CoDAS
ISSN 2317-1782 (Online version)

Case Report
Relato de Caso

Maria Carolina Ferreira[1]
Izabella Lima de Matos[1]
Maria Fernanda Capoani Garcia Mondelli[1]

Audiological findings in the Vogt-Koyanagi-Harada Syndrome

Achados audiológicos na Síndrome de Vogt-Koyanagi-Harada

Keywords

Uveomeningoencephalitic Syndrome
Uveitis
Hearing
Hearing Loss
Melanocytes

ABSTRACT

The Vogt-Koyanagi-Harada syndrome (VKHS) is a rare, multisystemic and autoimmune disease. The syndrome mainly affects the eyes, followed by bilateral chronic panuveitis, however, the syndrome may also affect the melanocytes tissues, for example, the eyes, inner ear, meninges and skin. The syndrome origin mechanism is not yet completely known. Commonly, the specific ethnic groups that are affected by the VKHS are as follows: Hispanics, Asians, Indians, Native Americans and ethnic groups from the Middle East. The audiological characteristics of the syndrome and the possible audiologist interventions for a specific case will be reported. The patient was attended at the Clinic of Speech Therapy, Faculdade de Odontologia de Bauru (FOB). She is 53 years old and presented audiological complaints. She was diagnosed with VKHS by a specialist doctor. Throughout the audiologist assessment, she presented bilateral sensorineural hearing loss, absent otoacoustic evoked emissions, 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. Therefore, before the hearing aid (HA) fitting, the audiologist should perform the hearing management, investigate if the patient takes the drug treatment and the occurrence of suggestive symptoms of the syndrome. These are some points that help in the reference to the specialist doctor and the audiologist strongly participates in what concerns the hearing.

Palavras-chave

Síndrome Uveomeningoencefálica
Uveíte
Audição
Perda Auditiva
Melanócitos

RESUMO

A síndrome de Vogt-Koyanagi-Harada (SVKH) é rara, multissistêmica e autoimune. Atinge principalmente os olhos, provocando uma panuveíte crônica bilateral, porém traz afecções em outras áreas e tecidos que são ricos em melanócitos, como olhos, orelha interna, meninges e a pele. Sua origem ainda não é totalmente conhecida. Geralmente, a SVKH atinge indivíduos de origem hispânica, do Oriente Médio, indianos, nativos americanos e asiáticos. Descrição dos aspectos audiológicos acometidos pela síndrome e as possíveis intervenções fonoaudiológicas para um caso específico. Paciente de 53 anos, sexo feminino, compareceu à Clínica de Fonoaudiologia, Faculdade de Odontologia de Bauru (FOB) com queixas audiológicas e diagnóstico médico da SVKH. A paciente apresentou perda auditiva sensório-neural bilateralmente, emissões otoacústicas evocadas ausentes e queixas vestibulares de vertigem postural e desequilíbrio ao andar, bem como queixa de zumbido agudo contínuo. O caso apresentado mostrou perda auditiva sensório-neural, vertigem, zumbido e acometimento ocular bilateral. Apesar do tratamento com corticoesteroide, a perda auditiva se manteve. Desta forma, precedente à indicação do AASI, o fonoaudiólogo deve atentar-se para o acompanhamento audiológico do caso, realização ou não de tratamento medicamentoso e ocorrência de sintomas sugestivos da síndrome, favorecendo o encaminhamento para o médico e participando ativamente do processo terapêutico envolvendo a audição.

Correspondence address:
Maria Carolina Ferreira – Alameda Doutor Octávio Pinheiro Brisolla 6-65, Jd. Pagani, Edifice Tapajós Apartment 51. Bauru – São Paulo – Brazil – 17012059
Telephone: +55 (49) 999495002
E-mail: mariaferreira@usp.br / mariacarol.mcf@gmail.com

Received: July 19, 2019.

Accepted: October 28, 2019.

Study conducted at the Speech Therapist Department, Faculdade de Odontologia de Bauru (FOB)- Universidade de São Paulo, Bauru, SP, Brazil.
[1] Faculdade de Odontologia de Bauru (FOB)- Universidade de São Paulo, Bauru, SP, Brazil.

Conflict of interests: none.
Funding source: Fundação de Amparo à Pesquisa do Estado de São Paulo (FAPESP – 2018/16349-6).

