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Cochleovestibular Syndrome as an Atypical Manifestation of Neuro-Behçet's Disease

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Abstract **Case Report**

Behcet's disease (BD) is a chronic multisystem disorder that can affect the skin, mucous membrane, and central nervous system. It can be characterized by symptoms such as cognitive impairments, psychiatric symptoms, and focal neurologic deficits, we present the case of a 56 years old neuro-BD with a 16 years history of cephalea, diplopia, generalized tonicoclonic comital seizures and amnesia stabilized on azathioprine 6 months later he developed a brutal deafness associated with tinnitus of the left ear and a spontaneous vertigo. This case report points out to the diagnostic and therapeutic challenges of BD especially when unusual symptoms are the prominent manifestations of the disease. It also discusses the different therapeutic options for patients with BD and cochleovestibular symptoms.

Keywords: Neurobehcet Disease, Hearing Loss, Vertigo, Tinnitus, Treatment.

INTRODUCTION

Behcet's disease (BD) initially described by Hulusi Behcet in 1937 is a chronic, remitting and inflammatory disease affecting mainly the mucocutaneous system, the eyes and joints, and the vascular and central nervous systems. The incidence of BD differs from region to region, being particularly common in the Mediterranean region, and in the Middle East and Far East. Vasculitis and secondary autoimmunity developing in response to the accumulation of immune complexes in small and intermediate blood vessels, have been identified in recent clinical studies as playing a major role in the pathogenesis of BD [1].

Today BD is considered as a multisystem disease often shows heterogenous clinical manifestations with variable involvement of many organs including central nervous system, heart, kidneys, and respiratory and gastrointestinal tracts. In recent years cochleovestibular system also was accepted among the others that BD may involve. The incidence of otological involvement in BD ranges from 12 to 80% in several studies [2].

CASE REPORT

A 56-year-old man whose symptomatology dates back to 16 years ago with the onset of helmet cephalea, Uveitis, generalized tonicoclonic comital seizures and amnesia associated with the onset of oral and genital aphthae. Brain MRI revealed a lesion involving the midbrain. Based on these criteria, a diagnosis of Neuro-Behçet's disease was established. The patient was put on boluses of methylprednisolone + colchicine for his disease and on valproic acid and levetiracetam as antiepileptic and then azathioprine for 1 year with stabilization of his disease.

The evolution was marked by the installation 6 months ago of a brutal deafness associated with tinnitus of the right ear and a spontaneous vertigo, the whole evolving in a context of apyrexia and conservation of the general state. On examination, the patient was conscious, hemodynamically and respiratorily stable. Vestibular examination was normal. Pure tone audiometry showed a mixed hearing loss in the right ear with an air bone gap at 15 dB on 3 successive audiograms spaced of 15 days (Figure1). The tympanogram was normal, the stapedial

reflexes were negative on the right ear. Acoustic otoemissions were normal. Auditory brainstem response showed a prolongation of the latency of the wave V. The eye-tracking test was abnormal with a low gain. The ocular saccades test was normal. The optokinetic reflex was abnormal with a low gain. The Head Shacking Test was negative. The caloric tests showed left hyporeflexia (Figure2).

