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ORIGINAL ARTICLE

Analysis of Audiometric Relapse-Free Survival in Patients With Immune-Mediated Hearing Loss Exclusively Treated With Corticosteroids☆

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KEYWORDS
Autoimmune disease;
Sensorineural hearing loss;
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Abstract
Objective: To describe the results in terms of audiometric relapse-free survival and relapse rate in immunomediated hearing loss patients treated exclusively with corticosteroids.
Method: Retrospective study of patients with audiometric relapses, monitored from 1995 to 2014, in two centres of the Community of Madrid.
Results: We evaluated 31 patients with a mean age of 48.52 years (14.67 SD), of which 61.3% were women. Most hearing loss was fluctuating (48.4%). Only 16.1% of patients had systemic autoimmune disease. There is a moderate positive correlation between the sex variable and the systemic involvement variable (Spearman's correlation coefficient = 0.356): specifically, between being female and systemic disease. The relative incidence rate of relapse in the first year was 2.01 relapses/year with a 95% CI (1.32–2.92). The mean survival time of the event (audiometric relapse) was 5.25 months (SD 0.756). With multivariate analysis, the only variable that achieved statistical significance was age, with a hazard ratio of 1.032 (95% CI; 1.001–1.063, $P = .043$).
Conclusions: Immune-mediated disease of the inner ear is a chronic disease with relapses. Half of the patients with immunomediated hearing loss treated exclusively with corticosteroids

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Abbreviations: dB, decibels; SD, standard deviation; CI, confidence interval; No., number; RT, TRrelative rate of relapse.

relapse before 6 months of follow-up. In addition, if a patient has not relapsed, they are more likely to relapse as each year passes. Analysis of the audiometric relapse-free survival will enable the effect of future treatments to be compared and their capacity to reduce the rhythm of relapses.

PALABRAS CLAVE
Enfermedades autoinmunes;
Hipoacusia neurosensorial;
Recaída;
Análisis de supervivencia

Análisis de supervivencia libre de recaída audiométrica en pacientes con hipoacusia inmunomediada tratados exclusivamente con corticoides

Resumen
Objetivo: Describir los resultados en términos de supervivencia libre de recaída audiométrica y el ritmo de recaída en pacientes con hipoacusia inmunomediada tratados exclusivamente con corticoides.
Método: Estudio retrospectivo de pacientes, con recaídas audiométricas, en seguimiento desde 1995 hasta 2014, en 2 centros de la Comunidad de Madrid.
Resultados: Se evaluaron 31 pacientes con una media de edad de 48,52 años (14,67 DE) de los cuales el 61,3% fueron mujeres. La mayoría de las hipoacusias fueron fluctuantes (48,4%). Solo el 16,1% de los pacientes presentaban enfermedad autoinmune sistémica. Existe una correlación positiva moderada entre ser mujer y presentar afectación sistémica (coeficiente de correlación de Spearman = 0,356). La tasa relativa de incidencia de recaída en el primer año en nuestra serie fue de 2,01 recaídas/año con un IC95% (1,32-2,92). El tiempo de supervivencia medio del evento (recaída audiométrica) fue de 5,25 meses (DE 0,756). Con el análisis multivariante, la única variable que consiguió significación estadística fue la edad, con una hazard ratio de 1,032 (IC95%; 1,001-1,063, p = 0,043)
Conclusiones: La enfermedad inmunomediada del oído interno es una enfermedad crónica con recaídas. La mitad de los pacientes tratados exclusivamente con corticoides recaen antes de los 6 meses de seguimiento. Además, si un paciente no ha presentado recaída, tiene más riesgo de recaer cada año que pasa. El análisis de la supervivencia libre de recaída audiométrica permitirá comparar el efecto de tratamientos futuros y su capacidad para reducir el ritmo de recaídas.

Introduction

Neurosensorial immune-mediated hearing loss is a progressive, bilateral disease with asynchronous presentation, which responds to corticosteroid treatment and which evolves with periods of inactivity and relapse. The course of the disease is different in every patient, sometimes different in each ear. It has been estimated that the incidence of immune-mediated diseases of the inner ear is low, under 1:5000 inhabitants/per year, which discourages studies. It may be associated with systemic diseases in 30% of cases.

The aetiology of immune-mediated hearing loss has not yet been demonstrated, despite efforts to define target antigens in the inner ear and the inflammatory response this unleashes in its attack.[1,2] Its diagnosis continues to be clinical, since no identifying marker exists,[3] and it is based on the response to treatment with oral corticosteroids or intratympanic drugs.

We may describe 3 patient groups, one with progressive bilateral and asymmetrical hearing loss, another with a fluctuating loss of hearing, and a last group, with repeated episodes of sudden deafness, which impair the hearing thresholds. The evolutionary consequence in any of these is major impairment of the hearing thresholds for which there is no appropriate and early treatment.

Responses have to be estimated interpreting the results of the audiometry before and after treatment, due to the fact that the inner ear cannot be visualised by any direct method, as occurs in autoimmune uveitis.[4] There are patients with immune-mediated hearing loss who initially respond to the corticosteroid and then stop doing so.

The use of corticosteroid saving agents when long-term corticosteroid treatments are required is accepted for all autoimmune diseases.

At present, there is no immune suppressant of choice in the long-term treatment of immune-mediated hearing loss,[5] but its potential modulator of the disease may be used to reduce the number of relapses, as occurs in autoimmune uveitis[6] as well as serving as the corticosteroid saving agent. Biological therapies (etanercept, rituximab,