Duazary / Vol. 20, No. 2 – 2023 / 152 - 158
DOI: https://doi.org/10.21676/2389783X.5374

Presentation of clinical cases

Sudden hearing loss as initial manifestation in patients with systemic autoimmune rheumatic diseases: Case series

Hipoacusia súbita como manifestación inicial en pacientes con enfermedades reumáticas autoinmunes sistémicas: serie de casos

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How to cite this article: Patiño-Arboleda M, Rueda-Gutiérrez J, Ocampo-Chaparro JM, Arbeláez-Cortés A. Hipoacusia súbita como manifestación inicial en pacientes con enfermedades reumáticas autoinmunes sistémicas: serie de casos. Duazary. 2023; 20 (2): 152-158. https://doi.org/10.21676/2389783X.5374

Received on July 29, 2022
Accepted on June 22, 2023
Posted online en June 30, 2023

ABSTRACT

Sudden hearing loss is an acute-onset entity secondary to multiple etiologies, including rheumatologic diseases. Typically, its involvement is unilateral associated with a sensation of ear plugging and tinnitus. The diagnosis is based on the history, physical examination, and audiometry. The first-line treatment is corticosteroids associated or not with other therapeutic agents according to their etiology. The prognosis depends on the degree of initial hearing loss, time of initiation of treatment, and factors inherent to the patient. We presented three clinical cases of sudden hearing loss as the initial manifestation of rheumatological disease.

Keywords: Sudden hearing loss; Rheumatic diseases; Autoimmune disease; Steroids; Ear.

RESUMEN

La hipoacusia súbita es una entidad de instauración aguda secundaria a múltiples etiologías, como las enfermedades reumáticas autoinmunes sistémicas (ERAS). Típicamente su afectación es unilateral, asociada con taponamiento ótico y acúfenos. El diagnóstico se basa en la anamnesis, exploración física y audiometría. El tratamiento de primera línea son los corticoesteroides sistémicos, junto con otros agentes terapéuticos según su etiología. El pronóstico depende del grado de hipoacusia, del tiempo de instauración del tratamiento y de factores inherentes al paciente. A continuación, se presentan tres casos clínicos de pacientes que debutaron con hipoacusia súbita como manifestación inicial de una ERAS.

Palabras clave: pérdida auditiva súbita; enfermedades reumáticas; enfermedades autoinmunes; corticoesteroides; oído

INTRODUCTION

Sudden hearing loss is defined as a sensorineural hearing loss more significant than 30 decibels (dB), at least in three or more consecutive frequencies of the tone audiometry, with an onset time of less than 72 hours[1]. It was first described in 1944[2,] and its incidence is increasing. It can present at any age, being more frequent between 43 and 53 years, without a predominance of sex[3]. Within the etiopathogenesis, three theories have been postulated[1]: the first and most studied is infectious, especially by viral agents; the second, due to alteration of the microcirculation of the inner ear; and the third refers to the association with immune-mediated diseases. The latter is supported by the response to immunosuppressive treatment with corticosteroids and the finding of specific antigens in the inner ear, such as type II collagen, β-tubulin, and heat shock protein 70, among others, whose damage may be mediated by autoantibodies, immune complexes or autoreactive lymphocytes; however, its etiopathogenesis and pathophysiology are still not entirely clear[4].

Sudden sensorineural hearing loss associated with systemic autoimmune rheumatic diseases (SARD) corresponds to less than 1% of all cases. On some occasions, it may be the first clinical manifestation of these[5], with a frequency of diverse appearance both in its sudden and non-sudden form of presentation (Table 1). The usual clinical presentation is usually unilateral involvement with or without tinnitus and vertigo, associated or not with specific characteristics of each SARD.

Table 1. Frequency of hearing loss in SARD.

SARD	**Frequency (%)**
Systemic sclerosis (Diffuse and limited)[6]	20-77
Rheumatoid arthritis[7]	25-72
Sjogren's syndrome[8]	46
Systemic lupus erythematosus[9]	6-70
Granulomatosis with polyangiitis[10]	8-63
Antiphospholipid síndrome	5-20

The diagnosis is based on an adequate anamnesis, a complete physical examination, acumetry through the use of tuning forks to differentiate between sensorineural or transmission hearing loss, and, finally, tonal audiometry for the objective evaluation of the functioning of the auditory system[12]. The first line of treatment is corticosteroids, associated with additional therapies that vary according to the suspicion or etiological confirmation of a SARD[13].

