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ORIGINAL ARTICLE

Otolith function assessment in patients with systemic sclerosis

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Speech audiometry;
Tympanometry;
Ear

Abstract *Introduction:* Audiovestibular dysfunction has been reported in connective tissue disease patients. Evaluation of the otolith organ function as a part of the vestibular system of the inner ear in systemic sclerosis (SSc) patients is still greatly uncovered in the literature.

Aim of work: To assess otolith function in SSc patients and to correlate the findings with the clinical characteristics and disease severity.

Patients and methods: Thirty SSc patients and 30 matched controls were included in this work. All patients were subjected to full history taking, clinical examination, assessment disease severity by modified Rodnan skin score (mRss), relevant laboratory and radiological tests. All patients had otolith function assessment using Vestibular Evoked Myogenic Potentials (VEMP) and auditory assessment done using pure tone audiometry (PTA), speech audiometry and tympanometry.

Results: Auditory assessment revealed bilateral sensorineural hearing loss (SNHL) in 11 (36.6%) patients an abnormal VEMP response was found in 24 (80%). The mean latencies of P13 and N23 were significantly prolonged than in the controls ($p < 0.001$). Sine scleroderma subtype was significantly more frequent in SSc patients with normal VEMP ($p = 0.04$). Skin thickening was significantly more frequent in those with abnormal VEMP ($p = 0.04$) and the mRss score was also significantly increased ($p = 0.007$). Those with hearing loss were significantly older in age ($p = 0.03$) and had a significantly longer disease duration ($p = 0.01$) and had a significantly increased frequency of digital pitting ($p = 0.02$) and ulcers ($p = 0.03$) compared to those with normal hearing.

Conclusion: Our results showed an evidence of inner ear involvement in SSc causing audiovestibular abnormalities.

1. Introduction

Systemic sclerosis (SSc) is an autoimmune connective tissue disease (CTD) described for the first time in the middle of the 19th century [1]. The term systemic sclerosis is used to

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describe patients who have common manifestations that link them together, whereas a highly variable clinical course exists that spans from mild and subtle findings to aggressive, life-threatening multisystem disease [2].

Systemic sclerosis (SSc) is characterized by fibrosis; mainly in the skin; vasculopathy with Raynaud's phenomenon as an almost universal symptom as well as the presence of autoantibodies. Vasculopathy is a major factor in the symptoms and morbidity of SSc, probably occurring as the result of an autoimmune insult triggered by an as yet unknown factor that causes inflammatory processes and ultimately fibrosis with collagen deposition. The involvement of vessels characterized by intimal proliferation of the arteries that is ubiquitous in this disease and this vasculature involvement is fundamental to organ damage and subsequent malfunction [3]. Insufficient angiogenesis with tissue ischemia and accumulation of extracellular matrix represent the hallmarks of SSc disease [4]. There is some evidence of hearing loss in other autoimmune diseases, mainly caused by cochlear – vestibular abnormalities, and with vasculitis as the most accepted hypothesis [5].

The vestibular system comprises two types of sensory organs – the semicircular canals and the otolith organs (the saccule and utricle). The two otolith organs are positioned perpendicular to each other and sense linear acceleration, head tilt, and gravity, with the primary role of providing input to the vestibulospinal reflex for postural stability [6]. The vestibular sensory epithelium of the otolith organ is located on the maculae of the saccule and utricle. The sensory cells, hair cells of the maculae, are embedded in a gelatinous layer and above this is a fibrous structure, the otolithic membrane, in which are embedded crystals of calcium carbonate called otoconia [7]. These delicate structures are liable to fibrosis in SSc, which in turn may lead to otolith dysfunction.

Vestibular Evoked Myogenic Potentials (VEMPs) are short-latency electromyograms evoked by high-level acoustic stimuli recorded from surface electrodes over the tonically contracted sternocleidomastoid (SCM) muscle, as they are used to evaluate otolith function [8]. There are few studies in the literature regarding hearing disorders [9], and vestibular dysfunction [10–12] in SSc. However, there is no study that evaluated otolith organ function as a part of vestibular system.

The aim of our work is to evaluate otolith vestibular function in patients with SSc and to correlate these findings with age, disease duration and different clinical parameters among patients with SSC.

2. Patients and methods

The present study included two groups; patient group included thirty with SSc patients who fulfilled the *2013 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR)* criteria for the classification of SSc [13], recruited over a period of 1 year from the Rheumatology and Rehabilitation Outpatient Clinic and/or Department, Kasr El-Aini Hospital, Cairo University. The Control group was comprised of thirty age and sex matched healthy adults. The study was approved by the *Institutional Review Board (IRB)*, and all cases signed a written informed consent.

The SSc patients were further subgrouped according to the classification system proposed by Le Roy et al. into limited cutaneous SSc (lSSc) and diffuse cutaneous SSc (dSSc) [14]. Scleroderma patients with ear trauma, operations or infections, or use of ototoxic drugs as salicylates, non-steroidal anti-inflammatory drugs (NSAID), quinine, all were excluded from our study.

All patients have been subjected to:

I. *Comprehensive history taking* including personal, present, family and past history. Systemic manifestations and the history of drug intake were also recorded.
II. *Thorough clinical examination* including cutaneous, gastrointestinal, cardiopulmonary, neurological, rheumatological and otological examination (including otoscopic examination).

Tenderness score was recorded according to *Ritchie articular index* [15]. The skin thickness was scored according to the *modified Rodnan Skin Score (mRss)*, [16] in which the body was divided into 17 areas and the degree of thickness is graded at each site, scoring each area from 0 to 3 where (0) is normal skin, and (3) is extreme thickening. The sum of the score of all palpated sites is documented with a maximum possible score of 51.

III. *Investigations:*

A- *Laboratory investigations* included complete blood picture (CBC), erythrocyte sedimentation rate (ESR), liver and renal function tests, urine analysis, creatine phosphokinase (CPK), rheumatoid factor using the latex agglutination method, antinuclear antibodies (ANA) and its pattern.

B- *Radiological investigations* included plain X-ray chest for detection of pulmonary infiltrates, interstitial lung fibrosis and dilated pulmonary artery; high resolution computerized tomography (HRCT) chest for detection and description of interstitial pulmonary fibrosis; and Doppler echocardiography for cardiac assessment.

C- *Auditory investigations*: All the following measures were done by an expert Audiologist:

1- *Pure Tone Audiometry (PTA): Air conduction* (AC) on the better ear was started as the patient wears earphones attached to the audiometer. Pure tones of controlled intensity were delivered to one ear at a time. The duration of the test tone is usually kept constant at some 1.5 s: Start at 1000 Hz at 40 dB, down by 10 dB until no response and then up by 5 dB until there is a response. After the threshold for the 1 kHz-tone has been recorded by the audiologist; the entire procedure is repeated with a 2 kHz tone, and further increasing the tone's frequency to 4, 8 and 12 kHz. After the high-frequency range has been tested, the same procedure is performed for the lower frequencies, starting again with a 1 kHz-tone, but decreasing to 0.5, 0.25 kHz.

The same procedure was used *for bone conduction* with frequencies 0.5–4 kHz. The headband is fitted with small plastic rectangles that are placed behind the ears so as to channel the tones through the bones of the skull. The patient feels the vibrations of the tones as they are transmitted through the bones to the inner ear. Results were plotted in the audiogram showing hearing loss as a function of frequency. Hearing was scaled according to the average threshold level into normal hearing (0–25 dB) and hearing loss which could be mild