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Original Research Paper

Audiology

VITILIGO AND SENSORINEURAL HEARING LOSS: A SINGLE CASE STUDY

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ABSTRACT Vitiligo is a systemic idiopathic disease characterized by the presence of sharply demarcated, discoloured spots caused by epidermal melanocyte loss or damage. This disease affects all races equally regardless of sex, with an incidence of 1% to 2%. Recent clinical and experimental studies support the theory that pathogenetic mechanisms of vitiligo could be a systemic event, as vitiligo is associated with ocular and auditory abnormalities including sensorineural hearing loss (SNHL). **Case Report:** The current study describes a 24year old male adult who was brought with complaint of reduced hearing sensitivity in both the ears since 2 to 3 and who was also diagnosed with vitiligo. There was no significant prenatal, perinatal and postnatal medical history as well as family history reported. No history of any noise exposure or any long standing medication. Previous audiological evaluation reveals bilateral hearing sensitivity within the normal limits. **Conclusion:** The patient presented in this case study had gone through multiple tests including Behavioural, Physiological & Electrophysiological evaluation. All the evaluation leads to the conclusion that vitiligo does have an effect on the auditory system and can lead to sensorineural hearing loss.

KEYWORDS : vitiligo, pure tone audiometry, oto acoustic emissions, auditory brainstem responses

INTRODUCTION

Vitiligo is a systemic idiopathic disease characterized by the presence of sharply demarcated, discoloured spots caused by epidermal melanocyte loss or damage. This disease affects all races equally regardless of sex, with an incidence of 1% to 2%. Vitiligo usually begins with a few small white patches that may gradually spread over the body over the course of several months. Vitiligo typically begins on the hands, forearms, feet, and face but can develop on any part of the body, including the mucous membranes (moist lining of the mouth, nose, genital, and rectal areas), the eyes, and inner ears. Sometimes the larger patches continue to widen and spread, but usually they stay in the same place for years. Many types of vitiligo have been identified: localized, including focal and segmental, and generalized, including acrofacial, vulgaris, and universal (Czajkowski et al., 2004). The causes for vitiligo includes: Autoimmune disorder, genetic disorder, neurogenic factors and self-destruction.

Melanin And The Auditory System

Melanin is a substance present in the skin that produces pigment. Each person will have a different amount of melanin in their skin. This variation is due to genetics and other factors. *Melanocyte* is a cell (as of the skin, eye, or hair follicle) that produces melanin. Under the evolutionary viewpoint, the embryonic origin of human melanocytes are from the neural crest, and they are located in the epidermis, hair bulbs of the skin, the uveal tract, retinal pigment epithelium of the eyes, leptomeninges, and the inner ear (Aydogan et al., 2006). The presence of otic melanocytes was first described by Alphonse Corti (1831). (Angrisani et al., 2009) these cells are primarily localized throughout the stria vascularis and modiolus of the cochlea, but they also exist in the vestibular organs (Nordlund et al.,2006) & (Shajil et al., 2006). Melanocytes may have an important role in the inner ear since hearing is affected in systemic disorders that affect pigmented areas such as the Vogt-Koyanagi and Waardenburg syndromes (Aydogan et al., 2006). Ardic et al., (1998) Melanin is believed to have several roles in the inner ear. It can bind to ototoxic drugs and inhibit their adverse effects on the cochlea. (Conlee, Bennett & Creel, 1995) have also reported that melanocytes of the inner ear can protect the cochlea against various stresses, especially loud noise. Melanocytes are also essential for creating endolymphatic potential, which is very important for cochlear hair cell function and normal hearing (Nin et al., 2016). The presence of melanocytes is not limited to the peripheral auditory system, abnormalities in the brainstem were found in both animals and humans with pigment disorders; additionally, neurons in the medial superior olivary nucleus of albino rabbits were shown to be 24% smaller than normal animals, and the branching density of its dendrites was significantly reduced (Ardic et al., 1998), so melanocytes are also present in the central auditory system.

Recent clinical and experimental studies support the theory that pathogenetic mechanisms of vitiligo could be a systemic event, as vitiligo is associated with ocular and auditory abnormalities including sensorineural hearing loss (SNHL).There are many supporting studies as which says that melanin deficiency can lead to sensorineural hearing loss (Fleissig et al.,2013) & (Dabbous, Medhat & Mesidy, 2020). Some reports, however, do not support a connection between hearing loss and vitiligo (Ozuer et al., 1998) & (Escalante et al., 1991). Since there were some discrepancies in the literature about the influence of vitiligo on auditory functions aim of the present study was to use multiple audiological tests including Behavioural, Physiological and Electrophysiological evaluation to analyse the connection between vitiligo and sensorineural hearing loss.

