Rare Diseases of the Immune System

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Lorenzo Emmi *Editor*

Behçet's Syndrome

From Pathogenesis to Treatment

Springer

Audio Vestibular Involvement in Behçet's Disease

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Paolo Vannucchi and Rudi Pecci

The audio-vestibular system is often involved in many autoimmune syndromes as Cogan's syndrome and Granulomatosis with polyangiitis (Wegener's granulomatosis) but an involvement in Behçet's disease is reported more frequently only in the last years. Behçet's disease was first defined by Hulusi Behçet in 1937. It is a refractory multisystem disorder mainly presenting with recurrent oral aphthae and genital ulcerations, skin lesions, and uveitis. The disease is a chronic inflammatory disorder involving the small vessels, which is of unknown etiology. It has a worldwide distribution with a prevalence ranging from 1:1,000 to 1:10,000 in Japan and Turkey to 1:500,000 in North America and Europe. The aetiopathogenesis is obscure but Behçet's disease is considered to be immune-mediated.

Audio-vestibular disturbance, including hearing impairment, tinnitus, and dizziness, is one of the multisystemic characteristics of Behçet's disease.

There are studies and case reports in the literature about the inner ear involvement, sudden cochlear sensorineural hearing loss (SNHL) and incidence of SNHL in Behçet's disease. In these studies, the incidence of HL has been reported as 12–80 %.

Auditory and vestibular lesions, among the clinical manifestations of central nervous system (CNS) involvement, were first described by Alajouaine et al. [1]; the authors described SNHL and gaze paretic nystagmus in a patient with Behçet's disease suffering from meningoencephalitis. Since then, other authors have shown that SNHL and dizziness are frequent symptoms in patients affected by Behçet's syndrome with or without the involvement of the CNS. Erdinç et al. [2] suggest

P. Vannucchi (✉) · R. Pecci
Department of Surgical Sciences and Translational Medicine, Unit of Audiology, Careggi Hospital, University of Florence, Largo Giovanni Alessandro Brambilla 3, 50134 Florence, Italy
e-mail: paolovannucchi@libero.it

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that presence of SNHL in Behçet's disease is prevalent and represents a fourth clinical symptom after oral, genital ulcers, and skin lesions: a statistically significant SNHL at high frequencies when compared with the speech frequencies has been reported and this high incidence of SNHL can be attributed to small venous disease that is seen in the majority of neuro-Behçet patients. Kulahli et al. [3] carried out a study to determine the characteristics and incidence of SNHL and vestibular disturbance in Behçet's syndrome with a large number (62) of patients, and they found that (1) the hearing and vestibular disturbances in Behçet's syndrome are more prevalent than previously recognized, (2) SNHL in high frequencies in Behçet's patients is an indicator of cochlear involvement, (3) there is a higher prevalence of central vestibular syndrome in Behçet's patients than it was thought before, and (4) HLA-B51 antigen may be able to be a prognostic factor for SNHL in Behçet's patients.

Behçet's disease is typically characterized by a lack of correlation between other organs and audio-vestibular involvement, which is in keeping with the multifocal nature of the disease process. Also, a lack of correlation was found between the auditory and vestibular lesions, but this may be explained by an understanding of the vascular supply: the common cochlear artery and anterior vestibular artery are the main branches of the labyrinthine artery and can be selectively involved by immunologically mediated inflammation. Furthermore, the incidence of dizziness is often more common than audiologic symptoms, probably due to the more diverse causes of dizziness, including peripheral and central vertigo.

In an interesting study, comparing Behçet's patients with sex and age matched healthy subjects, Süslü et al. [4] showed that in younger patients there could be a lesser ratio of SNHL compared to older ones. However, although hearing thresholds were within the normal limits according to pure-tone average—PTA- (at 500, 1,000, 2,000, and 4,000 Hz), hearing levels of the patients with Behçet's disease were found to be higher than the controls at most of the single frequencies and differences were seen to be significant. Furthermore, in the high frequencies (9,000–16,000 Hz) 63.4 % of the patients had hearing loss and the differences between the hearing levels of patient and control group tend to increase definitely in high frequencies. This finding points a cochlear involvement in Behçet's disease beginning and prominent in basal turn of the cochlea. In these patients it also could be found decreased responses with otoacoustic emission (OAE) and lower values of signal to noise ratio (SNR); this means that the physiological motility of the outer hair cells of the patients with Behçet's disease is lesser than the normal subjects. Meanwhile, normal hearing thresholds together with the decreased OAE responses could be accepted as the early findings of sub-clinical cochlear involvement in Behçet's disease. Finally, not only the duration of the disease, but also clinical features or other system involvements usually do not correlate with hearing levels. These data show that cochlear involvement can be regarded as an early sign and takes place even in patients without significant organ involvements.

Erbek et al. [5] suggest an association between delayed vestibular evoked myogenic potentials responses (VEMP) and Behçet's disease. Prolonged VEMP responses in patients with Behçet's disease maybe explained by chronic

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inflammation involving the pathways of the sacculo-collic reflex. Since abnormal VEMPs are not always associated with caloric weakness, it can be speculated that inferior and superior vestibular nerves might be damaged separately in Behçet's disease. Therefore, the VEMP response may be a useful diagnostic tool for vestibular evaluation in this disease.

Even Cinar et al. [6] more recently revealed that the prevalence of SNHL was significantly higher in the Behçet patients than in the controls. The duration of Behçet's disease had no significant impact on whether patients did or did not experience hearing loss. The authors reported hearing loss was the fourth most common clinical finding in the Behçet group, after oral ulcers, genital ulcers, and skin lesions, and they suggest the need for an adequate investigation of hearing in the routine follow-up of these patients.

Indeed, Behçet's disease may present with features other than the classic triad of symptoms. Raised awareness of the clinical features within the head and neck region will hopefully enable early diagnosis and treatment of this potentially serious condition.

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