THE HEARING SYSTEM PARTICIPATION IN AUTOIMMUNE DISEASES

First, we must emphasize the difficulty found for a complete survey on this issue, as sources available refer to physical and pathological explanations rather than focusing on clinical findings.[1]

It would be interesting to analyze this topic under two approaches: The first is when hearing symptoms appear before consolidation of diagnosis of autoimmune disease and will be the base for its confirmation. A clear example is the characterization of a unilateral acute chondritis. The second is that, when the diagnosis of the main disease is clear, involvement of the ear will be recorded as a rare or uncertain manifestation that is not yet considered among the most characteristic findings of the disease. Sudden deafness or a secretion expelled by auditory canal are examples.

Our idea is that the Brazilian Society of Rheumatology should enhance and update a former study focusing on establishing better data on the hearing system. Even the articles about systemic lupus erythematosus (SLE), condition in which such changes are more frequently observed, did not add safe data about the problem. The main autoimmune conditions with otologic manifestations are referred below.

Concerning rheumatoid arthritis, the literature cannot provide too much, as it is more focused on the deleterious action of drugs, such as anti-inflammatory and antipaludic drugs, which are frequent causes of problems in patient ears. There are superficial records about a higher frequency of mild or moderate deafness in patients that have had the disease for many years.

Immune-mediated inner ear disease (IMIED) produces neurosensorial deafness. The patients complain about the reduction of acoustic acuity or reduction of sound discrimination. In general, it is bilateral, rapidLy progressive. It may be associated with vestibular symptoms. The syndrome mechanism is not completely clear, but it is accepted as an immunologic nature. It is known that the endolymphatic sac is an immunocompetent organ, and circulating antibodies, against antigens of the inner ear, as well as viral endolymph antigens, are found in this condition. However, specificity and sensibility and the role of those autoantibodies in the pathologic process are poorly explained. Generally, early use and high doses of corticotherapy solve or reduce the problem. In case of persistence of manifestations resistant to corticotherapy, another option should be methotrexate, with a good, regular and, sometimes, inefficient result, according to several papers published.

The differential diagnosis for IMIED patients includes: Ménièrie's disease, Lyme disease, hereditary progressive loss, degenerative deafness, acoustic trauma, acoustic neurinoma, other posterior cranial fossa, multiple sclerosis, neurosyphilis, ototoxicity, hypercoagulability, hematologic disease, antiphospholipid antibody syndrome, and other autoimmune diseases.

Patients with neurosensorial deafness that behaves as a limited form, unilateral or bilateral of IMIED, affecting medium ear, have been observed at a high frequency in lupus. This manifestation may be associated with high titers of anticardiolipin antibodies. Some authors record the possibility of this clinical finding in up to 22% of cases, which generally are unobserved, due to the severity of the other symptoms of SLE.

We should remember that some drugs used in SLE, such as chloroquine and related products, and some diuretics may be responsible for a similar case history. Sometimes, those hearing symptoms may be associated with an acute aortic insufficiency. These findings are not correlated with the presence or elevation of titers of antinuclear or anti-DNA factor.

In relapsing polychondritis, the existence of mild to severe deafness is observed, due to the extension of the inflammatory process for the medium ear and Eustachian tube. There are also audiovestibular manifestations due to endolymphatic hydrops. It is an autoimmune episodic disease that presents recurring inflammation of cartilaginous structures. It may be associated with other autoimmune diseases and the doctor should not forget the need for a research of acquired immunodeficiency syndrome (SIDA) at the moment uncommon symptoms occur. Recurrent auricular chondritis is found in 85% of the patients and this is a very suggestive finding of this disease. Other manifestations in the hearing system may be: otitis externa, chronic myringitis, dysfunction of the Eustachian tube, conductive deafness, tinnitus, and sensorineural deafness. All of them improve with corticotherapy. Laboratory studies and biopsies may contribute to diagnosis. For a correct administration, a combined action of an otorhinolaryngologist and a rheumatologist should be required.

In systemic sclerosis, neurosensorial deafness may appear predominantly in the medium ear, but there is also a high incidence of mixed deafness.

In some vasculitis, a painful sensation in the inner ear, redness in the auditory canal (auditory canal hyperaemia), serous medium otitis etc. may occur. Curiously, there is an association between antimyeloperoxidase antibodies and

deafness in patients with polyarteritis nodosa or microscopic polyangiitis, which sometimes are the initial manifestations of the disease.

Corticotherapy should be immediately initiated, as well as therapy for Ménière's disease, if necessary. After two weeks, an audiogram should be performed. If the result is not satisfactory, two more weeks of high doses of corticoid are suggested. Alternatively, intratympanic dexamethasone injection may be indicated, considering, however, that this procedure may cause eardrum infection or persistent perforation. Even if aggressive corticotherapy shows an insufficient response, the patient should be maintained with small doses (5 to 10 mg of prednisone/day). When corticotherapy fails, a 2 mg/kg/day oral cyclophosphamide dose may be used. Plasmapheresis has been showing some value. Recently, biological agent etanercept 25 mg has been used twice a week, subcutaneously for 5 to 10 months. There will be a 58% improvement and 33% stabilization. Tinnitus, as well as dizziness, will improve 90%.

Cogan's syndrome is a rare systemic disease with unknown etiology associated with a pattern that is characteristic to eye and inner ear inflammation. It is a vasculitis with participation of blood vessels in several sizes, including great arteries. Audiovestibular symptoms were correctly identified in 1940. Typically, the beginning is unilateral and gradually becomes bilateral. It progressively leads to deafness in 50% of the patients. Deafness may occur in the beginning. Some of them have Ménière's disease, without deafness, but with dizziness, nausea, and ataxia. Aorta should be carefully examined.

In Wegener's granulomatosis, there may be a conduction deafness resulting from granulomatosis participation in nasopharynx. There is also Eustachian tube dysfunction, severe medium otitis that may be purulent, central perforation of the eardrum, and neurosensorial deafness.

Some infectious conditions, like otosyphilis, should be remembered in a careful anamnesis. It is important to consider that neurosyphilis and inner ear syphilis are not the same condition. In this manifestation, AIDS verification is mandatory.

Concerning Brazilian data, there are few publications. In 1983, Atra *et al.*[2] published the article "Study of hearing in SLE" in *Revista Brasileira de Reumatologia*; Gomides *et al.*[3] published another article on deafness in SLE in which they observed that the problem affects a significant part of the patients.

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