

Vogt-Koyanagi-Harada Incomplete Disease: A Case Report

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Received date: September 26, 2018; Accepted date: October 06, 2018; Published date: October 11, 2018

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Abstract

Introduction: Vogt-Koyanagi-Harada disease is a systemic autoimmune disease characterized mainly by a bilateral granulomatous panuveitis, in which antibodies are produced against tissues rich in melanocytes such as the retina, inner ear, meninges, skin, and hair, which explains its extraocular manifestations and its clinical presentation.

Case report: A 27-year-old Mexican woman presented with sudden reduced visual acuity, with no history of trauma or eye surgery, without evidence of another ocular disease, she also presented meningism and disacusia. Retinal detachment is evidenced with the help of imaging studies, in addition to cerebrospinal fluid pleocytosis. The final diagnosis during her stay in the hospital was VKH disease.

Discussion: The clinical case presented, based on current criteria for the diagnosis and classification of VKH disease is an incomplete VKH, and it was not possible to demonstrate the dermatological involvement in future stages to classify it as a complete VKH disease.

Conclusion: VKH disease is difficult to diagnose, because its diagnostic criteria are mainly exclusion and its clinical presentation is variable and insidious, which results in a generally late diagnosis and treatment, allowing the appearance and progression of sequelae.

Possibly, the diagnostic criteria for VKH disease should be subjected to a new review, to specify the most frequent clinical data in recent years, since they were published almost a couple of decades ago.

Keywords: Vogt-Koyanagi-Harada incomplete disease; Uveomeningeal syndrome; Retinal detachment; Systemic autoimmune disease

Introduction

The uveomeningeal syndromes are a heterogeneous group of disorders or conditions that share as a common characteristic, the affection of the retina, uvea and meninges [1], their causes can be infectious, due to neoplastic process or to systemic diseases.

This is a systemic autoimmune disease characterized mainly by a bilateral granulomatous panuveitis, in which antibodies are produced against tissues rich in melanocytes such as the retina, the inner ear, the meninges, skin and hair [2], which explains its extraocular manifestations and its clinical presentation.

It is known that VKH disease has a predilection for pigmented races, being less frequent in Caucasians, although its low prevalence in sub-Saharan Africans makes it consider that it is not the only ethnicity. It mainly affects the female gender between the third to fifth decades of life [3].

VKH disease has a clinical presentation divided into 4 phases consisting of the following:

Prodromal phase

Nonspecific symptoms among which are fever, headache, nausea, meningism, vertigo and disacusia, rarely include ataxia, confusion and neurological focalities. It usually lasts 3 to 5 days.

Acute uveitic phase

Decreased visual acuity, usually unilateral onset, then contralateral with a difference of 1-3 days, adds photophobia, iridocyclitis, retrokeratic deposits, ocular hypertension, vitreitis.

Choroiditis that results in rupture of the external hematorretinal barrier (pigmented epithelium of the retina), causing exudative retinal detachments. It can cause acute glaucoma. It lasts several weeks.

Convalescence phase

Retinal detachments gradually subside and uveitis begins to subside. There is a pigmentary redistribution that gives rise to an eye fund "like sunset" (sunset glow fundus). It can last for months.

Chronic recurrent phase

It can interrupt the convalescent phase with recurrent anterior uveitis. There is formation of synechiae, pupillary block, iris atrophy, cataracts and glaucoma.

Systemic manifestations

Neurological symptoms: Headache, meningism, encephalitis, paralysis of the cranial nerves, aphasia, there may even be changes in personality, loss of consciousness and seizures. They usually appear in the prodromal phase.

Auditory symptoms: Tinnitus, hypoacusia and vertigo, mainly. They can be present in all phases.

Dermatological manifestations: Vitiligo, alopecia and poliosis; generally in the convalescence phase.

The diagnosis is made through clinical and image criteria (Figure 1) established and published by the American Uveitis Society in 2001 [4].