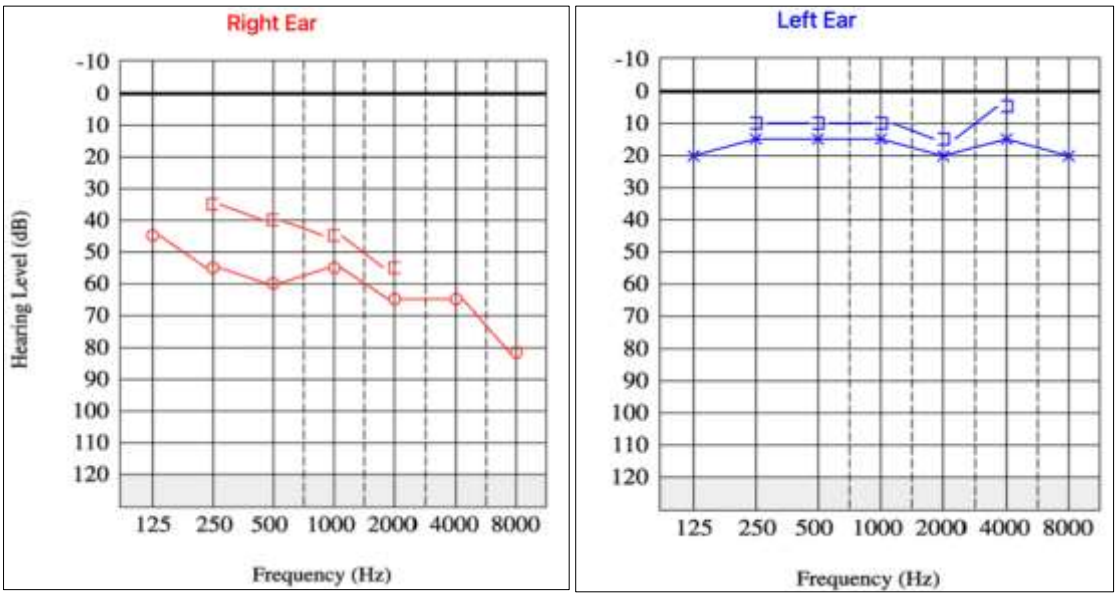

Figure 1: Pure tone audiometry: mixed hearing loss in the right ear with an air bone gap at 15 dB

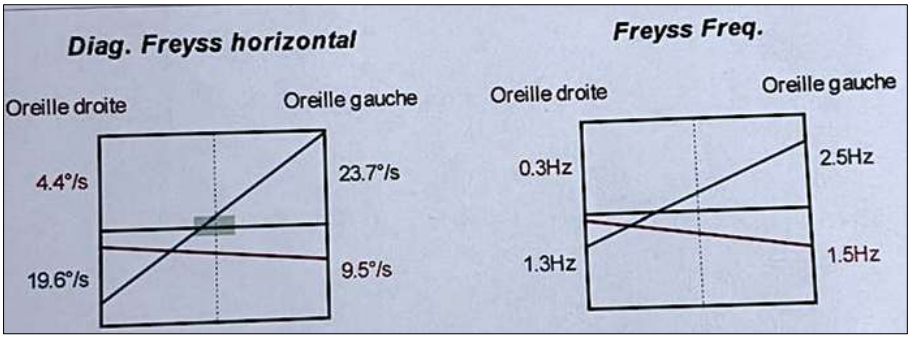

Figure 2: Caloric tests: right Hyporeflexia

The patient underwent a therapeutic adjustment. He was also fitted with a conventional hearing aid in the left ear and underwent 20 sessions of vestibular rehabilitation. This therapeutic approach led to partial recovery of cochleovestibular function, with complete resolution of vertigo and noticeable improvement in hearing loss and tinnitus.

DISCUSSION

First described in 1937 by Dr Halusi Behçet in Istanbul, Behçet's disease was prevalent along the Silk Road, but became widespread with immigration. It is a chronic relapsing inflammatory disorder of unknown aetiology, affecting small vessels of nearly all organs. The underlying histopathologic findings include leukocytoclastic vasculitis, fibrinoid necrosis of postcapillary venules, or perivascular neutrophilic accumulations. Originally described as a classical triad of oral ulcers, genital ulcers and ocular disease including anterior uveitis, it is now acknowledged to be multisystemic. Central nervous system involvement occurs in 10–25% of patients and can span the entire neuraxis. Audiovestibular symptoms include vertigo, sudden sensorineural hearing loss, tinnitus and orthostatic imbalance. Vertigo lasting several days, occurring alone or with hearing impairment, has been described. The prevalence of otological involvement ranges between 12–80% in separate studies. Audiometry in these patients showed mild sensorineural hearing loss without specific attributes that enable its recognition. Based upon diverse auditory brainstem response abnormalities ranging from normal waveforms in the presence of sensorineural hearing loss (implying cochlear pathology) to delayed I–III or I–V interpeak latencies (indicating brainstem involvement), the auditory pathways are thought to be involved at multiple sites. Caloric weakness implying abnormal horizontal canal function and prolonged VEMP peak latencies (suggesting central vestibulopathy) have been reported [3].