INTRODUCTION

Vogt-Koyanagi-Harada syndrome (VKHS) is a rare disease, probably with an autoimmune and multisystemic etiology which reaches the tissues rich in melanocytes such as the eyes, inner ear, meninges and, the skin[1]. Commonly, the disease causes a chronic bilateral panuveitis and the eyes are the most affected. The visual consequences of the disease show up associated with other neurological, audiological and cutaneous disorders[2]. The disease etiology and pathogenesis is not yet totally known. It has been supposed that a central component could lead to a systemic disorder. The infectious and autoimmune basis could also be explained by the inflammations and destruction of melanocytes found in a range of tissues[3]. A previous study in the topic of otorhinolaryngology showed that the autoimmune system could damage the inner ear melanocytes and could cause auditory and balance dysfunctions[3,4].

The first descriptions about the disease are from the XXI century and were made by an Arabic doctor, Ali Ibn Isa[2]. Some cases described in that moment referred to different diseases, however, other cases exclusively referred to the VKHS[3]. In the beginning of the 20th century, the VKHS was described by a Swiss doctor, Alfred Vogt (1906) and, later, the disease was described by two Japanese researchers, Einosuke Harada (1926) e Yoshizo Koyanagi (1929)[1]. Currently, the cases described by Vogt, Koyanagi and Harada are considered the same disease with a wide spectrum of clinical variations named in honor of the three authors[2,3].

Specific ethnic groups more commonly present the disease, such as:

The Asian population (east and southeast of the continent), the Middle East, Indians, Hispanics and Native Americans. The African population rarely present the disease, suggesting that pigmentation is not the mainly etiologic component[1,3].

Regarding all the VKHS cases involving uveitis, the representative value in Japan is approximately 7%, 1-4% in the US and 3% in Brazil. In the US, the disease incidence is between 1.5 to 6 per million patients and in Japan, a mean of 800 new cases per year is registered[5]. Mostly VKHS patients present the disease between the second and the fifth decade of life, and women are the most affected , however, this is not true for the Japanese population[1,5].

Regarding the otological manifestations of the syndrome, 24% of the 67 patients studied who presented the VKHS had tinnitus complaints in Brazil and of the 137 patients studied in Japan, 54% of them presented the same complaints[5]. The hearing symptoms are found in 30% and 75% of VKHS cases which can be categorized as sensorineural hearing loss within the framework of clinical manifestations of the syndrome[3].

The VKHS mainly occurs in four steps: (1) prodromal, this phase is observed a few days prior to the ocular inflammation and it lasts for up two weeks and the patient can present neurological manifestations and ophtamological disorders, such as photofobia, headache, fever; (2) ophthalmic/acute, mainly characterized for the bilateral visual turbidity, bilateral uveitis, ocular ache, photofobia, conjunctival hyperemia, disacusia and serous multifocal retinal displacement; (3) chronic/convalescent, characterized by the melanocytes tissues depigmentation, such as the skin, eyes, poliosis, vitiligo, disacusia and uveitis; (4) chronic recurrent phase is characterized by recurrent granulomatous anterior uveitis and choroidal thickening with a less uveal thickening than that presented in the acute phase[6].

Overall, the determined criteria for the syndrome diagnosis are: (1) absence of surgery or ocular traumas previous to the uveitis; (2) absence of another ocular disease; (3) bilateral ocular disease; (4) neurological/audiological manifestations; (5) dermatological manifestations that did not precede the neurological/ocular manifestations[7].

The syndrome main diagnosis is clinical due to its variations and to help the diagnosis of the disease, a specific classification is defined by the International Nomenclature Committee: complete VKHS, incomplete or probable[8].

The patient that presents the complete VKHS shows all the criteria cited above, from number 1 to 5[7]. In the incomplete VKHS stage, the patient should present bilateral ocular disease, such as the three initial criteria associated to the number 4 or 5[7]. Regarding the probable VKHS stage, the patient should present isolated bilateral ocular disease associated to the three initial criteria[7]. The above mentioned topics help in the syndrome diagnosis. The differential diagnosis should be performed until the ophthalmic/acute stage due to some prodromal stage symptoms similar to a viral infection[8].

The differential diagnosis is important to perform due to the association between panuveitis with neurological, audiological, and dermatological symptoms that may also occur in patients with another disease. Some diseases should be differentiated from VKHS: Systemic lupus erythematosus, sarcoidosis, Behcet Disease, syphilis, tuberculosis, herpes, fungal infections, viral diseases, sympathetic ophthalmia, posterior acute multifocal epitheliopathy, secondary retinal multiple displacement, systemic arterial hypertension, vestibular neuronitis, Lyme disease and Cogan Syndrome[6,7].

