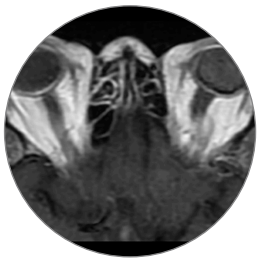

Vogt-Koyanagi-Harada Syndrome Cerebral and Ocular Manifestations

Manifestaciones cerebrales y oculares del Síndrome de Vogt-Koyanagi-Harada

Luis Felipe Ovalle Daza[1]
Catalina Wilches Vanegas[2]

DOI: https://doi.org/10.53903/01212095.129

Summary
Vogt-Koyanagi-Harada syndrome, formerly kwon as uveomeningoencephalitic syndrome, is a rare disease, with very few cases reported in the literature. It is an uncommon autoimmune entity with multi-organ involvement. Magnetic resonance imaging is important to identify periventricular white matter lesions, brain stem and brain peduncles involvement, as well as pachymeningeal enhancement and diffuse choroidal thickening. Despite being a rare entity, it is important for the radiologist be acquainted with its imaging findings.

Key words (MeSH)
Uveomeningoencephalitic syndrome
Magnetic resonance imaging
Central nervous system

Resumen
El síndrome de Vogt-Koyanagi-Harada, antes conocido como síndrome uveomeningoencefálico, es una enfermedad rara, con muy pocos casos descritos en la literatura. Es una patología autoinmune infrecuente, con un compromiso multiorgánico. La resonancia magnética es importante para identificar las lesiones de la sustancia blanca periventricular, la afectación del tallo cerebral y los pedúnculos cerebrales, así como el realce paquimeníngeo y el engrosamiento difuso de la coroides. A pesar de ser una entidad poco frecuente, es importante que el radiólogo esté familiarizado con sus características en imágenes.

Palabras clave (DeCS)
Síndrome uveomeningoencefálico
Imagen por resonancia magnética
Sistema nervioso central

Introduction

Vogt-Koyanagi-Harada syndrome (VKHS) is a rare multi-organ autoimmune disorder, with few cases described in modern literature (1). VKHS mainly affects the ocular pigmented tissue, and causes alterations to the auditory tissue, integumentary tissue and central nervous system (CNS). It is characterized by bilateral uveitis, retinal detachment, vitiligo, alopecia, hearing loss, tinnitus, headache and meningismus (2). Magnetic resonance imaging (MRI) allows the identification of choroidal involvement -with typical findings of diffuse thickening-, without damage to the sclera, associated with retinal detachment (3). Regarding the CNS, initial reports describe high signal lesions in T2-weighted sequences of the periventricular white matter, while more recent literature has documented lesions in the brain stem and cerebral peduncles, as well as pachymeningeal enhancement (4,5). In the case presented here we describe the typical brain MRI findings in a patient with VKHS.

Clinical Case

A 63-year-old female patient diagnosed with VKHS presents to the Radiology Department of the institution to which the authors belong, with uveitis associated with loss of visual acuity in the left eye and decreased visual acuity in the right eye, in addition to areas of skin depigmentation in the upper and lower limbs detected on physical examination. The patient reported occasional holocranial headache and bilateral hypoacusis.

Discussion

VKHS was first described in the 10th century by the Baghdadi ophthalmologist Ali Ibn Isa, who described the characteristic physical appearance of ocular inflammation, depigmentation of hair, eyebrows and eyelashes. It is believed that a genetic basis may be a predisposing factor, as there is a higher incidence in patients with more pigmented skin; however, a characteristic inheritance pattern has not yet been found (2). One theory suggests that VKHS has variable clinical manifestations, which depend on the stage of the syndrome at the time of presentation. The pathophysiology of this disease has not been fully elucidated, but it is thought to be related to an autoimmune response caused by T lymphocytes against melanocyte-related protein tyrosinases, which are found in the uvea, retina and leptomeninges (6).

VKHS develops in four phases: prodromal, acute, convalescent and chronic recurrent uveitis. The prodromal phase presents as a viral infection and can last from a few days to a few weeks. In this phase the clinical manifestations are mainly extraocular,

[1]Fourth year resident in radiology radiology, Fundación Universitaria Sanitas. Bogotá, Colombia.

[2]Medical radiologist, Clínica Reina Sofía. Bogotá, Colombia.

