Content available at: https://www.ipinnovative.com/open-access-journals

IP Journal of Otorhinolaryngology and Allied Science

Journal homepage: https://www.joas.co.in/

Case Report

An audiological profile in vogt-koyanagi-harada syndrome

Aksha Dayal[1], Imran Ansari[1], Debadatta Mahallik[1,*], Hansa Banjara[1]

[1] *Dept. of Otorhinolaryngology and B.ASLP Course, Pt. Jawahar Lal Nehru Memorial Medical College, Raipur, Chhattisgarh, India*

ARTICLE INFO

Article history:
Received 20-09-2022
Accepted 27-12-2022
Available online 04-01-2023

Keywords:
Otoacoustic emission
Auditory Brain stem response
VKH syndrome

ABSTRACT

The Vogt-Koyanagi-Harada syndrome (VKHS) is a rare, multisystemic, granulomatous inflammatory and autoimmune disease that affects the pigmented structures such as eyes, ears, skin, and hair. The syndrome mainly affects the eyes, followed by bilateral chronic panuveitis. A 34 years old female came with complaints of reduced vision and reduced hearing sensitivity to our hospital. She was diagnosed with VKHS in the ophthalmology department. Throughout the audiologist assessment, she presented bilateral sensorineural hearing loss, absent otoacoustic evoked emissions, and complaints about postural vertigo and acute tinnitus. The specific case reported presented sudden sensorineural hearing loss, vertigo, tinnitus, and bilateral ocular disease. Even though drug treatment was performed, the hearing loss remained.

This is an Open Access (OA) journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprint@ipinnovative.com

1. Introduction

The Vogt-Koyanagi-Harada Syndrome is an uncommon disease which affects tissues with high melanocyte counts like the skin, inner ear, meninges, and eyes, is likely having an autoimmune and multisystemic etiology.[1] This disease has a characteristic feature of bilateral, chronic, diffuse panuveitis associated with characteristic neurological, auditory and integumentary features.[2] Ali Ibn Isa, an Arabic Doctor was the first to publish the earliest reports of the disease in 12^{th} century.[3] In 1906, Alfred Vogt,[4] a Swiss ophthalmologist, while he was still a resident at Basel University Hospital, documented a case report of a 16-year-old patient having iridocyclitis and poliosis. Yoshizo Koyanagi[5] in 1929, described 16 cases with similar features, including 6 new cases. Among the abnormalities he listed were idiopathic bilateral anterior uveitis, dysacusis, vitiligo, polio, and alopecia, as well as a prodromal phase of headache, fever, and disorientation. He also postulated that an "anaphylactoid" reaction against pigment constituted the root of the disease. In 1926, Einosuke Harada[6] characterised a condition with a prodromal period of malaise and meningeal irritation, bilateral uveitis, bilateral spontaneously resolving retinal detachments, lymphocytosis of the cerebrospinal fluid, and dysacousia. In 1932, Babel from the Department of Opthalmology at Geneva University Hospital admitted that Vogt-Koyanagi Syndrome is identical to the Harada's description of the condition. The cases described by Vogt, Koyanagi, and Harada are today recognised as a single syndrome with a broad range of clinical variants and the name was combined into VKHD Syndrome.[7]

2. Geographical Distribution

Darkly pigmented people are more likely to develop the condition. The most commonly impacted groups are Asians, Native Americans, and Hispanics. In Caucasians, it is unusual. Black people with sub-Saharan African ancestry are disproportionately impacted, showing that the level of

* *Corresponding author.*
E-mail address: debadatta.mahallik@gmail.com (D. Mahallik).

https://doi.org/10.18231/j.ijoas.2022.033
2582-4147/© 2022 Innovative Publication, All rights reserved.

skin pigmentation is not the only aetiologic element in the pathophysiology of VKH disease.[8–10]

2.1. Prevalence

VKHS is most common in individuals between the ages of 20 and 50, yet instances have been observed in children as young as 4 years old, and its advancement is more abrupt in these circumstances.[11,12] With a 2:1 male to female ratio, women are typically more affected in comparison to men.[13]

