Unilateral sensorineural hearing loss as immune-mediated inner ear disease
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INTRODUCTION

Immune-mediated inner ear disease (IMIED) is a syndrome of bilateral sensorineural hearing loss (SNHL) characterized by rapid progression over days to months (1), that is caused probably by specific or aspecific antibodies or immune cells which are attacking the inner ear (2). Hearing loss involves mostly both ears (80%), with symmetric or asymmetric auditory thresholds (3,4).

IMIED is diagnosed after clinical suspicion and response to corticosteroids (3). Therefore the most rational strategy for a correct diagnosis of IMIED should consider, in addition to the typology of SNHL, differential diagnosis and immunosuppression response, also the results of some immunological laboratory tests, that can be specific (OTTObst *) and aspecific (5,6,7).

The aim of our study was to investigate the positivity of aspecific immunological tests not only in patients suffering from bilateral SNHL, but also in patients suffering from progressive or sudden unilateral SNHL.

RESULTS

72 patients were observed at our Clinic because suffering from suspected IMIED. They were 24 males and 48 females, with a mean age of 46.8 years (min 14, max 72). In total, the aspecific immunological tests resulted positive in 48 patients (66.67%). In Group A (n=26), patients resulted positive for aspecific immunological tests (66.7%) (Figure 1): 80.8% was affected by symmetric SNHL and 19.2% by asymmetric SNHL. 13 patients (33.3%) resulted negative for aspecific immunological tests, 61.5% affected by symmetric SNHL and 38.5% by asymmetric SNHL (Figure 1). In Group A1, 72.4% of patients resulted positive for aspecific immunological tests and 27.6% were negative. In Group A2, 50% resulted positive and 50% negative (Figure 2A).

In Group B, aspecific tests were positive in 66.7% of patients and negative in 33.3% of patients (Figure 1). In Group B, among the 22 patients affected by sudden SNHL (Group B1) 59.1% resulted positive for aspecific immunological tests, and 40.9% resulted negative; among the 11 patients affected by progressive SNHL, the 81.8% resulted positive for aspecific immunological tests, while the 18.2% resulted negative (Figure 2B).

Considering together the bilateral progressive and unilateral progressive SNHL (19+11 patients), a 70% of positivity of immunological tests has been found. ANA resulted positive in 30.7% of Group A and 33.3% of Group B; ENA resulted positive in 12.8% of Group A and 3% of Group B; Anti-smooth-muscle resulted positive in 23.1% of Group A and 27.3% of Group B; Anti-tyroglobulin antibodies resulted positive in 10.2 of Group A and 24.2% of Group B; Anti-thyroid peroxidase (anti-TPO) antibodies resulted positive in 18% of Group A and 24.2% of Group B. In Group B, 17.14% (6) ANA positivity was found in 17.14% of the patients, while ASMA resulted always negative.

In regard to the antibodies titers scores of ANA and ASMA, significant statistical differences have not been assessed between Group A and B, for both parametric and non-parametric tests (Figures 1A and 2A).

Among Group A patients with positivity for aspecific auto-antibodies (n=26), 3 (11.5%) had systemic autoimmune disease associated to thyroid disease, 7 (26.9%) had only systemic autoimmune disease and 3 (11.5%) had only thyroid disease. Among Class A patients with positivity for aspecific auto-antibodies (n=13), nobody had systemic autoimmune disease associated to thyroid disease, 2 (15.4%) had only systemic autoimmune disease and nobody had only thyroid disease.

Among Class B patients with positivity for aspecific auto-antibodies (n=22), 1 (4.5%) had systemic autoimmune disease associated to thyroid disease, 2 (9%) had only systemic autoimmune disease and 7 (31.8%) had only thyroid disease. Among Class B patients with negativity for aspecific auto-antibodies (n=11), nobody had systemic autoimmune disease associated to thyroid disease, 1 (9%) had only systemic autoimmune disease and nobody had only thyroid disease (Figure 4).

As far as the correlation between thyroid pathology and positivity for anti-thyrogbulin and anti-TPO, Group B showed this correlation in 88.9% of patients with high value of autoantibodies, while Group A in 66.7% of patients with inferior values.

DISCUSSION AND CONCLUSIONS

Aspecific tests positivity in suspected IMIED showed to be high (66.67%) in unilateral and bilateral groups. ANA and ASMA titers and distributions resulted similar.

IMIED can be unilateral or bilateral. The detection of serological non specific inner ear autoantibodies may be useful to suggest a non-organ-specific autoimmune disorder, not only in bilateral SNHL but also in unilateral type, especially in the progressive form.

Although there is no consensus about the use of diagnostic tools to correctly identify an IMIED, an immunological evaluation of patients would be helpful in view of a therapeutic approach, eventually based on steroids for primary and recurrent disease.

REFERENCES


MATERIALS AND METHODS

A prospective case series study was performed. Patients with Meniere's disease, retrococlear pathologies or aged over 65 years were excluded. All the patients have undergone pure tone audiometry and the following battery of blood exams to evaluate the immunological response: ANA, ENA screening, anti-thyroperoxidase (anti-TPO), anti-thyroglobulin and antibody against smooth muscle (ASMA).

Clinically, the patients were divided into two groups: the first one, Group A, with bilateral hearing loss and the other one (Group B), with unilateral hearing loss. The incidence of aspecific antibody positivity was evaluated globally, in individual groups and also in the different hearing loss evolution. A scale has been assigned to the autoantibodies with higher positivity, ANA and ASMA: 1 for slight positivity, 2 for mild positivity and 3 for high positivity (Fig.1B). A statistical two pair sample test (t test) has been applied for the evaluation of statistical difference between ANA and ASMA positivity scores of the two Groups A and B.

The results were compared with a control group, Group C, composed of 35 normal subjects. The incidence of systemic autoimmune disease has been evaluated in Group A and Group B: the correlation between systemic autoimmune disease and aspecific autoantibody positivity or negativity was analyzed.