearing loss unresponsive to intensive treatment, cochlear implantation remains a valuable strategy[14]. However, surgical treatment can only be implemented after the inflammation is under control.

In the case of our patient, the first-line therapy with high-dose glucocorticosteroids for 2 weeks proved ineffective. Due to progressive hearing loss, we decided to implement three treatment methods simultaneously: bDMARD (infliximab – in a dose increased to 5 mg/kg body weight due to a high risk of permanent deafness), conventional DMARD (methotrexate) and HBOT. The choice of infliximab was determined by available research results indicating its greatest effectiveness in the management of vestibulocochlear disorders. Methotrexate was included in the treatment regimen to reduce the formation of antibodies against infliximab and improve long-term

outcomes. Due to its proven effectiveness in sudden sensorineural hearing loss and good safety profile, the patient was also scheduled for HBOT. As a result of this combined therapy, hearing normalized and labyrinthine and vision disorders resolved. After a year of treatment, owing to a significant increase in aminotransferase activity, we had to discontinue methotrexate and then infliximab. A major challenge was to decide on the patient's further therapeutic approach, as complete discontinuation of treatment could result in disease relapse, while continuation of glucocorticosteroids could involve the development of numerous side effects. Ultimately, taking into account the patient's reproductive plans, it was decided to introduce azathioprine, which has been continued to this day. Due to the lack of studies regarding the length of maintenance therapy in Cogan's syndrome, it seems appropriate to follow the recommendations for the treatment of other vasculitides.

In summary, owing to the unfavourable prognosis in the case of vestibulocochlear system involvement in steroid-resistant Cogan's syndrome, the implementation of bDMARDs (especially infliximab) together with conventional immunosuppressants in second-line treatment is worth considering. Combining pharmacotherapy with HBOT may promote better final results. The key to success is knowledge of this rare disease, and therefore rapid diagnosis and implementation of intensive treatment.

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