The treatment is mainly based on systemic corticosteroids during the acute phase, there is no established consensus of which medication and which route to use, however there are multiple reports of improvement with intravenous methylprednisolone for three days with follow-up with oral prednisone for at least 6 months.

Other treatment options include immunosuppressants such as azathioprine, cyclosporine, methotrexate and the use of immunoglobulins [3,5], and the use of monoclonal antibodies such as adalimumab has been documented, which is associated with a lower risk of recurrence and visual impairment [6].

The complications that occur are related to the duration of the disease, recurrences and the age of the patient at the time of suffering from the disease. The complications that usually occur are cataracts, glaucoma, choroidal neovascularization and subretinal fibrosis [4].

Case Report

Female patient of 27-years of age, of Mexican nationality, with no pathological personal history of contributory illness, she began a month before her medical attention, with pulsatile-type holocranial headache, to which an episode of visual acuity loss was added suddenly with duration of approximately six hours before the spontaneous remission referred by the patient.

Four weeks later, the patient presented with paresthesias in the lower lip, dysarthria and bilateral hearing loss. She then presented with headaches of the same characteristics as 4 weeks ago, for which she went for assessment to the emergency department.

At her arrival at the emergency department, the patient is oriented, with a Glasgow of 15 points, without apparent involvement of cranial nerves, strength preserved in extremities, 12 hours later has a decrease in the Glasgow coma scale score to 12 points, in addition to meningeal signs.

A simple skull tomography was performed where there is no evidence of pathology, lumbar puncture was performed to study cerebrospinal fluid with report of rock water appearance, proteins 62 mg/dl, glucose 73 mg/dL (central glycaemia 84 mg/dL), 138 cells per mm³, 70% mononuclear, Gram stain, negative.

Empirical treatment was started with ceftriaxone, vancomycin and acyclovir without showing marked improvement.

1. Exclude intraocular surgery or penetrating ocular trauma
2. Exclude other ocular diseases
3. Bilateral ocular involvement (A or B)
 - A. *Early manifestations*
 1. Evidence of diffuse choroiditis.
 - a. Focal areas of subretinal fluid or
 - b. Bullous serous retinal detachment
 2. Equivocal fundus findings, both of the following must be present
 - a. Fluorescein angiography: focal areas of delay in choroidal perfusion, multifocal of pinpoint leakage, large placoid areas of hyperfluorescence, pooling within subretinal fluid and optic nerve staining
 - b. Ultrasonography: diffuse choroidal thickening without evidence of scleritis
 - B. *Late manifestations*
 1. History suggestive of prior presence of findings from 3A or either or both 2 and 3 below or multiple signs from 3
 2. Ocular depigmentation
 - a. Sunset glow fundus
 - b. Sugiura sign
 3. Other ocular signs
 - a. Nummular chorioretinal depigmented scars or
 - b. Retinal pigment epithelium clumping or
 - c. Recurrent or chronic anterior uveitis
4. Neurologic/auditory findings such as
 - a. Meningismus
 - b. Tinnitus
 - c. Cerebrospinal fluid pleocytosis
5. Dermatologic manifestations
 - a. Vitiligo
 - b. Alopecia
 - c. Poliosis

Complete VKH = The criteria from 1 to 5 must be present
Incomplete VKH = Criteria 1 to 3, and at least one of criteria 4 or 5
Probable VKH = Only eye disease, criteria 1 to 3

Figure 1: Diagnostic criteria for VKH disease.

On the third day of hospital stay, visual acuity decreased until total amaurosis, so the ophthalmology service was consulted, who reported diminished visual acuity, found a pupillary response to luminous stimulus, pupillary red reflex present, there is no exudates or hemorrhages, however there is bilateral retinal detachment with abundant subretinal fluid, hyperemic sclera and effacement of papillary margins, arterial pulse present and white-yellow lesions in posterior pole.