Department of Radiology and Diagnostic Imaging of the Clínica Reina Sofía. Bogotá, Colombia.

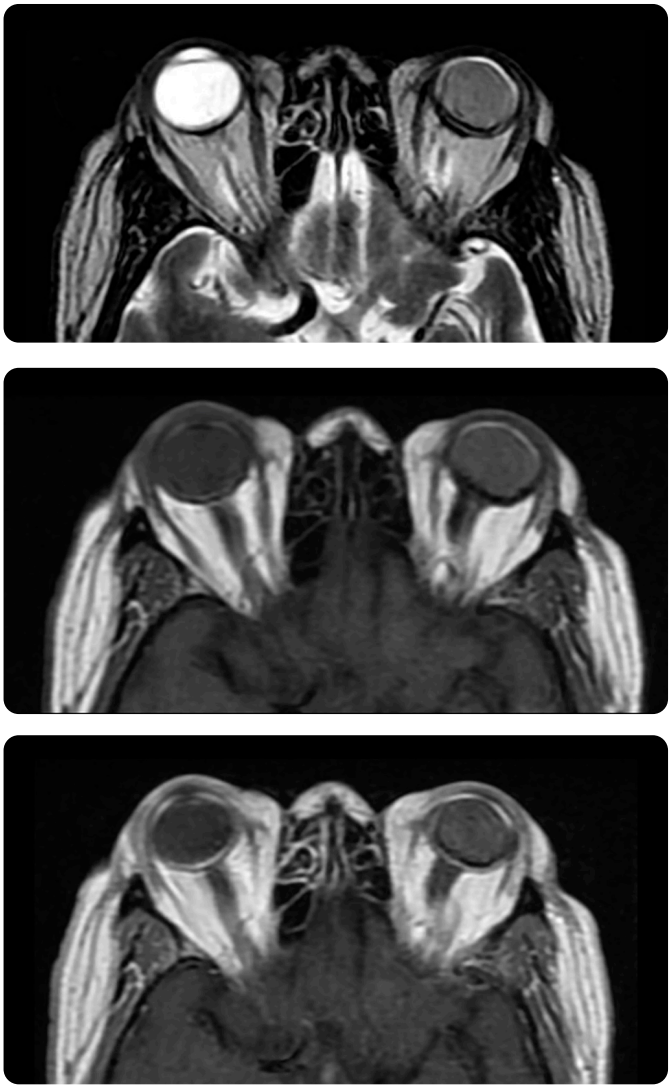

Figure 1. a) Axial MRI with T2 information: bilateral choroidal thickening; alteration in the morphology and in the signal of the left eyeball, decrease in size, due to old changes of detachment of the left retina and surgical absence of the crystalline lens. Phakectomy with intraocular lens in the right orbit. (b) MRI with T1 information and (c) with contrast medium: bilateral choroidal thickening and enhancement.

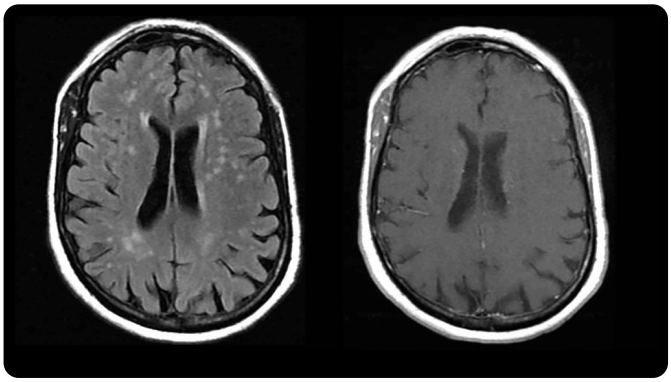

Figure 2. MRI, axial FLAIR and spin echo with T1 information and with contrast medium: multiple hyperintensities in the periventricular white matter, bilateral semioval and subcortical centers, these lesions do not enhance with the administration of contrast medium and may correspond to age-related changes. No pachy- or leptomeningeal relay is observed.

such as headache, meningismus, fever, nausea, vertigo and hearing disturbances.

The acute uveitis phase follows the prodromal phase and in this phase begins the sudden alteration of visual acuity in both eyes, secondary to granulomatous uveitis and choroidal thickening. The convalescence phase occurs several weeks or months after the acute uveitis phase and may last several months. In this phase, choroidal depigmentation, vitiligo and poliosis are observed. Finally, the chronic recurrent phase occurs in some patients, develops between 6 and 9 months from the onset of symptoms, and is characterized by exacerbations of uveitis, which is resistant to systemic corticosteroid therapy (7).

