Diffuse Systemic Sclerosis Presenting as Meniere's Disease-Like Symptoms as Part of Autoimmune Inner Ear Disease

Joy Feld MD[1], Avi Shupak MD[2,3,4] and Devy Zisman MD[1,4]

[1]Rheumatology Unit, Carmel and Lin Medical Centers, Haifa, Israel
[2]Otoneurology Unit, Lin Medical Center, Haifa, Israel
[3]Department of Otolaryngology, Head and Neck Surgery, Carmel Medical Center, Haifa, Israel
[4]Rappaport Faculty of Medicine, Technion-Israel Institute of Technology, Haifa, Israel

KEY WORDS: systemic sclerosis (SSc), autoimmune inner ear disease (AIED), sensorineural hearing loss (SNHL), Meniere's disease

IMAJ 2015; 17: 263–264

Ear involvement has been described in systemic sclerosis (SSc), where hearing loss and vestibulopathy coexist with other disease manifestations [1,2]. We present an unusual case of SSc where Meniere's disease as part of autoimmune inner ear disease (AIED) was the first presentation of diffuse SSc. The literature regarding audiovestibular involvement in SSc is reviewed.

PATIENT DESCRIPTION

A 44 year old previously healthy woman presented after 2 months of recurrent vertigo, nausea and vomiting, in addition to intermittent right ear tinnitus and fullness. Physical examination was positive for first-degree left beating spontaneous nystagmus. Otoneurological laboratory evaluation indicated right ear mixed sensorineural and conductive hearing loss and right peripheral vestibulopathy involving both the semicircular canals and otolithic organs. A diagnosis of definite Meniere's disease was reached. Treatment with a low salt diet, acetazolamide 250 mg once a day and betahistine 16 mg three times/day was recommended. The vertigo attacks resolved. Follow-up audiometry showed progressive bilateral mixed hearing loss. Serological evaluation revealed solely antinuclear antibodies. A diagnosis of AIED was reached. Prednisone 1 mg/kg and aspirin 100 mg/day resulted in fair improvement. Methotrexate 20 mg/week was added.

Three years after her initial presentation, while on a regimen of 20 mg/week methotrexate and 5 mg/day prednisone, the patient developed Raynaud's phenomenon, accelerated skin fibrosis with progressive sclerodactyly, arthritis and dyspepsia. Scl-70 antibodies became apparent on the immunological evaluation. The diagnosis of diffuse SSc was reached. Gradually other disease manifestations appeared, including severe skeletal and cardiac blunt myopathy, lung fibrosis, esophageal dysmotility, "watermelon" stomach and paralytic ileus.

Sequential treatment with high dose steroids, cyclophosphamide, mycophenalate mofetil, IV immunoglobulins and bosentan all failed to attenuate the progressive diffuse SSc. Five years after her presentation the patient died of a fatal arrhythmia.

COMMENT

The prevalence of bilateral sensorineural hearing loss (SNHL) among SSc patients ranges from 20% to 69%, while a conductive component contributes to the hearing loss in 3–11% of them [1,2]. Dizziness and vertigo have been reported in 21–75% of patients [2], with a specific diagnosis of benign paroxysmal positional vertigo (BPPV) in 17% [3].

Hearing loss and vestibular derangement have been mostly reported in patients with an established diagnosis of SSc and not as the presenting symptom of the disease [4]. In addition to the present case, to the best of our knowledge there is only one other description of AIED as the initial symptom of SSc. In that case of limited SSc, bilateral sudden sensorineural hearing loss preceded the appearance of scleroderma skin changes by several months [4].

Another unique feature of our patient was her initial Meniere's disease-like symptoms. Meniere's disease-like symptoms have not been previously described in SSc although they have been reported in other rheumatic diseases. The severity of hearing impairment in SSc patients is highly variable, ranging from asymptomatic high frequency loss not affecting speech reception and discrimination to a flat audiogram with profound hearing loss [2].

Most previous publications [2] did not find a significant correlation between hearing loss and age, duration, type and severity of SSc systemic manifestations of the disease, presence of autoantibodies and drug therapy. A single cohort study reported a significant correlation between the presence of pulmonary hypertension digital ulcers and abnormal audiometric tests in a limited number of SSc patients [1]. In another study that included diffuse SSc patients, older age correlated significantly with the SNHL severity [2].

Although cochlear, middle ear and vestibular end-organ involvement has been suggested as the common etiology for hearing loss, dizziness and vertigo, other factors have also been described in SSc patients, such as auditory neuropathy, cranial nerve VIII involvement and central vestibular abnormalities.

It has been suggested that the reported impairment of multiple vestibular end-organs points to peripheral vestibulopathy of ischemic origin or an intralabyrinthine autoimmune process [1,3]. As in other manifestations of SSc, immunosuppressive therapy is effective in AIED when the disease phase consists mainly of inflammatory processes in contrast to the fibrotic and ischemic phases, which are generally irreversible.

The first line of treatment of AIED is high dose corticosteroids. Case reports have been published on the positive response to azathioprine, mycophenolate mofetil, cyclophosphamide, plasmapharesis and intratympanic infliximab. A randomized study found that methotrexate was no more effective than a placebo. Clinical trials with etanercept have yielded mixed results [5].

In conclusion, AIED might be a rare presenting symptom of SSc. Ear involvement is variable and initially can imitate known otoneurological entities including Meniere's disease. Thus, a high index of suspicion is recommended, including awareness of systemic autoimmune disease manifestations and immunological studies.

Correspondence
Dr J. Feld
Rheumatology Unit, Carmel Medical Center, Haifa 34362, Israel
Phone: (972-4) 825-0486
Fax: (972-4) 826-0213
email: joyfeld@gmail.com

References

1. Amor-Dorado JC, Arias-Nunez MC, Miranda-Filloy JA, Gonzalez-Juanatey C, Llorca J, Gonzalez-Gay MA. Audiovestibular manifestations in patients with limited systemic sclerosis and centromere protein-B (CENP-B) antibodies. *Medicine* (Baltimore) 2008; 87 (3): 131-41.
2. Monteiro TA, Christmann RB, Bonfá E, Bento RF, Novalo-Goto ES, Vasconcelos LG. Hearing loss in diffuse cutaneous systemic scleroderma. *Scand J Rheumatol* 2011; 40 (6): 467-71.
3. Amor-Dorado JC, Barreira-Fernandez MP, Arias-Nunez MC, Gomez-Acebo I, Llorca J, Gonzalez-Gay MA. Benign paroxysmal positional vertigo and clinical test of sensory interaction and balance in systemic sclerosis. *Otol Neurotol* 2008; 29 (8): 1155-61.
4. Deroee AF, Huang TC, Morita N, Hojjati M. Sudden hearing loss as the presenting symptom of systemic sclerosis. *Otol Neurotol* 2009; 30: 277-9.
5. Buniel MC, Geelan-Hansen K, Weber PC, Tuohy VK. Immunosuppressive therapy for autoimmune inner ear disease. *Immunotherapy* 2009; 1 (3): 425-34.