Sudden sensorineural hearing loss in atypical Cogan's syndrome: A case report

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ABSTRACT

The Cogan's syndrome (CS) is a very uncommon inflammatory condition that appears in young adults without a gender predisposition. It can be presented as typical, with interstitial non-syphilitic keratitis and Ménière-like audiovestibular manifestations. An atypical form of CS involves uveitis, scleritis, episcleritis, and systemic vasculitis symptoms.

This is a case of a 41-year old male who reported eye redness, photophobia, and gait instability. His condition progressed to a sudden sensorineural hearing loss, tinnitus, and constant vertigo accompanied by cephalgia. He was evaluated by the Otolaryngology, Ophthalmology, and Neurology departments giving rise to CS, Ménière's syndrome, acoustic neuroma, glaucoma, multiple sclerosis, and meningioma as differential diagnoses. The patient was prescribed with oral and intravenous steroids, ophthalmic antibiotics and steroids, as well as oral omeprazole. The patient was discharged, without his eye manifestations, and for his clinical progress and underwent conventional and speech audiometry, otoacoustic emissions, nystagmography, tympanometry, and auditory steady-state response, that showed a vestibular dysfunction and a severe sensorineural hearing loss. His follow-up, six months later resulted with a normal vestibular function and an improvement from severe to mild sensorineural hearing loss.

The Cogan's syndrome is a rare condition which can leave the patient with permanent incapacitating sequelae including profound hearing loss. Effective treatment is necessary to avoid complications and improve the patient's condition and life quality.

1. Introduction

Cogan's syndrome (CS) is an inflammatory condition without a very clear origin. It was first described in 1945 by Dr. David Cogan, who identified concomitant ocular and audiovestibular symptoms in patients without evidence of syphilitic symptoms [1–3].

Clinically, this syndrome occurs in two different clinical presentations. The typical form of CS comprises three main characteristics. Ocular symptoms, mainly interstitial non-syphilitic keratitis; audiovestibular symptoms similar to the clinical manifestations of Ménière's syndrome, sudden presentation of tinnitus and vertigo accompanied by sensorineural hearing loss; and an interval of less than two years between ocular and audiovestibular symptoms [3,4].

There is evidence that CS is associated with symptoms such as conjunctivitis, scleritis and episcleritis. In addition, it may present with a systemic involvement with aortic vasculitis, although it could occur in any vessel, and along with Crohn's disease [5,6].

There are about 300 cases published in the medical literature describing this syndrome. The most serious sequel due to the lack of specific treatment is a definitive hearing loss, which affects the quality of life of the patient to a great extent. As a physician, it is vital to consider this condition as a differential diagnosis to avoid this type of complications.

This paper describes the case of a patient who presented ocular and audiovestibular symptoms at a community practice medical center in northeastern Mexico and review relevant literature.

2. Case presentation

A 41-year-old male patient born in northeastern Mexico who works in a mechanical diesel workshop, a daily smoking habit and without relevant family history. He was referred to the Otolaryngology...
Department due to sudden bilateral hearing loss.

The patient starts its condition three weeks prior to his visit with red eyes, photophobia, and gait instability. Three days after its debut he went to a medical center that prescribed him otical prednisolone without improvement. Eight days later, the patient suffered bilateral sudden hearing loss, postural vertigo that progressed to constant vertigo, tinnitus, headache, and gait instability, and three days after, he went to a first level clinic where he was administered acyclovir (750mg/8 hours) and referred to our clinic.

In our medical center, the patient was evaluated by Ophthalmology, Neurology, and Otolaryngology of the clinic. The physical exploration revealed anterior uveitis, episcleritis, vertigo, hearing loss, and gait instability. The differential diagnoses were: sudden sensorineural hearing loss, glaucoma, multiple sclerosis, acoustic neura, meningioma, Ménière’s syndrome, and CS.

Several audiologic studies were performed, a normal A type curve in the tympanogram, transient evoked otoacoustic emissions at 75 dB that evidenced a “no pass” result, nystagmography indicating a vestibular dysfunction with abnormal sacadic eye movement test, altered optokinetic response, and bilateral hyporeflexia in the caloric test. The patient also underwent a complete pure-tone audiometry showing a moderate-severe bilateral sensorineural hearing loss with an average of 64 dB in all frequencies. An auditory steady-state response was performed, and its results confirmed the 64 dB sensorineural hearing loss as shown in Fig. 1.

Laboratory studies were performed showing an abnormal creatinine clearance level of 50.93 mL/min, 10.4 k/ul leukocytes, 1.12 k/ul monocytes, mean corpusular hemoglobin at 31.6 pg, red cell distribution with of 15.4%, lactate dehydrogenase at 245U/L, erythrocyt sedimentation rate at 39 mm/h, a positive c-reactive protein and IgG for Rubella. The rest of the laboratory studies, including HIV, hepatitis B and C, rheumatoid factor, Venerale Disease Research Laboratory (VDRL), lupus erythematosus cells, anti SSA/anti Ro and SSB/anti La, anti-nuclear antibodies, anticytoplasmic antibodies, and anti-neutrophil cytoplasmic antibodies, showed negative results.

Initial treatment with acyclovir was suspended and the patient was administered oral prednisolone (75mg/day) and omeprazole (40mg/day); ophthalmic prednisolone and tobramycin; and intravenous dexamethasone (8mg/6 hours). The medication was tolerated without any complaints.

During the first week after admission, the patient refers the disappearance of his right ear tinnitus and bilateral hearing loss, and improvement on his left ear tinnitus. The eye redness and cephalae resolved completely.