A previous study assessed 14 medical records of patients with VKH disease to analyze in which stage of the disease the patients received the first specialized attendance[8]. From the 14 medical records assessed, only five of them received the first attendance in the ophthalmic stage of the disease, the maximum period for the early diagnosis of the syndrome; seven patients received the attendance in the convalescence stage and three in the recurrent chronic stage. Therefore, an early diagnosis and treatment are essential due to their relevant role in the prognosis of the syndrome.

The intervention for the VKHS is based on the treating of active ocular inflammation, the syndrome progression, and the possible ocular implications. Hence, the intervention is performed with the administration of systemic immunosuppressive agents associated to the corticosteroids[6,7]. Commonly, the early intervention is administered with oral prednisone and gradual decrease in the absence of recurrent clinic symptoms[6,7]. When the inflammation is severe, the treatment is performed with methylprednisolone intravenous pulse therapy for three or five days, followed by the administration of oral prednisone. The treatment with a low dose of corticosteroids should be kept for years until the end of the treatment[6,7].

Therefore, considering the audiological findings of VKHS and its rarity, the early diagnosis and treatment are relevant and the audiologist performs an important role during the intervention and referral to another specialized professional due to the common delay in the search for a doctor. Thus, the present case report aims to highlight the audiological aspects affected by the syndrome and the possible audiologist intervention for a specific case.

PRESENTATION OF THE CLINICAL CASE

The present case report was performed after the approval of the Ethics Research Committee, Faculdade de Odontologia de Bauru (FOB), Universidade de São Paulo (USP-number 3.396.812) and after the participant's signature in the Authorization Term, giving permission to use the data of her medical record in the present study. The Ethics Research Committee of the Institution approved the absence of an Informed Consent Form.

Anamnesis

A 54-year-old woman was assessed in the Speech Therapy Clinic of FOB with complaints of sudden hearing loss. In 2014, when the patient was 49 years old, she sought medical care due to the loss of visual acuity and hearing, and the diagnosis of VKHS and hearing loss was performed by the health professional. In the same year, the patient presented the onset of glaucoma and cataract. The treatment with prednisone was begun and kept until the present. In 2015, the patient reported that one year after the syndrome diagnosis, her visual acuity in the left eye partially improved, as well as her hearing compared with the previous year. The patient reported no family history of the syndrome and absence of parental consanguinity.

The patient did not understand what was being said on the telephone and reported that she constantly had to increase the television volume. She worked in the production department of a factory for nearly one year. The patient also referred acute and continuous tinnitus in the both ears and vestibular symptoms, such as crisis postural vertigo and imbalance when walking, leaning to the right side (Table 1)

Table 1. Main audiological findings

Clinical audiological findings	Occurrence in the case reported
Bilateral sensorineural hearing loss	(+)
Tinnitus	(+)
Vertigo	(+)
Dizziness	(-)
Inner ear impairment (cochlea)	(+)

Legend: (+): present; (-): absent.

Pure tone and vocal audiometry

The pure tone audiometry showed severe sensorineural hearing loss in the right ear and moderate sensorineural hearing loss in the left ear. Regarding the logoaudiometry, the speech recognition percentage index (SRPI) showed a good recognition and the speech recognition threshold was compatible with the pure tone audiometry (Table 2).

Table 2. Logoaudiometry results

	RE	LE
SRT	55dB	60dB
SRPI (monosyllables)	88% in 85dB	80% in 80dB
SRPI (syllables)	92%	96%

RE: Right ear; LE: Left ear; dB: Decibel; SRT: Speech Recognition Threshold; SRPI: Speech Recognition Percentage Index.

Immittance acoustic measures

The acoustic immittance measures showed an A curve and presence of stapedial acoustic reflexes contralateral and ipsilateral bilateral.

Otoacoustic Evoked emissions (OEA) and Brainstem Auditory Evoked Potential

There were absence of transient otoacoustic evoked emissions, and distortion showed in both ears. The brainstem auditory evoked potential showed integrity of the hearing pathway when the stimulus was presented bilaterally.

Selection and hearing aids (HA) fitting

In the year of 2017, the patient was fitted with a HA at another health system. The patient used a behind-the-ear HA less than one year due to the fact that it was broken.

