Report Of A Case Of Sudden Unilateral Audiovestibular Disturbance In A Patient With Adamantiades Behçet’s Disease And Review Of The Literature.

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Citation


Abstract

Introduction: Adamantiades-Behçet’s disease is generally known as an autoimmune disease characterised by multisystem generalized vasculitis with perivascular infiltration. Typical loci of manifestations are the mucous membranes, skin and eyes, as well as the joints. The central nervous system is often involved in Adamantiades-Behçet’s disease and diverse neurological manifestations have been described. Disequilibrium or isolated hearing impairment is also reported in the literature, especially in the Middle East. Case Report: We present a case of a patient, with a 20 year history of mucocutaneous and ocular manifestations of Adamantiades-Behçet’s disease. The patient presented with a rarely described episode of sudden unilateral, low and middle frequency hearing deterioration, tinnitus and peripheral vestibular disturbance. He received steroid therapy, which lead to significant remission of all his symptoms. Discussion: Inner ear involvement in patients with Adamantiades-Behçet’s disease in Middle East populations is documented, and there has been an effort the past few years to determine its characteristics. Predominant patterns of the auditory and vestibular impairment and potential predictive markers for their appearance have been suggested, but there exist cases with distinct and uncommon characteristics. The administration of corticoides improves the symptoms of the disease especially those involving the inner ear.

INTRODUCTION

Adamantiades-Behçet’s disease was originally described as a clinical triad of recurrent aphthous stomatitis, genital ulceration, and uveitis,1 but today it is considered a multisystem disease, characterized by chronic, generalized vasculitis with perivascular infiltration that may involve many organs - skin, joints, veins, arteries, central nervous system, gastrointestinal tract, and lungs (Table 1).

Only 5% of patients with Adamantiades-Behçet’s disease present with neurological complaints and symptoms, but 25–30% eventually develop neurological manifestations, which are more frequently found in patients from northern Europe and North America. The most common lesions are: meningoencephalitis, benign intracranial hypertension, a multiple sclerosis-like picture, pyramidal involvement, and psychiatric disturbances (Table 2).6, 7, 8
Auditory and vestibular lesions, among the clinical manifestations of central nervous system involvement in patients with Adamantiades-Behçet’s disease, were first described by Alajouanine in 1961. Since then, other studies have tried to determine the incidence, pathogenesis and characteristics of these lesions and the detailed relationship between them.

We present the case of a 45 year old male with Adamantiades-Behçet’s disease who had inner ear involvement without any other neurological symptoms and discuss its rare characteristics compared to the predominant patterns of the disease in the present literature.

CASE REPORT

A 45-year-old man presented to our clinic with an episode of true rotatory vertigo. He reported that approximately two days before attending the hospital he experienced tinnitus and hearing loss accompanied by fullness in his left ear. He had no symptoms of diplopia, dysarthria, dysphagia, or sensorimotor changes and his laboratory findings were within normal limits. The otorinolarygologic examination was positive for decreased left vestibular function - first grade horizonto-rotatory nystagmus directed to the right side, seen under Frenzel’s glasses, was present and eliminated by visual fixation and caloric testing was positive. Audiometry revealed moderate hearing loss located in the low and middle frequencies with a peak at 4 KHz (Audiogram 1, Table 3).

Otoscopic examination findings were normal, with intact, transparent and mobile tympanic membranes. Past medical history was unremarkable, with no mention of noise exposure, ototoxic drugs usage, ear surgery, cranial trauma, metabolic diseases or other factors associated to hearing deterioration and vestibular disturbance. The patient reported no allergies and was not at that time taking any medications. He had a 20-year history of recurrent oral aphthae, uveitis and five episodes of scrotal ulceration (Adamantiades-Behçet’s disease according to the International Study Group for Behçet’s Disease criteria). He had presented no other dermamatologic manifestation of the disease and no symptoms of other organ, central nervous system or inner ear involvement during his past medical history. He had a negative pathergy test, was HLA B51 positive and had received treatment by his Rheumatologist, according to the occasional clinical presentation of his disease during that period of time - topical glucocorticoids in the form of mouthwash for the oral aphthae, systemic glucocorticoid therapy (prednisone, 1 mg/kg per day) and azathioprine (2 to 3 mg/kg per day) for the uveitis.