In India, with a prevalence rate of 21.08 percent, VKH is the common cause of panuveitis. In a retrospective study done between January 1992 and December 1994, J Biswas, S Narain, D Das and S.K. Ganesh found that Uveitis is less common in children below 10 years (3.61%) and in adults over 60 years of age (6.44%), whereas it is majorly affecting the patients in their forties (23.57%). In this study they also found that men were predominantly more affected as compared to women, whereas in other similar studies women were more affected. They hypothesized that such discrepancy is due to the reality that in developing countries like India, men are economically active hence are more likely to get medical assistance as compared to women.[14]

2.2. Etiology

According to immunological and histopathological studies, VKH is an autoimmune inflammatory condition mediated by T cells that target melanocytes.[15,16] These activated T cells likely initiate the inflammatory process through the generation of cytokines, IL 17 and IL 23, in individuals with altered tolerance to melanocytes from deficient T regulatory cells.[17]

3. Stages of Progression

There are four stages to the VKH Syndrome:

1. *Stage: - Prodromal Phase,* this phase is seen a few days before the ocular inflammation and lasts for up to two weeks. The patient may exhibit neurological symptoms of ophthalmological problems, such as photophobia, headaches, and fever.
2. *Stage: - Acute Uveitis Phase,* which is primarily characterised by bilateral visual turbidity, bilateral uveitis, ocular pain, photophobia, conjunctival hyperemia, disacusia, and serous multifocal retinal displacement.
3. Stage: - *Chronic/convalescent Phase,* depigmentation of the melanocyte tissue is the characteristic feature of this stage.
4. Stage: - *Chronic recurrent phase:* with less uveal thickening than the acute phase, this stage is characterised by recurrent granulomatous anterior uveitis and choroidal thickening.[18]

3.1. Diagnostic criteria

In 1978, during the annual meeting of the American Uveitis Society, first attempt was made to formulate diagnostic criteria for VKH Syndrome. In order to meet these requirements, a patient had to have no prior history of ocular trauma or surgery as well as one of the following signs from at least three of the following categories: bilateral chronic iridocyclitis; posterior uveitis, including exudative retinal detachment, forme fruste of exudative retinal detachment (optic disk hyperemia or edema, “subretinal macular edema”), or “sunset glow” fundus; neurologic symptoms or signs of tinnitus, neck stiffness, cranial nerve or central nervous system problem, or cerebrospinal fluid pleocytosis; and cutaneous finding, such as alopecia, poliosis, or vitiligo. These criteria were not apt for the diagnosis of the disease in the initial stage as the criteria includes only the chronic features of the disease. Hence a new diagnostic criterion was formulated based on the concept of Babel that VKH Syndrome is a single entity with quite distinct clinical features depending upon the stage at which the patient is examined. Regardless of the stage at which the patient is examined, and despite the variations in the clinical manifestation of the disease, it is termed as VKH Syndrome, even though all the possible signs of the disease are not manifested. Due to this, the criteria are made to enable the classification of Vogt-Koyanagi-Harada syndrome into "complete" and "incomplete" groups based on the range of symptoms present. The new recommended criteria provide the designation of “Probable” when the diagnosis is questionable but the findings are strongly suggestive.[18]

VKH Disease is such a condition that affects multiple system of the body. Hence for the accurate diagnosis of the Disease, involvement of the other system other than ocular system is necessary.[18] The extraocular manifestation of the disease includes:

1. Neurological Findings include meningismus, headache, fever, cerebrospinal pleocytosis, nausea abdominal pain, stiffness of the neck and back, or a combination of these features.[19]
2. Integumentary Findings – It includes depigmentation of choroid, eyebrows, eyelashes, hair, and skin resulting in poliosis, vitiligo, and alopecia.[20,21]
3. Audiological findings include hearing loss, tinnitus, and vertigo.[22]

4. Case Report

4.1. Clinical case presentation

The present case was performed at Pt. J.N.M Medical College, Raipur Chhattisgarh after the participant’s consent, gave permission to use the data of her medical record in the present study. Following are the data obtained:

4.2. Patient's case history

A 35 years old lady referred from the Department of Opthalmology for hearing evaluation to our department. She was admitted in our hospital for chronic uveitis and was diagnosed with VKH Syndrome based on the American Uveitis Society Diagnostic Criteria.

