Sensorineural hearing loss as the first manifestation of Sjögren's syndrome

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Introduction

Dysfunction of inner ear, vestibulocochlear nerve or central brain processing centers leads to sensorineural hearing loss (SNHL). Autoimmune inner ear disease (AIED) is a rare but potentially treatable cause of hearing loss, characterized by progressive evolution over weeks to months.[1] In 1958, an antigen-antibody reaction was described in 13 patients with progressive bilateral SNHL,[2] and later, in 1979, the term autoimmune SNHL was proposed after an idiopathic bilateral progressive SNHL had markedly improved with steroids therapy.[3] Since there are no defined diagnostic criteria for AIED, it remains a diagnosis of exclusion, supported by clinical suspicion and responsiveness to corticotherapy. About one third of the cases of AIED is secondary to a systemic autoimmune disease and may, rarely, be the first manifestation of the latter.[4,5]

Method

Description of a clinical case, based on the data referred to in the clinical process.

Results

We present the case of a 65-year-old woman who attended the otorhinolaryngologist due to progressive and cumulative unilateral left hearing loss over one month, without any other accompanying symptoms. She had been diagnosed with breast cancer eleven years before, being successfully treated with chemotherapy and right mastectomy. She was medicated for arterial hypertension and hypothyroidism. Her son had been recently diagnosed with a cerebellopontine angle tumor. She had no history of vertigo or ocular problems. Otoscopy was normal but the Weber test lateralized to the right and the Rinner test were both positive. The neurological examination was normal, including no cerebellar or vestibular findings. The remaining physical test was unremarkable.

An audiogram was performed, with left hearing loss described as -70 dB in the 8000 Hz range, confirming the suspicion of SNHL (Figure 1A). The routine laboratory workup and magnetic resonance imaging were normal.

She was started on 60 mg of prednisolone (1 mg/kg/day) with immediate response (Figure 1B). Steroid tapering was started and, after four weeks, she stopped the treatment. A relapse of symptoms occurred during the following month, with deterioration of hearing levels registered in the audiogram (Figure 1C). Corticotherapy was restarted and AIED was considered the most likely diagnosis, therefore she was referred to the autoimmune diseases clinic to exclude a systemic autoimmune disease.

She complained of dry eyes and mouth but denied arthralgia, parotid swelling or cold extremities, and had no history of recurrent abortion. The physical examination remained normal aside from mucosal dryness confirmed by the Schirmer test (< 5 mm in 5 minutes). Laboratory tests showed normal liver and thyroid function, with positive thyroid peroxidase antibodies. Antinuclear antibodies were positive – she had anti-mitochondrial M2 antibodies but anti-Ro and anti-La where negative. Other autoimmune tests, including rheumatoid factor, anti-neutrophil cytoplasmic, anti-centromere, anti-neuronal antibodies, and lupus anticoagulant, were negative. She was observed by an ophthalmologist who reported no significant findings. The thyroid echography showed a microgranular pattern. She was treated with pilocarpine and a scintigraphy was performed, showing hypofunction of the salivary glands. The biopsy revealed significant lymphoid infiltrate – class III on the Chisholm Mason scale – confirming Sjögren's syndrome (SS).

After four months of low dose corticotherapy (5 mg of prednisolone), she had no symptoms of hearing loss and her left audiogram showed a 50 dB improvement (Figure 1D).