In 2018, the patient was fitted with a completely new bilateral HA in the canal (CIC), with an omnidirectional microphone, automatic control of volume, WDRC compression and NAL-NL2 prescriptive rule. The real ear measurement was adjusted according to the target and the patient requirements and the patient made effective use of the HA verified by the datalogging. In the same year, the Client Oriented Scale of Improvement (COSI™) to assess the most important hearing situations for the patient was performed.

DISCUSSION

A previous study found that people of half age are mainly affected by the syndrome[9]. The VKHS is characterized by the ocular manifestations, however, hearing loss may be associated[10]. The patient of the present study presents moderate and severe sensorineural hearing loss associated with other otological symptoms, such as other studies, which found sensorineural hearing loss of different levels in the VKHS[9,10].

Similar to the case described, the vestibular-cochlear nerve is one of the main affected and it is responsible for the sudden bilateral sensorineural hearing loss and there is an absence of tympanic factors that could suggest a hearing loss with conductive components[7]. The progression of hearing loss would be fast and would cause comprehension of words impairment, however,

the patient described in the present study presented positive results for the SRPI[7].

The autoimmune physiopathology of the syndrome in the inner ear could explain the hearing loss in any frequency, such as in the case presented due to the possible autoimmune reaction against the inner and outer ear hair cells and a disorder in the vascular stria. Commonly, the disorders in the vascular stria cause flat hearing loss, however, high frequency hearing loss could initially exist[11].

The transient and distortion of product otoacoustic emissions were absent bilaterally and were compatible with the diagnostic hearing loss, such as a previous study that also found absent otoacoustic emissions in the studied patients that presented moderate or severe hearing loss[12].

In a previous study, a sample of 24 participants with VKHS diagnosis was assessed and showed notable otologic manifestations in three patients that presented tinnitus, two presented sudden hearing loss and one presented vertigo[10].

Previously, twenty patients with VKHS were assessed and six presented sudden hearing loss, four presented fluctuating hearing loss, seven reported tinnitus complaints, and five showed clinical manifestations related to the vestibular system (dizziness or vertigo), such as the case presented in the present study[4]. The same study evaluated the balance of the 20 participants and 14 presented nystagmus and 15 showed disorders during the balance test. These findings suggest peripheral vestibular disorders and hearing/vestibular impairments associated to the VKHS, such as the case reported by the present study.

Currently, the administration of corticosteroids and immunosuppressants has been recommended for the treatment of SVKH, with the aim of reducing inflammation caused by the syndrome[5,12]. A study indicated that the interval between hearing loss and the initiation of appropriate treatment plays an important role in the outcome of the syndrome[12]. In patients who started treatment with corticosteroids just after the start of hearing loss, the impairment was restored throughout the treatment. However, the patients who started the treatment several weeks after the start of hearing loss did not show responses to it. Thus, there is no hearing restoration prognosis if the impairment of the inner ear was severe, such as in profound hearing loss.

The usual otological manifestations of SVKH have a central origin and develop mainly in the ophthalmic/acute stage, with no complications or recurrences, commonly presenting a positive prognosis with remission of the symptoms when early and appropriate treatment is performed[7].

In the specific case presented, the patient has been treated with prednisone since the diagnosis of the syndrome in 2014, however, the hearing loss, moderate on one side and severe on the other, remains until the present moment. Even if the diagnosis is made in the ophthalmic/acute stage of the syndrome, the limit in which early diagnosis and treatment should occur[8], suggest the greater involvement of the inner ear presented by the patient could have limited the remission of hearing loss, causing the necessity to still use a HA.

The appropriate therapeutic guidance process depends on the medical diagnostic assertiveness of the syndrome and the multiple therapeutic approach regarding the vestibular symptoms[7]. Due to the otological changes concomitant with the syndrome, audiological exams can be used to diagnose the impairment in most cases, as well as assist in the therapeutic process and prevent indications of inadequate sound amplification devices[7-9].

FINAL COMMENTS

The case described in the present study presented sudden bilateral sensorineural hearing loss, as well as tinnitus, vertigo and bilateral ocular involvement. Despite the treatment with corticosteroids, hearing loss remained, and the early diagnosis and intervention could help in a better hearing prognosis.

However, the patient has adapted to using the HA and there was an improvement of the visual acuity of the left eye.