Audiological history includes complaints of sudden hearing loss with no conductive history. the patient also had the complaint of subjective type vertigo with tinnitus in both ears. She had difficulty hearing during a telephone call. She faced difficulty in understanding speech in a noisy environment.

5. Results

5.1. Pure tone audiometry

Audiometry was performed using an inter-acoustic AC40 audiometer. The PTA for each ear is determined separately using circumaural headphone TDH 39 at frequencies of 250, 500, 1000, 2000, 4000, and 8000 Hz for Air Conduction, and at 250, 500, 1000, 2000, and 4000 Hz for Bone Conduction using B71 bone vibrator, with masking where needed. The PTA measured using a 4-frequency average, showed bilateral mild to moderate sensorineural hearing loss.

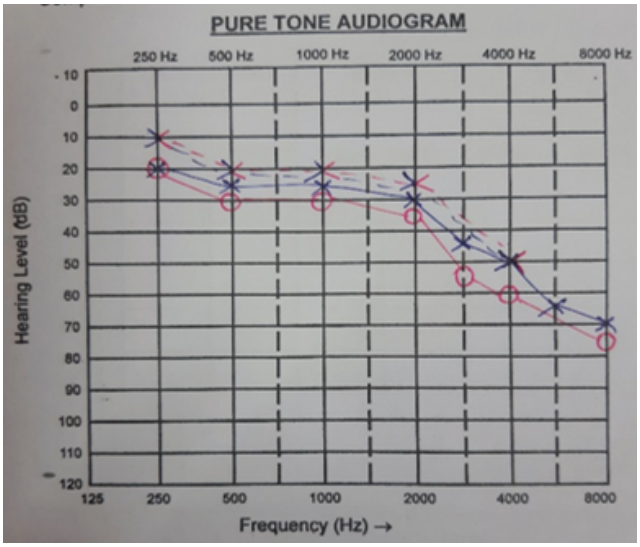

Fig. 1: -Hearing threshold of both ears

5.2. Immittance acoustic measures

Impedence Audiometry was done using Interacoustic tympstar in which acoustic probe tip were inserted inside the ear getting the hermatic seal. Findings were "bilateral A type tympanogram with bilateral absence of acoustic reflex" (Figure 1).

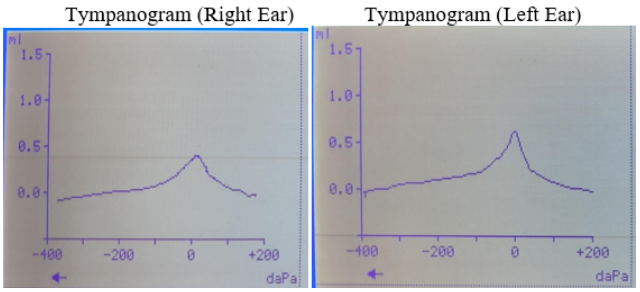

Fig. 2: Tympanogram (Right Ear), Tympanogram (Left Ear)

5.3. Oto-acoustic Emission (OAE)

DPOAE done using GSI-Audera. An acoustic probe enclosed in a soft rubber covering and containing a miniature microphone and a speaker was placed in the distal portion of the external auditory canal. The stimuli applied were a 65-dB sound pressure level (dB SPL) tone of f 1 and a 55-dB SPL tone of f 2 (f 2 > f 1; f 2 /f 1 = 1.22). The DPOAE response levels were measured at the frequency 2f 1 –f 2. The noise floor levels were also calculated at each frequency. DPOAE were recorded for 3 frequency pairs at 2,000, 3,000 and 4,000 Hz. Findings were "bilateral DPOAE refer(absent) suggestive of impaired functioning of the outer hair cells".