Below are three clinical cases of patients who debuted with sudden hearing loss as the initial manifestation of SARD.

CLINICAL CASES

1. A 50-year-old woman was admitted to the rheumatology clinic due to a clinical picture that began four months earlier, consisting of sudden hearing loss in the right ear associated with tinnitus. As a pathological history, he referred to hypothyroidism, and, in the review of symptoms by systems, he presented a history of a subjective dry eye of several months' evolution. In the previous evaluation by otorhinolaryngology, the audiometry documented right sensorineural hearing loss, and treatment was started with prednisolone 1 mg/kg/day, along with oral acyclovir, with improvement in his clinical picture. However, one month later, she presented a new episode of severe contralateral sensorineural hearing loss, which required intratympanic infiltration of corticosteroids, achieving partial improvement. Complementary tests revealed antinuclear antibodies (ANA) with a cytoplasmic pattern at titres of 1:320 and positive anti-SSA/Ro at 71.1 U/ml (<15 U/ml). Schirmer's test was positive for dry eye: 4 mm in the right eye (RE) and 5 mm in the left eye (LE). It was classified as Sjögren's syndrome, associated with sudden hearing loss. Due to the persistence of severe left sensorineural hearing loss, he was hospitalized to start pulsing methylprednisolone at a dose of 500 mg/day for three days, followed by oral prednisolone with a gradual reduction to 5 mg/day, hydroxychloroquine 200 mg/day, and azathioprine 50 mg/day.

2. A 69-year-old woman admitted to the rheumatology clinic was referred by an otorhinolaryngologist for an etiological study of sudden hearing loss. She referred a clinical picture that began seven months earlier, consisting of sudden hearing loss in the left ear, associated with a sensation of ear plugging and vertigo. Within the pathological history, he reported hypothyroidism and, upon review of symptoms by systems, subjective sensation of dry mouth for several months.

She provided audiometry that showed severe left sensorineural hearing loss and magnetic resonance imaging (MRI) of the brain that showed enhancement of the left labyrinth with the contrast medium. Within the extension studies, the Schirmer test was positive for dry eye (OD: 8 mm, LE: 4 mm), and the minor salivary gland biopsy was compatible with Sjögren's syndrome (focus score: 1.14). Treatment was started with prednisolone at 1 mg/kg/day and hydrochlorothiazide, acyclovir, and hydroxychloroquine 200 mg/day as an immunomodulator.

3. A 63-year-old woman with a clinical picture that began four months ago, consisting of tinnitus and right otalgia, for which she received oral antibiotic management without improvement. Fifteen days before the consultation, she presented sudden bilateral hearing loss, more remarkable in the right ear, associated with holocranial headache and vertigo. Within the pathological history, she referred arterial hypertension and commented on several months of dry cough and runny nose. He initially consulted the otorhinolaryngologist, finding bilateral sensorineural hearing loss on audiometry, so prednisolone was started at 1 mg/kg/day, and he was referred to rheumatology for etiological studies. Complementary tests were C-reactive protein (CRP) at 108 mg/L (<5 mg/L), erythrocyte sedimentation rate (ESR) 64 mm/h, ANA homogeneous pattern at titres of 1:320; positive anti-MPO (anti-myeloperoxidase) antibodies >100 ng/ml (<5 ng/ml). Chest radiography showed subsegmental atelectasis in the lingula, without pulmonary infiltrates or nodules; paranasal sinus X-rays and brain MRI were normal. A picture consistent with vasculitis associated with ANCA (anti-neutrophil cytoplasmic antibodies) was considered, with positive anti-MPO, possibly granulomatosis with polyangiitis, supported by a history of upper respiratory tract symptoms and hearing impairment due to sudden bilateral hearing loss. He was hospitalized for the initiation of methylprednisolone pulses at 500 mg/day for three days, together with the first cycle of intravenous cyclophosphamide at 750 mg. She was discharged with prednisolone with a gradual reduction to 5 mg/day, and cyclophosphamide was continued until completing six cycles with the same dose. The clinical evolution was favorable, and maintenance therapy was started with azathioprine 50 mg/day.

Below is a summary of the clinical characteristics, paraclinical studies, treatment, evolution, and timeline of occurrence of events (Figure 1 and Table 2).