Thus, prior to the HA indication, the audiologist must recommend the audiological monitoring of the case and investigate whether the patient undergoes drug treatment since it can bring hearing improvements.

The professional should also pay attention to the patient who does not have the diagnosis of SVKH, but who has suggestive symptoms to make the required referral to the specialized professional, assisting in the early diagnosis of the syndrome and actively participating in an interdisciplinary team in the therapeutic process regarding hearing.

REFERENCES

1. Damico FM, Kiss S, Young LH. Vogt-Koyanagi-Harada Disease. Seminars in Ophtalmology. 2005;20(3):183-90. DOI: 10.1080/08820530500232126.
2. Greco A, Fusconi M, Gallo A, Turchetta R, Marinelli C, Macri GF et al. Vogt-Koyanagi-Harada Syndrome. Autoimunity Reviews. 2013;12(11):1033-38. PMid: 23567866. DOI: 10.1016/j.autrev.2013.01.004.
3. Mota LAA, Santos AB. Síndrome de Vogt-Koyanagi-Harada e o seu acometimento multissistêmico. Rev Assoc Med Bras.2010;56(5):590-5. DOI: 10.1590/S0104-42302010000500023.
4. Kimura H, Ohashi N, Aso S, Watanabe Y. Clinical Study of the Role of Melanocytes in the inner ear of patients with Harada's Disease. ORL. 1996;58(4):233-7. PMid: 8883112. DOI: 10.1159/000276843.
5. Lavezzo MM, Sakata VM, Morita C, Rodriguez EEC, Abdallah SF, da Silva FTG et al. Vogt-Koyanagi-Harada disease: review of a rare autoimmune disease targeting antigens of melanocytes. Orphanet Journal of Rare Diseases. 2016;11(1):29. PMid: 27008848. PMCID: PMC4806431. DOI: 10.1186/s13023-016-0412-4.
6. Baltmr A, Lightman S, Tomkins-Netzer O. Vogt–Koyanagi–Harada syndrome–current perspectives. Clin Ophthalmol. 2016;10:2345-61. PMid: 27932857. PMCID: PMC5135404. DOI: 10.2147/OPTH.S94866.
7. Carneiro SG, da Silva DL, Palheta ACP, Neto FXP, Nunes CTA, Ferreira TO et al. Síndrome de Vogt-Koyanagi-Harada: Revisão de Literatura. Arq Int Otorrinolaringol. 2008;12:419-25.
8. da Costa GFS, Biancardi AL, Xavier CA, Provenzano G, Júnior HVM. Vogt-Koyanagi-Harada syndrome: evaluation of the disease phase in which patients receive the first specialized attendance. Rev Bras Oftalmol. 2018;77(2):85-8. DOI: 10.5935/0034-7280.20180018.
9. Rivera VR, Garrigues HP, Pinazo RG. Sensorineural Hearing Loss Evolution in Vogt-Koyanagi-Harada Syndrome. Acta Otorrinolaringologica (English Edition). 2011;62(6):465-8. DOI: 10.1016/j.otoeng.2010.09.001
10. Ondrey FG, Moldestad E, Mastroianni MA, Pikus A, Sklare D, Vernon E et al. Sensorineural Hearing Loss in Vogt-Koyanagi-Harada Syndrome. The Laryngoscope. 2006;116(10):1873-6. PMid: 17003710. DOI: 10.1097/01.mlg.0000234946.31603.fe.

11. Ruiz-Allec LD, Peñaloza-López Y, Ocaña-Plante NR, Valdez-González T, López-Star E. Peripheral and central audiologic findings in patients with Vogt-Koyanagi-Harada Syndrome. Acta Otorrinolaringologica (English Edition). 2009;60(4):253-9. PMid: 19814971. DOI: 10.1016/j.otorri.2009.01.002.

12. Dousary SAL. Auditory and vestibular manifestations of Vogt–Koyanagi–Harada disease. The Journal of Laryngology & Otology. 2011;125(2):138-41. PMid: 20880417. DOI: 10.1017/S0022215110001817.

Authors' contribution

MCF performed the data collection, analysis and interpretation of the data, analysis of the theoretical content, revision of the literature, manuscript writing and manuscript submission; ILM performed the critical revision of the intellectual content, analysis of the theoretical content, revision of the literature and manuscript writing; MFCGM participated as supervisor of the research, conception of the study project, data analysis, correction and approval of the final version of the manuscript.