Case Report

A 24 year old adult male was brought to the clinic with a complaint of reduced hearing sensitivity in both the ears since 2-3 years. There was no significant prenatal, perinatal and postnatal medical history as well as family history reported. No history of any noise exposure or any long standing medication. The earlier audiological evaluation findings were as follows:

1) Pure tone audiometry – Bilateral hearing sensitivity within the normal limits
2) Speech audiometry – Speech reception threshold (SRT) and Speech discrimination scores (SDS)
3) Tympanometry and Reflexometry – Bilateral 'A' type tympanogram with both ipsilateral and contralateral reflexes present at 500Hz, 1000 Hz and 2000Hz suggestive of no middle ear pathology.
4) Oto acoustic emissions – Bilateral DPOAE's present suggestive of normal outer hair cell function.

Ear	SRT	SDS
Right ear	25dBHL	100%
Left ear	25dBHL	100%

Method

A detailed case history was obtained followed by which Otoscopic evaluation to visualize the external auditory canal and tympanic membrane status. All the audiological evaluations were carried out in a sound treated room. Pure tone audiometry was carried out with MAICO MA42 dual channel audiometer. Both air conduction thresholds and bone-conduction thresholds were measured. The thresholds (minimum level of hearing) for air conduction were estimated using standard headphone TDH39 (770 Park Ave, Huntington, NY11743, US), from frequency range 250Hz to 8 KHz, with intensity level -10 dBHL to 120 dBHL, and the pure tone average was measured using an average of three frequencies, that is, 500Hz, 1 KHz, and 2 KHz. Bone conduction was tested with test frequencies from 250Hz to 4 KHz, with intensity level from -10 dBHL to 70 dBHL (with standard bone conductor B71, Radio ear). Speech audiometry was performed, including speech reception threshold using spondee words, and speech discrimination score, using phonetically balanced words.

Tympanogram and Acoustic reflex thresholds were assessed using GSI Tympstar instrument. Tympanogram was measured using a 226 Hz probe tone, with sweep pressure starts point at -200 dapa to end point of +400 dapa. Ipsilateral and Contralateral acoustic reflex were obtained using pure tone activator stimuli 500 Hz, 1 kHz, 2 kHz and 4 kHz at 90dBHL, 100dBHL and 105dBHL. The patient was instructed to sit quiet and not to move during the procedure. Distortion product Oto acoustic emissions (DPOAEs) were recorded using IHS Jr instrument to check to outer hair cell (OHC) function. Tone pair was sequential, with an F2/F1 ratio of 1.2. Intensity of the tone pairs was 65 and 55 dB SPL for L1 and L2, respectively. Two separate runs per ear were collected for determining repeatability. Validity and reliability of normal outer hair cell function was determined by analysing each distortion product frequency separately. Passing criteria for DPOAE was 6 dB SNR. ABR was measured using IHS Jr. ABR Instrument. Three electrodes were placed on the vertex (non-inverted), the ipsilateral mastoid (inverted), and the contralateral mastoid (ground). Inter electrode impedance was kept below 5kΩ. The acoustic stimuli were rarefaction clicks with 0.1 ms duration that were delivered monaurally through ER-3A insert earphones presented at a repetition rate of 31.1/sec. Responses to 2048 clicks were preamplified and band pass between 100-3000 Hz. The analysis time of the screen was set 12 ms. The electrode impedance was regularly monitored. Recording was obtained in a sound proof condition. Duplicate recordings were made to check reproducibility. Absolute latency values of waves I, III and V and interpeak latencies (IPL) of I-III, III-V, and I-V, were measured.

Dermatological evaluation by a specialized dermatologist included the following: detailed dermatological history from the patients, including family history of vitiligo, type, site, duration, onset and course of vitiligo. Assessment of vitiligo were carried out by vitiligo area severity index (VASI) (Hamzavi et al., 2004) and vitiligo disease activity score (VIDA) (Njoo et al., 1999).