In the study made by Kulahi and Al Sixty-two patients with Behcet's syndrome were included, 34 men and 28 women whose mean age was 33.7 (15–60). Sixty-two healthy normal control subjects (38 male and 24 female) were included. Patient and control groups were questioned about any history of audio-vestibular disturbance and underwent physical and ENT examination and the following audiologic tests: pure tone audiometric test (0.25, 0.5, 1, 2, 4, and 6 kHz), tympanogram, speech discrimination, short increment sensitivity index, tonedecay test, auditory brainstem response. Vestibular system was evaluated by videonistagmogram and caloric test. Cranial and brainstem magnetic resonance imaging (MRI) of patients who have vestibular disturbances were practiced to examine the central nervous system. Both the patient and the control groups were tested with the HLA-B51 antigen. Pure tone audiogram showed sensory-neural hearing loss (P25 dB hearing level in at least two frequencies) in 20 of the 62 (32%) Behcets patients while the control group were normal. There was a hearing loss involving high frequencies in the audiograms of Behcets patients with hearing disturbances. The recruitment investigation tests and auditory brain stem response confirmed cochlear involvement in all 20 patients. Caloric stimulation tests revealed a normal vestibular function in all patient and control group. In electronystagmography, 21 (34%) patients had hypometric or hypermetric saccades and smooth pursuit tests showing that 4 (6%) patients had pathological changes while the control group was normal. HLA-B51 antigen was found positive in 15 of 20 Behcets patient with hearing loss. Conclusion: The hearing and vestibular disturbances in Behcets syndrome is more prevalent than previously recognized; Hearing loss in high frequencies in Behcets patients is an indicator of cochlear involvement in this disease; There is a higher prevalence of central vestibular syndrome in Behcets patients than it was thought before; HLA-B51 antigen may be able to be a prognostic factor for sensorineural hearing loss in Behcet' s patients [4].

In a study by Marsili *et al*., a 15-year-old-boy with six months of history of fever, dizziness, tinnitus and ataxia who had an autoimmune inner ear disease associated to BD was successfully treated with the anti-tumor necrosis factor (TNF)-α at 24 mg/m2 subcutaneously every other week. His symptoms completely resolved soon after the therapy was introduced, Follow-up audiometry, laboratory evaluation and ocular screening recorded normal values. At two-year follow-up, the boy continues adalimumab treatment and is symptom free [5].

Utsunomiya *et al*., reported a 52-year-old female patient whose BD was revealed by acute bilateral panuveitis, oral aphta, genital ulcers and glaucoma secondary to uveitis. These symptoms were controlled with systemic immunosuppressive agents. Later, she developed a sudden right-sided hearing loss. Prednisolone 80 mg/day was administered and her hearing loss improved immediately. Prednisolone was gradually reduced and discontinued over two weeks. Thereafter, from August 2014 to October 2017, she experienced five episodes of unilateral or bilateral sensorineural hearing loss without any recurrent BD-related signs, and GC treatment was repeated when hearing loss relapsed, glucocorticoids were discontinued due to severe glaucoma deterioration, TNF-α blockers (adalimumab) were introduced without relapse and with improvement in hearing loss [6].

Elidan *et al*., suggest that cyclosporine might serve as an important mode of treatment of sensorineural hearing loss on an inflammatory "autoimmune" background, he conducted a study that involved thirty-five patients with Behcet's disease (20 under cyclosporine treatment and 15 under the conventional therapy), 12 patients suffering from endogenous uveitis, and 35 normal subjects were evaluated audiologically before entering the study and were followed up for at least a year. Twenty-eight Behcet patients (80%) showed some degree of hearing loss. The averaged pure tone audiogram of the Behcet group showed statistically significant auditory deficits in comparison with that of the control group. None of either the Behcet group or the uveitis group showed any hearing deterioration during the follow-up period. Five Behcet patients under CyA therapy demonstrated improvement in their hearing [7].

CONCLUSION

Even though the diagnosis of BD is very rare, we have documented a significant case of neuro-BD that manifested with sensorineural hearing loss, Recently, the triad of oral–genital–ocular symptoms in BD has been replaced by one of a multifaced disease with additional features, including involvement of the CNS, skin, joints, intestines and blood vessels [8, 9], Cyclosporine and Anti-TNF-α agents can be promising drugs for Autoimmune sensorineural hearing loss and in order to avoid the adverse effects of GC [7, 6].

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