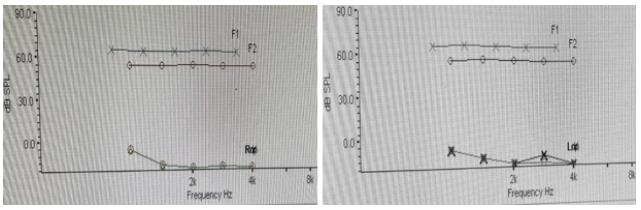

Fig. 3: - DP-Gram of both ears.

5.4. Auditory brainstem response (ABR)

ABR has done using GSI-Audera using conventional montage with insert earphones in which findings were, "bilateral replicable wave V could be traced till 50dB nHL suggestive of Bilateral Mild Hearing Loss."

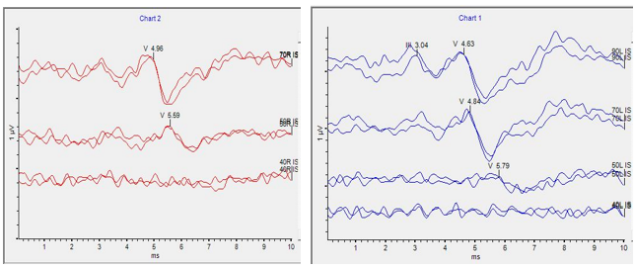

Fig. 4: ABR wave form of both ears.

6. Discussion

Anterior uveitis, exudative retinal detachment, depigmented fundal lesions, as well as the presence of neurological, auditory, or dermatologic manifestations, are clinical characteristics of VKH Syndrome. Ocular and extraocular signs and symptoms are needed for the diagnosis of VKH Syndrome.[19]

The present study is supported by the previous studies on VKH Syndrome which states that the VKH syndrome is characterized with ocular disease and associated with hearing loss[2,18,19]. The present patient, showed bilateral mild sensori-neural hearing loss, supported to previous study done by Dousary Al S[23]. The hearing loss was sensori-neural in nature, there is no involvement of middle ear conductive pathology, as confirmed from impedance findings Fig 2. Absence of Bilateral DPOAE, presence of bilateral wave V till 50 dB nHL in ABR is correlating with audiometric findings, suggests bilateral mild to moderate SN hearing loss. Presently she is under treatment of with corticosteroids and immunosuppressant for reducing the inflammation caused by the syndrome. A study revealed that the outcome of the syndrome is immensely affected by the time period between hearing loss and the initiation of the treatment. The hearing impairment is resolved if the treatment is started just after the hearing loss[23]. Although there is no improvement if the treatment is started after several weeks of hearing loss. Thus, there is no hearing restoration prognosis if the impairment of the inner ear was severe, such as in profound hearing loss.

The case described in the present study presented sudden bilateral sensorineural hearing loss, tinnitus, vertigo and bilateral ocular involvement. Hence early diagnosis and intervention could help in a better hearing prognosis.

7. Source of Funding

None.

8. Conflict of Interest

None.