Declaration on ethical aspects

To carry out and participate in this study, the signing of the informed consent by each of the patients was taken into account; in addition, the procedures followed were by the ethical standards of Resolution 8430 of 1993 of the Ministry of Health and Social Protection. of Colombia and the Declaration of Helsinki.

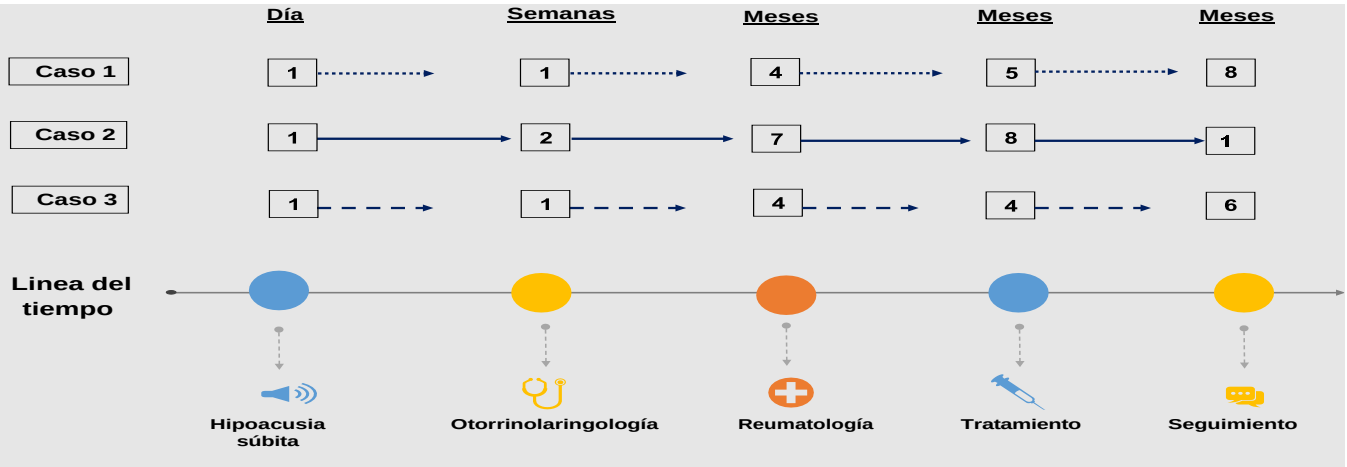

Figure 1. Event occurrence timeline.

Table 2. Description of clinical cases.

CASE	1	2	3
Sex	Women	Women	Women
Age	50 years	69 years	63 years
Origin	Cali, Colombia	Cali, Colombia	Cali, Colombia
Background	hypothyroidism	Hypothyroidism	Arterial hypertension
Symptoms	Bilateral sudden hearing loss otic tamponade Dry Eye	Sudden left hearing loss otic tamponade Dry mouth Vertigo	Bilateral sudden hearing loss otic tamponade headache Vertigo runny nose Dry cough
Tone audiometry	Bilateral sensorineural hearing loss: RE 40 dB, LE: 56 dB	Left sensorineural hearing loss: RE 20 dB, RO:60 dB	Bilateral sensorineural hearing loss: RE 81 dB, LE: 65 dB
Speech audiometry	100% Discrimination: DO at 50dB RO at 80 dB	100% Discrimination: DO at 30dB RO at 80 dB	100% Discrimination: DO at 100dB RO at 80 dB
Paraclinical studies	• Positive Schirmer test • ANA cytoplasmic pattern • Anti-SS-A/Ro positive	• Positive Schirmer test • Salivary gland biopsy: focus score 1.14	• ANA homogeneous pattern • anti-MPO positive • anti-ena negative • anti-DNA negative
Diagnosis	Sjogren's syndrome	Sjogren's syndrome	ANCA positive vasculitis
Treatment	• intravenous methylprednisolone • oral prednisolone • Acyclovir • hydroxychloroquine • Azathioprine	• oral prednisolone • Acyclovir • hydroxychloroquine • hydrochlorothiazide	• intravenous methylprednisolone • oral prednisolone • Cyclophosphamide • Azathioprine
Last evaluation	hearing improvement tinnitus onset	Hearing improvement and dry symptoms	hearing improvement

Abbreviations: ANA: antinuclear antibodies; anti-MPO: anti-myeloperoxidase antibodies; dB= decibel; RE: right ear; LE: left ear.