During his present admission, after consulting with his Physician, we decided to administer a corticosteroid regimen (prednisone, 1 mg/kg per day for a period of 10 days). The therapy lead to significant improvement of all the symptoms (retreat of the vertigo, reduction of the tinnitus and significant improvement of the hearing impairment and the ear fullness sensation) seven days later (Audiogram 2, Table 4).
The patient was released 10 days after admission and on post release follow up 6 months later showed no sign of relapse.

DISCUSSION

Adamantiades-Behçet's disease has an incidence that ranges from 1:10,000 in Japan to 1:500,000 in North America and Europe. It affects mainly young adults, with males having more severe disease. A genetic susceptibility of the disease has been noted in certain populations in the Mediterranean and the Middle East, with a significant HLA B51 positivity among patients.\(^2,3\) Since the first description by Alajouanine\(^9\) who described hearing loss and gaze paretic nystagmus in a patient with Adamantiades-Behçet's disease suffering from meningoencephalitis, many authors have subsequently reported audio-vestibular disturbances in Adamantiades-Behçet's patients, especially of Middle and Far East origin, with or without central nervous system involvement.\(^9,10,11,12\)

Adamantiades-Behçet's disease is autoimmune in nature and is characterised by multisystem generalized vasculitis with perivascular infiltration of T lymphocytes, B lymphocytes and neutrophils. These cells secrete proinflammatory cytokines such as TNF-alpha, IL-1, IL-6, cytokines that may cause vascular endothelial injury and dysfunction that may affect multiple organs.\(^13\) Due to the multifocal nature of the disease process there seems to be no correlation between audiovestibular and other organ involvement.\(^14,15\) Moreover auditory and vestibular symptoms may or may not coexist in a patient, as the auditory system and the vestibular system can be separately affected.

The later may be explained by an understanding of the vascular supply.\(^10,15\) The cochlea, sacculae, and posterior canal are supplied by the common cochlear artery, whereas the utricles, together with the anterior and horizontal canals, are supplied by the anterior vestibular artery.\(^10,15\) 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, causing distinct clinical manifestations. Thus, a lesion of the anterior vestibular artery would manifest as an isolated vestibular abnormality in routine clinical vestibular testing, whereas involvement of the common cochlear artery would show as an abnormality in audiologic testing.\(^10\)

The incidence of hearing loss in Adamantiades-Behçet's disease according to reports in the literature varies considerably, between 12% to 80%.\(^12,14,15,16,17,18,19,20,21,22,23\) Similarly the incidence of vestibular involvement is reported to be between 20% to 40%.\(^9,16,19,24\) Despite the fact that a consensus has not been reached, most studies suggest no statistically significant relationship between the age or the disease duration and inner ear involvement.\(^10,11,12,15,21,22\) Furthermore a statistically significant lower frequency of positive pathergy test and a higher prevalence of HLA-B51 antigen in Adamantiades-Behçet's patients with hearing loss should be noted. These findings make pathergy test and HLA-B51 potential prognostic factors for sensorineural hearing loss development in these patients.\(^12,21\)

Whereas in the most recent literature the predominant pattern of auditory involvement was characterized by a progressive bilateral high-frequency sensorineural hearing loss, with a down slope (4, 8, 10, and 12kHz),\(^15,21,22,25\) in the present case the hearing deterioration was unilateral and localized mainly in low and middle frequencies, with relative sparing of the hearing in high frequencies (audiogram 1). Furthermore its sudden onset is very rarely described in the literature. Considering the studies with the largest groups of patients we discover that none of the 63 patients in the study of Erdinç,\(^15\) none of the 27 in the study of Mahdi\(^22\) and only one of the 62 in the study of Kulahli\(^21\) experienced sudden unilateral hearing loss. In addition the auditory impairment was accompanied by vestibular disturbances, as a result of the vasculitic process, that apparently affected both the common cochlear artery, causing hearing deterioration and the anterior vestibular artery producing symptoms of a typical peripheral vestibular dysfunction. The relatively young age of the patient further
supports the literature findings that suggest no relationship between the age of the patient and inner ear involvement, while the fact that he was HLA B51 positive and pathergy test negative is in accordance with the prognostic value of these parameters. Conventional corticosteroid therapy was sufficient in relieving the patient’s symptoms and there was no need for the use of immunosuppressive regimens.

References
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