The patient continued his clinical improvement for two more days, and neither tinnitus, vertigo, nor subjective hearing loss were present. The lab results showed a normal erythrocyt sedimentation rate at 2 mm/h, normal urinalysis, and complete blood count showed 13900 leucocytes per ul.

The patient was discharged due to the clinical progress and prescribed oral prednisone (75mg/day) and omeprazole (40mg/day) for one more week. The steroid therapy was tapered slowly for two months to avoid corticosteroid-induced toxicity.

The patient underwent control audiological studies six months later. Pure tone audiometry and auditory steady-state response showed significant improvement from an average of 64 to 35 dB of sensorineural hearing loss as shown in Fig. 2. The tympanogram displayed a normal A type curve with inner ear pressure and volume in the normal range, transient evoked otoacoustic emissions reported normality, nystagmography adequate vestibular function with normal saccadic eye movement test, appropriate optokinetic response, and bilateral normoreflexia in the caloric test.

3. Discussion

The patient of this case is considered a young adult, according to his age, coinciding with the characteristics described in the literature. We found evidence in three large series of cases that this condition occurs in this age group with averages between 28.6 and 38 years of age. In addition, these series conducted by Duke University, the Mayo Clinic and the French National Society of Internal Medicine found a male-female relationship of 16/31, 27/33 and 19/13, respectively. These series support the claim that it occurs in young adults and that there is no predilection for gender [7–9].

The manifestations in the CS are usually initially ocular and later the audiovestibular system becomes affected. The ocular symptoms in the typical form of the disease are non-syphilitic interstitial keratitis, red eye, pain and photophobia, diplopia and foreign body sensation. The audiovestibular symptoms are in most cases severe, disabling and generally bilateral. They consist of sensorineural hearing loss, tinnitus, vertigo that can lead to vomiting, nystagmus, and ataxia [9,10].

There are ocular symptoms that are associated with non-syphilitic interstitial keratitis, giving rise to the concept of atypical CS. Some of them are anterior or posterior uveitis, scleritis, episcleritis, corneal ulceration, peripheral ulcerative keratitis, conjunctival or subconjunctival hemorrhage, exophthalmos, papillary edema, oculumotor paralysis, among others [11,12].

In addition to the ocular and audiovestibular symptoms, the atypical CS presents systemic manifestations such as fever, asthenia and adynamia. About 30% of patients report rheumatological symptoms such as myalgias, arthralgias and sometimes arthritis. There are patients who manifest symptoms of medium and large vessel vasculitis, especially in

Fig. 1. Pure-tone audiometry and auditory steady-state response before treatment.
PTA: Pure-tone audiometry
ASSR: Auditory steady-state response.
the aorta. There have been cases in which the digestive tract is affected with hemorrhages and diarrhea due to mesenteric arteritis [12–14].

At the time of the work, no evidence was found in the literature that Cogan’s syndrome has a family predisposition.

The diagnosis of this condition is mainly clinical. Laboratory studies that are used to diagnose autoimmune diseases such as antinuclear antibodies and anti–citrullinated protein antibody are not present in CS. However, there is evidence of the relationship between the typical CS and some markers that continue to be studied. The anti Hsp-70 antibody has a high sensitivity to diagnose the typical syndrome. In such a way that if a negative result is obtained (before starting immunosuppressive treatment), the diagnosis of typical CS can be excluded [15,16].

There is no evidence of guidelines or criteria for treatment, which is why it is based on case reports and series of cases where there is improvement of symptoms and decrease in complications. The treatment is directly related to the severity of the symptoms, from topical steroids for mild ocular inflammation, to systemic immunosuppressants to counteract sensorineural hearing loss, severe ocular involvement and systemic symptoms [17].

The literature reports that it should be started with 1mg/kg/day of prednisone for 2–4 weeks or until the patient improves. If improvement is not achieved, the dose of the steroid should be increased for 2 to 3 more weeks. A failure in treatment indicates the need to administer an additional immunosuppressive treatment such as methotrexate was used at a dose of 15–25 mg/week, azathioprine at 1.5–2.5 mg/kg/day, cyclophosphamide at 2–3 mg/kg/day, and cyclosporine A at 5 mg/kg/day [17].

There is evidence that the tumor necrosis factor (TNF) alpha is very relevant in the pathophysiology of the disease. It is a cytokine that occurs in situations of infection and inflammation by activated macrophages. This induces the increase of acute phase reactants such as C-reactive protein. Infliximab is a monoclonal chimeric anti-TNF alpha antibody and has been shown to improve the symptoms of patients with systemic vasculitis such as Takayasu’s disease. Likewise, in a French series with 62 patients with CS they found that patients with refractory audiovestibular symptoms resolved in 80% at 6 months when they were treated with infliximab [17,18].

4. Conclusion

The research team suggests the physicians to consider the CS as a differential diagnosis for audiovestibular and ocular symptoms. A profound sensorineural hearing loss is the most frequent and devastating complication. This rare condition affects the quality of life of the patients if not treated effectively. However, if treated correctly, the patients improve satisfactorily.

Ethical approval

The case report was approved by the local research and ethical committee.

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Author contribution

Jose Luis Treviño-Gonzalez, research team coordinator, data collection and analysis, final approval.

German A. Soto-Galindo: data collection and analysis, literature research, paper writing and corrections.

Rafael Moreno-Sales: data collection and analysis, final approval.

Josefina A. Morales Del Angel: data collection and analysis, literature research.

Conflicts of interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Jose Luis Treviño Gonzalez MD PhD.

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