There is still no consensus on the definitive diagnostic test for VKHS, which limits it to a combination of paraclinical studies. VKHS is classified as complete, incomplete and probable, based on the following criteria: 1. Uveitis without a history of trauma or ocular surgery. 2. Uveitis without clinical or paraclinical evidence of other ocular disease. 3. Bilateral uveitis with retinal detachment. 4. Auditory or neurological (not visual) involvement. 5. Cutaneous findings (8). Complete VKHS meets all 5 criteria. Incomplete VKHS meets the first 3 criteria, with criteria 4 or 5. VKHS is considered probable when the involvement is ocular only, only fulfilling the first 3 criteria (8). Despite being a systemic pathology, up to 54% of patients with VKHS only have ocular symptoms in the early stage of the syndrome. Considering the above, extraocular manifestations of full-blown VKHS may appear months or years after the initial ocular manifestations (9).

MR images are a useful tool in the diagnosis of VKHS; in addition to bilateral ocular findings (choroidal enhancement on contrast-enhanced images, with diffuse thickening, sclera respect and retinal detachment) (Figure 1), high diffuse periventricular signals are also observed on T2 and FLAIR sequences (Figure 2) (3). In addition, meningeal enhancement has recently been described as another finding of this disease in brain MRI, but it was not found in the patient of this case (4). The ability of MRI to demonstrate meningeal enhancement fits the theory of melanocyte-rich tissue involvement, and may explain early clinical symptoms, such as headache occurring in up to 82% of patients, before the onset of frank neurological symptoms, so this enhancement may be an early marker of CNS involvement (10).

Conclusion

VKHS is a rare autoimmune disease, affecting multiple organs. MRI is useful to identify ocular and brain involvement, so it is important for the radiologist to be familiar with its characteristics and the different imaging findings.

References

1. Lohman BD, Gustafson CA, Mckinney AM, Sarikaya B, Silbert SC. MR imaging of Vogt-Koyanagi-Harada syndrome with leptomeningeal enhancement. AJNR. 2011;32(9):E169-71.
2. Moorthy RS, Inomata H, Rao NA. Vogt-Koyanagi-Harada syndrome. Surv Ophthalmol. 1995;39(4): 265-92.
3. Ibanez HE, Grand MG, Meredith TA, Wippold FJ. Magnetic resonance imaging findings in Vogt-Koyanagi-Harada syndrome. Retina. 1994;14(2):164-8.
4. Han HJ, Kim HY, Park JH, Lee EJ, Kim Do G, et al. Magnetic resonance imaging of pachymeningeal enhancement in Vogt-Koyanagi-Harada disease. Neurol Sci. 2010;31(6):785-8.
5. Hashimoto T, Takizawa H, Yukimura K, Ohta K. Vogt-Koyanagi-Harada disease associated with brainstem encephalitis. J Clin Neurosci. 2009;16(4):593-5.

6. Rao NA. Mechanisms of inflammatory response in sympathetic ophthalmia and VKH syndrome. Eye. 1997;11:213-6.
7. O'keefe GAD, Rao NA. Vogt-Koyanagi-Harada disease diagnostic criteria. Surv Ophthalmol. 2016;62(1):1-25.
8. Read RW, Holland GN, Rao NA, Tabbara KF, Ohno S, et al. Revised diagnostic criteria for Vogt-Koyanagi-Harada disease: report of an international committee on nomenclature. Am J Ophthalmol. 2001;131(5):647-52.
9. Rao NA, Gupta A, Dustin L, Chee SP, Okada AA, et al. Frequency of distinguishing clinical features in Vogt-Koyanagi-Harada disease. Ophthalmology. 2010;117(3):591-9.
10. Rao NA, Sukavatcharin S, Tsai JH. Vogt-Koyanagi-Harada disease diagnostic criteria. Int Ophthalmol. 2007;27(2-3):195-9.

Correspondence

Luis Felipe Ovalle Daza
Carrera 17 # 96-61, apto 501
Bogotá, Colombia
pipeovalle17@gmail.com

Received for evaluation: November 30, 2020
Accepted for publication: January 19, 2021