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Letter to the editor

Auditori damage as part of neurological signs in Behçet disease

Daño auditivo como parte de las manifestaciones neurológicas en la enfermedad de Behçet

To the Editor:

Behçet's disease (BD) is a multisystemic inflammatory disease with a chronic progression characterized by flares and no associated autoantibodies.[1] Among the recognized characteristics of the disease, due to their severity and frequency are the neurological complications. Among these, involvement of the inner ear is one of the most frequent, although on many occasions it goes unrecognized. We present the case of a 43-year-old woman, allergic to metoclopramide, who was diagnosed with BD in 1998 based on the presence of recurrent oral and genital ulcers, axial and peripheral joint pain, recurrent lower limb phlebitis, and lymphocytic meningitis on 3 occasions. After that she presented a visual deficit and a homonimal right superior quadrantopsia was confirmed. Physical examination was normal, including a retinal examination. Laboratory testing (blood count, erythrocyte sedimentation rate [ESR], biochemistry, C-reactive protein [CRP], ANA [antinuclear antibody], ENA [extractable nuclear antigen], antineutrophic cytoplasm antibodies, urine, cerebrospinal fluid analysis) was normal or negative. The study of the thrombophilia showed a deficit of activated C protein without mutation of Leyden´s factor V. Anticardiolipin antibodies were negative on 3 occasions and no testing was performed looking for antibeta 2 glycoprotein antibodies. Central nervous system magnetic resonance (MR) with gadolinium did not show any pathologic findings and visual potentials were compatible with a right perichiasmatic lesion. Treatment was started with prednisone at a descending dose (from 30 mg/day to a maintenance dose of 5 mg/day), azathioprine (100 mg/day), colchicine (0,5 mg), and dicloverine chloride (5 mg, 1 tablet BID), acenocumarine, diclofenac (50 mg/BID), and rabeprazol (20 mg/day).

The patient came to regular visits every 4 to 6 months and referred episodes in which her joint pain worsened, without other signs of activity, with stabilization of the visual deficit. During her 2007 check-up visit, she was routinely interrogated and examination revealed a retromastoidal adenopathy of an approximate diameter of 0.5 cm, unfixed and with mild pain; no other adenopathies were detected; cardiac and respiratory examination, abdominal palpation, and peripheral pulses were all normal. Laboratory tests (ESR, CRP, ANA, ENA, serum proteins, Epstein-Barr and cytomegalovirus serology, and urine testing) were all normal. Cervical ultrasound informed that a "solid mass approximately 6 mm in diameter was located in the right retromastoidal region." The patient was referred to the Ears Nose and Throat Department for a biopsy of the lymph node and during direct interrogation she reported "hearing loss" and "muffled sounds." A computerized tomography (CT) and an MR of the ear and auditory canals was requested, evidencing hypertrophy

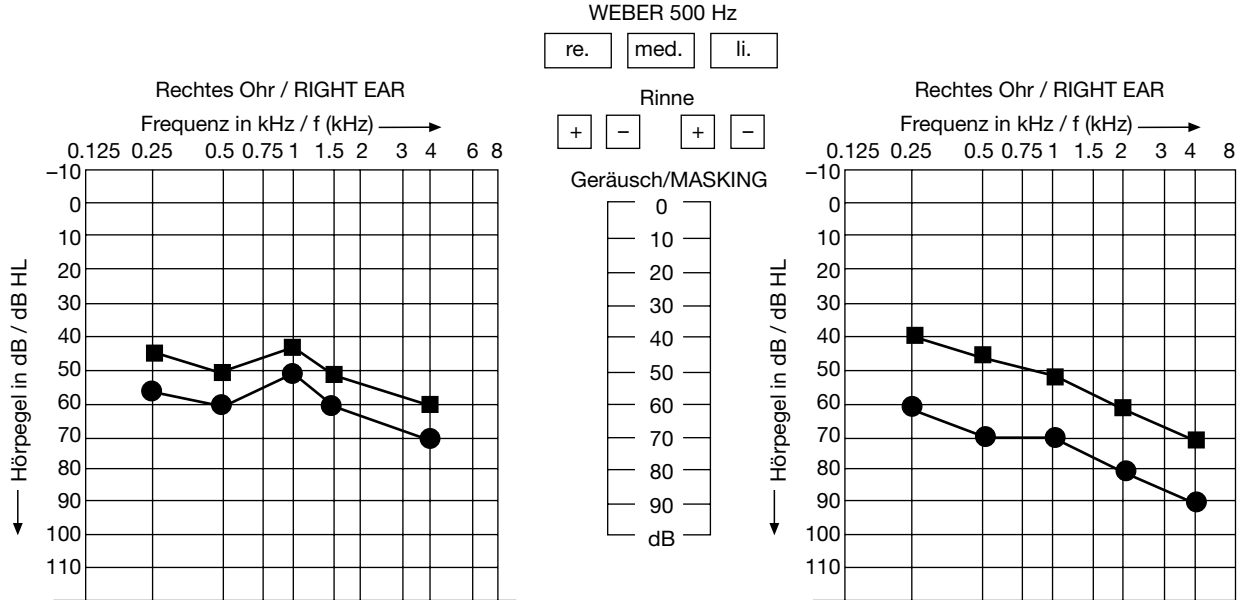

Figure. A) Right ear: perceptive hypoacusia at 50 dB with a fall starting at 1000 Hz down to 70 dB with a presbiacusia type curve. B) Left ear: perceptive hipoacusia with a fall starting at 1000 Hz down to 70 dB, reaching 90 dB. C) Left and right auditory pathways presenting perceptive endocochlear hypoacusia.

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of the mucosa of the left inferior mastoid cells; this was considered incidental. This complementary testing ruled out box occupation or VIII cranial nerve involvement. Otoscopy was normal. Biopsy informed of a "non-specific chronic lymphadenitis." Audiometric tests and hearing potentials (left and right auditory pathway presented endocochlear perceptive hypoacusia) confirmed the diagnosis of perceptive hypoacusia. In later audiometric studies, and because the patient had been under different steroid treatment regimens, the hearing loss was considered to be irreversible (Figure).

Most studies estimate that in BD the prevalence of neurosensitive hearing loss[2–4] oscillates between 12% and 80%, and that the prevalence of vestibular affection[5] occurs around 20%–40%. Symptoms that oblige us to suspect this neurosensitive affection are hearing loss, "muffling" of sounds or tinnitus. Neurosensitive damage can develop slowly and these symptoms can pass undetected for months or years, as occurred in our patient. The presence of "muffling" referred to by our patient when interrogated directly led to this suspicion. It seems that perivasculitic or thrombotic phenomenons are the anatomopathological substrate of this problem. Some studies consider that neurosensitive hearing loss is related to the severity of disease,[6] and that this is more frequent in patients with multi-organ affection, as was the case in our patient. When reviewing the literature we have not found evidence that evaluates the role of steroid or immunosuppressive treatment on hearing loss in BD. However, it seems logical to suggest that the control of the inflammatory process using high dose steroids in the initial phases of the process can prevent irreversible damage.

As a conclusion to the present case and review, we would like to emphasize that auditory affection seems to be a relatively frequent manifestation of BD, in which the clinical suspicion generated by directed interrogation would lead to an early diagnosis and the possibility of avoiding it becoming irreversible.

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