References

1. Damico FM, Kiss S, Young LH. Vogt-Koyanagi-Harada Disease. *Seminar Ophthalmol*. 2005;20(3):183–90.
2. Greco A, Fusconi M, Gallo A, Turchetta R, Marinelli C, Macri GF. Vogt-Koyanagi-Harada Syndrome. *Autoimmunity Rev*. 2013;12(11). Available from: https://rarediseases.org/rare-diseases/vogt-koyanagi-harada-disease/.
3. Pattison EM. Uveomeningoencephalitic syndrome (Vogt-Koyanagi- Harada). *Arch Neurol*. 1965;12:197–205. doi:10.1001/archneur.1965.00460260087010.
4. Vogt A. Frühzeitiges Ergrauen der Zilien und Bemerkungen über den sogenannten plötzlichen Eintritt dieser Veränderung. Early graying of cilia and remarks about the so-called sudden occurrence of these change. *Klin Monatsbl Augenheilkd*. 1906;44:228–42.
5. Koyanagi YD. Alopecia und Poliosis bei schwerer Uveitis nicht traumatischen Ursprungs. Dysacusis, alopecia and poliosis in severe uveitis without traumatic origin. *Klin Monatsbl Augenheilkd*. 1929;82:194–211.
6. Harada E. Clinical study of nonsuppurative choroiditis: a report of acute diffuse choroiditis. *Acta Soc Opthalmol Jpn*. 1926;30:356–78.
7. Herbort CP, Mochizuki M. Vogt-Koyanagi-Harada disease: inquiry into the genesis of a disease name in the historical context of Switzerland and Japan. *Int Ophthalmol*. 2007;27(2-3):67–79.
8. Yang P, Ren Y, Li B, Fang W, Meng Q, Kijlstra A. Clinical characteristics of Vogt- Koyanagi-Harada syndrome in Chinese patients. *Ophthalmology*. 2007;114(3):606–20.
9. Rajendram R, Evans M, Rao NA. Vogt-Koyanagi-Harada disease. *Int Ophthalmol Clin*. 2005;45(1):115–49.
10. Sukavatcharin S, Tsai JH, Rao NA. Vogt-Koyanagi-Harada disease in Hispanic patients. *Int Ophthalmol*. 2007;27(2-3):143–51.
11. Ondrey FG, Moldestad E, Mastroianni MA, Pikus A, Sklare D, Vernon E. Sensorineural hearing loss in Vogt-Koyanagi-Harada syndrome. *Laryngoscope*. 2006;116(10):1873–9.
12. Berker N, Ozdamar Y, Soykan E, Ozdal P, Ozkan SS. Vogt-Koyanagi-Harada syndrome in children: report of a case and review of the literature. *Ocul Immunol Inflamm*. 2007;15(4):351–8.
13. Tabbara KF, Chavis PS, Freeman WR. Vogt-Koyanagi-Harada syndrome in children compared to adults. *Acta Ophthalmol Scand*. 1998;76(6):723–9.
14. Biswas J, Narain S, Das D. Pattern of uveitis in a referral uveitis clinic in India. *Int Ophthalmol*. 1996;20(4):223–8.
15. Norose K, Yano A. Melanoma specific Th1 cytotoxic T lymphocyte lines in Vogt-Koyanagi-Harada disease. *Br J Ophthalmol*. 1996;80(11):1002–8.
16. Sugita S, Sagawa K, Mochizuki M. Melanocyte lysis by cytotoxic T lymphocytes recognizing the MART-1 melanoma antigen in HLA-A2 patients with Vogt-Koyanagi-Harada disease. *Int Immunol*. 1996;8(5):799–803.
17. Chen L, Yang P, Zhou H. Diminished frequency and function of CD4+CD25high regulatory T cells associated with active uveitis in VogtKoyanagi-Harada syndrome. *Invest Ophthalmol Vis Sci*. 2008;49(8):3475–82.
18. Read RW, Holland GN, Rao NA. Revised diagnostic criteria for Vogt-Koyanagi-Harada disease: report of an international committee on nomenclature. *Am J Ophthalmol*. 2001;131(5):647–52.
19. Sugiura S. Vogt-Koyanagi-Harada disease. *Jpn J Ophthalmol*. 1978;p. 1127. doi:10.1007/978-3-642-88589-1_229.
20. Goto H, Rao NA. Sympathetic ophthalmia and Vogt-Koyanagi-Harada syndrome. *Int Ophthalmol Clin*. 1990;30(4):279–85.
21. Beniz J, Forster DJ, Lean JS. Variations in clinical features of the VogtKoyanagi-Harada syndrome. *Retina*. 1991;11(3):275–280.
22. Dousary SA. Auditory and vestibular manifestations of Vogt-Koyanagi-Harada disease. *J Laryngol Otol*. 2011;125(2):138–41.

Author biography

Aksha Dayal, Student

Imran Ansari, Audiologist https://orcid.org/0000-0001-8509-850X

Debadatta Mahallik, Audiologist https://orcid.org/0000-0002-4889-7131

Hansa Banjara, Professor and Head

Cite this article: Dayal A, Ansari I, Mahallik D, Banjara H. An audiological profile in vogt-koyanagi-harada syndrome. *IP J Otorhinolaryngol Allied Sci* 2022;5(4):141-144.