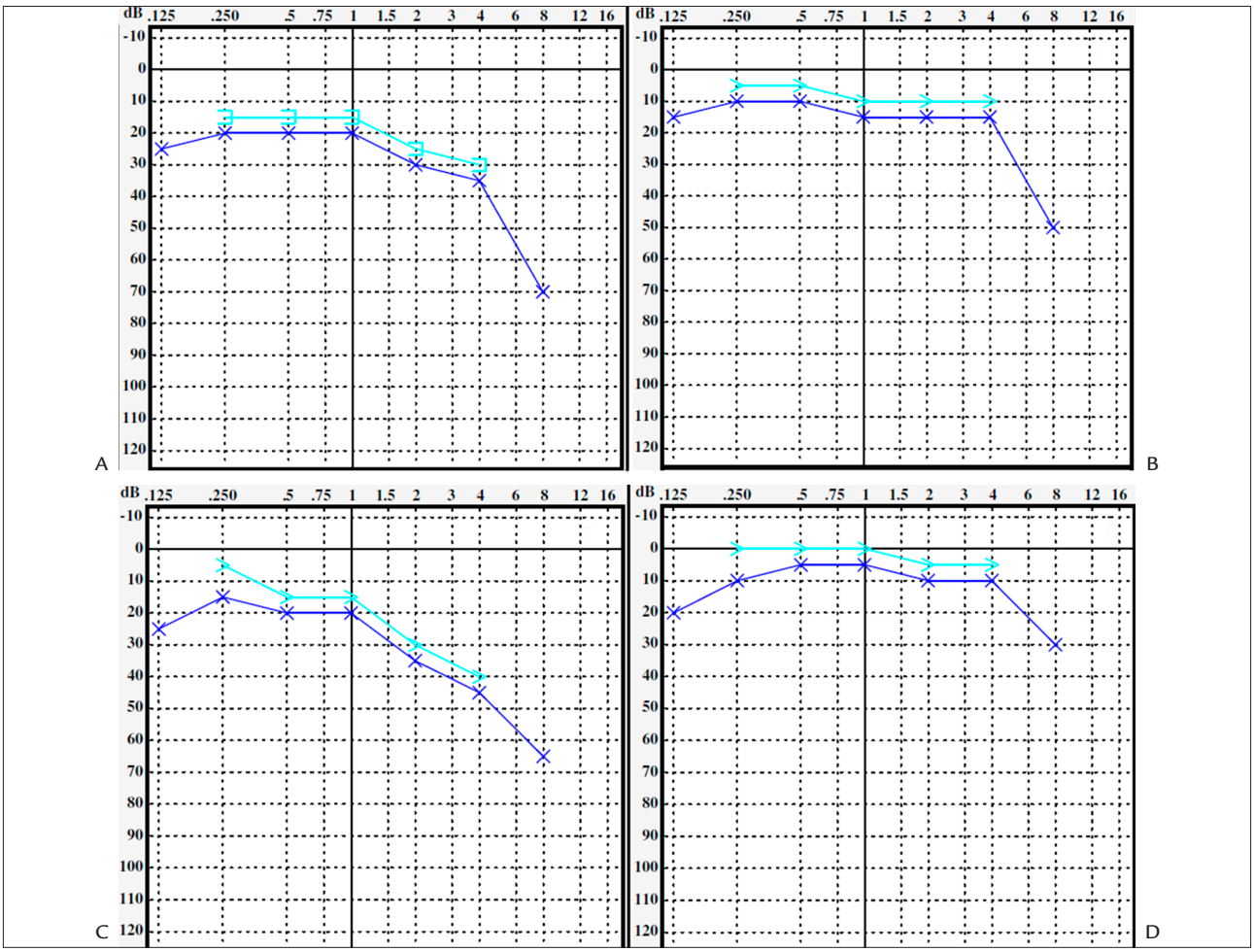

FIGURE 1 A. First audiogram, with left hearing loss described as -70 dB in the 8000 Hz range, confirming the suspicion of SNHL. B. Audiogram on 60 mg of prednisolone. C. Audiogram after stopping steroids, showing deterioration of hearing levels. D. Audiogram after four months of low dose corticotherapy, showing a 50 dB improvement.

Discussion

AIED is a SNHL which may have uni- or bilateral involvement and presents with progressive deterioration of auditory function over weeks to months. The time course is very important for the differential diagnosis between sudden hearing loss (hours to days) and presbycusis (years). It usually affects people in between the third and the sixth decade of life. Vestibular symptoms may be present.[3,5,6]

SNHL can be the initial manifestation of a systemic autoimmune disease such as SS, systemic lupus erythematosus, granulomatosis with polyangiitis (Wegener's),[7] polyarteritis nodosa, rheumatoid arthritis, and Cogan's syndrome.[8,9] There is a significant association between thyroid autoimmunity and Ménière's disease, which can itself be very difficult to distinguish from AIED in the first months,[10] but our patient had no vestibular symptoms. The relationship between SS and AIED was studied by Tumiati et al., who suggested the performance of audiometric tests in patients with this syndrome.[11] In this case, despite seronegative anti-Ro and anti-La, there were clinical, imaging, and histological findings that allowed the diagnosis of SS. Although anti-mitochondrial antibodies are characteristic of primary biliary cirrhosis, it is known that their presence in SS patients can occur, presenting a higher risk of liver involvement,[12] which increases the importance of surveillance by a multidisciplinary team. Systemic corticosteroids should be initiated as soon as possible since the prognosis is time dependent.[5]

Conclusion

This case, in which the medical findings led to the diagnosis of SS, illustrates the importance of a multidisciplinary approach and clinical suspicion of an autoimmune

cause for progressive SNHL. After a response to corticotherapy, the possibility of an association with autoimmune systemic diseases should be thoroughly investigated.

Key messages

- Autoimmune inner ear disease is a diagnosis of exclusion, supported by clinical suspicion and responsiveness to corticosteroids.
- It is secondary to an autoimmune disease in one third of the cases.
- Although it is an unusual etiology of hearing impairment, it is important to recognize it, as early diagnosis and treatment can have a marked effect on the clinical outcome.

Notes

- No funding to declare.
- No animal or human studies were carried out by the authors for this article.
- The authors declare that none of the presented data allows for the identification of the patient.

Conflict of interest

The authors declare no conflict of interest.

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