DISCUSSION

Three clinical cases of patients with SARD are presented, two with Sjögren's syndrome and one with ANCA-associated vasculitis, who presented with sudden hearing loss as the initial manifestation of their disease.

Sudden hearing loss is sensorineural hearing loss of less than 72 hours[1]. Among the most frequent causes are infections, trauma, toxins, and metabolic or hematological disorders. However, even though SARD corresponds to less than 1% of the causes[5], it is necessary to consider them within the differential diagnosis. As evidenced in the cases described, timely diagnosis and treatment are associated with improved auditory functioning. However, a shorter time between the onset of symptoms and evaluation by rheumatology would have been ideal.

Within the approach to a patient with sudden hearing loss, it is essential to prepare a detailed clinical history, together with a complete physical examination that includes hearing evaluation through acumetry to evaluate the airway and bone in order to differentiate according to the Weber and Rinne tests between transmission and sensorineural hearing loss[14]. Audiometry is essential for both diagnosis and follow-up. The airway and bone pathway curves typically descend simultaneously in sensorineural hearing loss[15]. The type of hearing loss documented

in the three cases described was sensorineural. This form of involvement occurs more frequently in patients with sudden hearing loss secondary to cochlear involvement of the inner ear in SARD.

Inner ear involvement has been reported in different ERAS. It was first described by Cogan in 1940, with a higher frequency of presentation in women than in men and higher between the fourth and sixth decade of life,[16] something similar with the cases presented.

Interdisciplinary management is essential when a SARD is suspected of causing sudden hearing loss. Sjögren's syndrome is associated with sicca symptoms, granulomatosis, and polyangiitis with upper respiratory symptoms. However, sudden hearing loss is less frequent than its initial form of presentation[17,18]. A study that evaluated the association between hearing loss in patients with Sjögren's syndrome found that of 60 women and three men included, with a mean age of 49 years, the prevalence of hearing loss was 95.2%, with all the neurosensory type cases[19]. On the other hand, a study that described the frequency, type, and course of hearing loss in granulomatosis with polyangiitis found that of 36 patients, mostly men, 56% had hearing loss, 47% being sensorineural, with an appearance both at the beginning of the disease and during a relapse[10]. The cases described demonstrate how the auditory manifestations represent an entity that must be sought and identified early within the group of SARD.

The role of autoimmunity in the pathogenesis of sudden hearing loss has been studied for some time[20]. So far, six antigens are known in the inner ear, capable of being recognized by the immune system. The mechanism of ear damage may be secondary to direct recognition of the antigen by an autoantibody, by deposition of immune complexes, or by antigenic presentation to an autoreactive T lymphocyte-associated with the release of cytokines and other inflammatory molecules[21]. The pathophysiological origin of the three cases mentioned may be secondary to one or more of these mechanisms.

First-line treatment is the use of corticosteroids administered systemically, intratympanically, or combined[4] with specific therapeutic agents depending on the causative SARD. This has been related to better hearing function and control of the underlying disease[11,22]. The three cases described had a favorable clinical course after starting corticosteroids and additional immunosuppressive therapy in two of them.

This series of cases demonstrates the importance of carrying out a comprehensive approach in the study of patients with sudden hearing loss, with particular emphasis on identifying the individual profile through clinical history, complete physical examination, and correct interpretation of paraclinical studies, which allows determining an etiological entity, such as that of autoimmune origin. Shortening the time between the onset of symptoms, the identification of hearing loss, the evaluation by the relevant specialty, and the start of timely treatment are associated with a better hearing prognosis. However, this ideal approach represents a challenge for health services and professionals.

CONCLUSIONS

Sudden hearing loss is a rare entity with multiple etiological groups, among which are SARD in low prevalence. The interdisciplinary approach to this group of patients allows timely identification through a detailed clinical history, physical examination including auditory function, search for systemic manifestations of some underlying disease, and rational use of complementary tests guided to diagnostic suspicion. of a possible etiological entity and establishment of an effective treatment, thus improving the prognosis and quality of life of the patient.

DECLARATION ON CONFLICTS OF INTEREST

The authors declare that does not exist a conflict of interest.

AUTHORS' CONTRIBUTION

First author: Analysis, writing of the manuscript, and final approval.
Second author: Analysis, writing of the manuscript, and final approval.
Third author: Analysis, writing of the manuscript, and final approval.
Fourth author: Analysis, writing of the manuscript, and final approval.

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