Vitiligo Area Severity Index:

The percentage of vitiligo involvement is calculated in terms of hand units. One hand unit (which encompasses the palm plus the volar surface of all digits) is approximately equivalent to 1% of the total body surface area. The degree of pigmentation is estimated to the nearest of one of the following percentages: 100% - complete depigmentation, no pigment is present; 90% - specks of pigment present; 75% - depigmented area exceeds the pigmented area; 50% - pigmented and depigmented areas are equal; 25% - pigmented area exceeds depigmented area; and 10% - only specks of depigmentation present.

Vitiligo Disease Activity Score:

VIDA score is a six-point scale for evaluatingvitiligo activity. Individuals own opinion is the base in VIDA score. VIDA score based on patients' opinion divided in 6 stages. Lower VIDA scores indicate less activity (Njoo et al., 1999). Grading is as follows:

+4: Activity of 6 weeks or less period,
+3: Activity of 6 weeks to 3 months
+2: Activity of 3 to 6 months,
+1: Activity of 6 to12 months
0: Stable at least for 1 year and,
-1:Stable at least for 1 year with spontaneous repigmentation.

RESULTS

Otoscopic Examination:

Otoscopic examination revealed bilateral cone of light present with no other abnormalities present for the ear canal and tympanic membrane.

Pure Tone Audiometry:

Pure tone audiometry revealed bilateral mild to moderate sensorineural hearing loss (Figure.1).

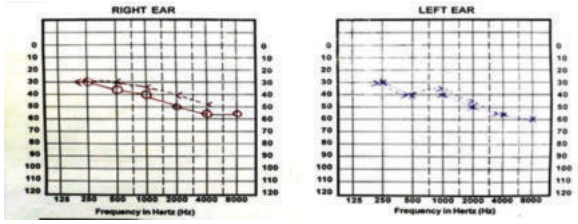

Figure1. Pure Tone Audiometry Findings

Speech Audiometry:

SRT and SDS results are as follows (Figure.2)

Ear	SRT	SDS
Right ear	55dBHL	85%
Left ear	55dBHL	80%

Figure.2 Speech Audiometry Results

Immittance And Reflexometry:

Bilateral 'A' type tympanogram with ipsilateral reflex present at 500Hz and absent contralateral relexes suggestive of no middle ear pathology (Figure.3).

Measures	Right	Left
Tympanogram type	A	A
Ear canal volume	1.6 cc	1.8 cc
Static compliance	1.5 ml	1.7 ml
Peak pressure	10 dapa	9 dapa

Figure.3 Immittance And Reflexometry Results

Oto Acoustic Emission:

Distortion product oto acoustic emission revealed bilateral absent oae's suggestive of outer hair cell dysunction (Figure.4).

Auditory Brainstem Responses:

Revealed bilateral clear and replicable Vth peak obtained till

60dBnHL which leads to the impression mild to moderate hearing loss(Figure.5). At higher intensities the absolute latencies of wave V was prolonged for both the ears as well the inter peak latencies I-III, III-V and I-V were also found to be prolonged bilaterally.

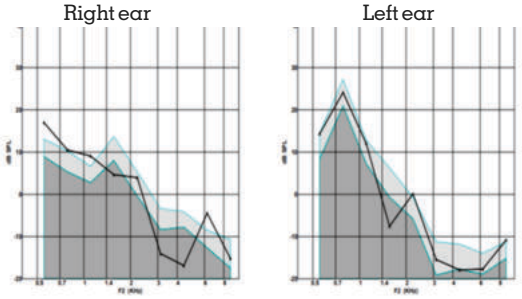

Figure.4 DPOAE Findings Of Right Ear And Left Ear Respectively.

	Frequency (Hz)			
	500	**1000**	**2000**	**4000**
Right ipsi	105	NR	NR	NR
Right contra	NR	NR	NR	NR
Left ipsi	105	NR	NR	NR
Left contra	NR	NR	NR	NR

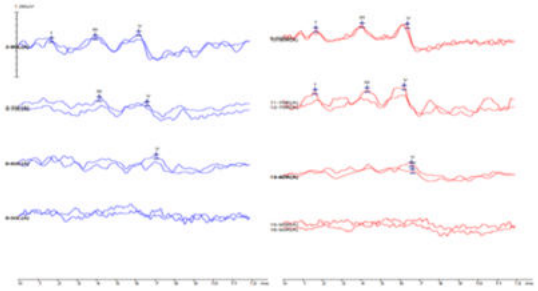

Figure.5 ABR Waveforms Of Both Right And Left Ears

The Dermatological evaluation revealed the following findings:

1) Vitiligo area severity index (VASI): Score of -50 - ~25 (Much worse)

2) Vitiligo disease activity score (VIDA): A score of +2 (Active in past 6 months)

DISCUSSION

Vitiligo is a common, chronic, depigmenting skin disease constituted by the destruction of melanocytes (epidermis, the mucous membranes, eyes, and in some hair bulbs) (Shajil et al., 2006). Moreover, involvement of leptomeninges, retinal pigment epithelium, uveal tract, and inner ear melanocytes is reported. An association between vitiligo and ocular, hearing, and autoimmune pathologies is reported in the literature (Dawoud et al., 2017) & (De jong, Adelman & Gross, 2017). There are discrepancies in the literature about the effect of melanin on auditory system. Some authors state that melanin influence hearing whereas others question such influence. It is hypothesized that in vitiligo patients, there occurs a synchronous loss of melanocytes, both in skin and in inner ear, with the resultant loss of their homeostatic and protector function, and increased vulnerability of inner ear to various damaging forces (Hilding & Ginzberg, 1977). (Garber et al., 1982) have reported and association of reduced cochlear melanin level with increased susceptibility to noise-induced hearing loss, audiogenic seizures and increased levels of auditory fatigue.

In the present study PTA showed mild to moderate sensorineural hearing loss which is comparable with Mahdi et al., (2012) who also found that patients with vitiligo had lowered hearing thresholds starting from 2 to 8 kHz compared with controls. This was explained by possible alteration of the inner ear pigment cells that lead to sensorineural hearing loss. Similarly, Fleissig et al., (2013) reported incidence of mild and minimal sensorineural hearing loss in individuals affected with vitiligo. Dawoud et al., (2017) in their study also revealed that 23/30 (77%) of vitiligo patients had normal PTA thresholds and 7/30 (23%) patients had mild to moderate SNHL.

DPOAE evaluation revealed bilateral absence of OAEs which is well correlated with Anbar et al., (2015) study in which they examined 53 patients with vitiligo and normal audiograms, 60% of patients had cochlear dysfunction based on DPOAEs, often bilateral, while their control group had no abnormality. The presence of OAEs is a reliable indicator of structural integrity of the outer hair cells (OHCs) in the cochlea, and their absence might reveal possible subclinical OHC damage corresponding to a particular conventional frequency region (i.e. a cochlear lesion before any evidence occurs in pure tone audiometry).

Auditory brainstem response revealed prolongation of absolute latencies of wave III and V in both the ears, as well as the inter peak latencies I-III, III-V and I-V bilaterally. These findings were well supported by Koura et al., (2018) who found statistically significant prolonged wave III, wave V, and interpeak latencies of I–III and I–V latencies of BAEP findings in both ears of 18 (60%) of 30 patients and in the left ear of two (6.6%) patients in the vitiligo group compared with the control group. Aydogan et al., (2006) proposed that the delay of wave III, wave V, and interpeaks of I–V, I–III, and III–V of ABR in patients with vitiligo may refer to the pathology of the superior olivary nucleus and upper brainstem or inferior colliculus and to abnormal synaptic activity and a delayed synchronization of the action potential along the pathway from the auditory nerve to the cochlear nucleus and from the cochlear nucleus to the superior olivary nucleus and inferior colliculus (Ozuer et al.,1998).

CONCLUSION

The present study revealed that melanin may have an important role in the maintenance of the function of the auditory system and also that vitiligo patients require routine monitoring for auditory functions for early identification of SNHL. Both Older and younger subjects with vitiligo might be at a higher risk for audiological abnormalities. These patients should also be informed regarding the associated risk with noise and ototoxic drug exposure. Also the degree of hearing loss can vary according to the severity of vitiligo.

In summary, audiological evaluation of patients with vitiligo should include audiometry and very high-frequency audiometry, OAE and ABR measurements. Incomplete evaluation might result in under detection of hearing loss. Additionally, patients should take extra precautions to prevent acoustic trauma and avoid ototoxic drugs, especially during active disease, as their auditory system may be less protected due to non-functional melanocytes. To fully understand the mechanism by which melanocytes function in relation to hearing, we recommend further research among vitiligo patients to gain a better understanding of the audiology functions and cell mechanisms in the absence of functional melanocytes in the ear.

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