Subsequently, magnetic resonance imaging was performed, where a bilateral retinal detachment can be observed as the only anomaly (Figures 2 and 3), which is corroborated by optical coherence tomography (Figures 4 and 5).

So that in the presence of the meningeal syndrome accompanied by visual and auditory affection without evidence of infectious or traumatic etiology, in addition to the imaging findings, an uveomeningeal syndrome, caused by VKH disease, is integrated, initiating treatment based on intravenous corticosteroids at high doses for 3 days, obtaining significant improvement in all signs and symptoms, with the exception of visual acuity that had a torpid evolution, treatment with azathioprine was initiated and partial improvement in visual acuity was obtained.

Was not possible to escalate treatment to monoclonal antibodies because not having the resource.

Patient is discharged with outpatient treatment and follow-up appointments, however it has not been presented.

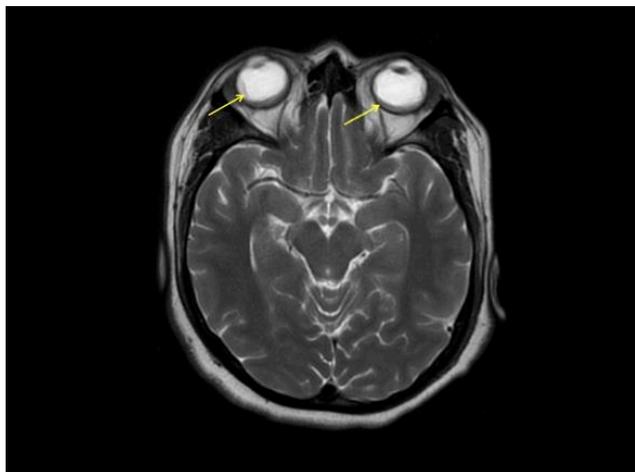


Figure 2: Magnetic resonance, bilateral retinal detachment (arrows) is observed.

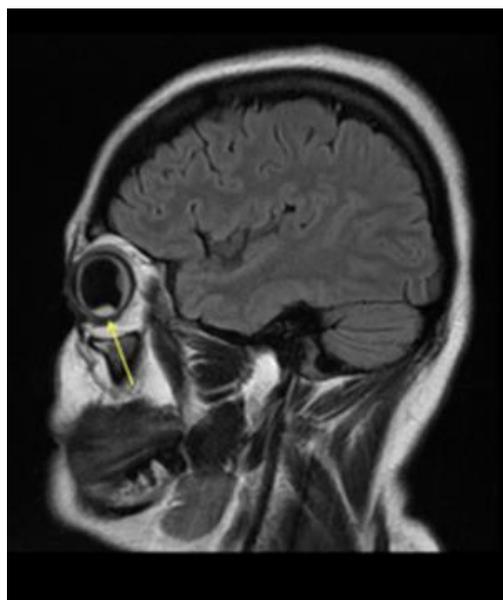


Figure 3: Flair magnetic resonance with retinal detachment (arrow).

Discussion

The exposed clinical case meets criteria to classify it as incomplete VKH disease, as it has ophthalmological implications that do not have a history of trauma or ocular surgery prior to the onset of symptoms, absence of evidence suggestive of another ocular disease, bilateral ocular involvement, neurological and auditory manifestations, supporting the diagnosis in the presented imaging studies and intense pleocytosis in the cerebro-spinal fluid. It is not possible to detect tegumentary manifestations since these are usually characteristic of the convalescence phase and are almost always detected at follow-up appointments. In this particular case, it was not possible to corroborate a complete VKH disease or discard it for the reasons already explained

in the presentation of the case, being classified only as incomplete VKH disease. It has been described as an important prognostic factor the evolution of visual acuity as a response to a month of treatment [7], so the inability to follow the patient, deprives us of prognostic data of the particular case, as well as the detection of late-onset sequelae and their subsequent study.

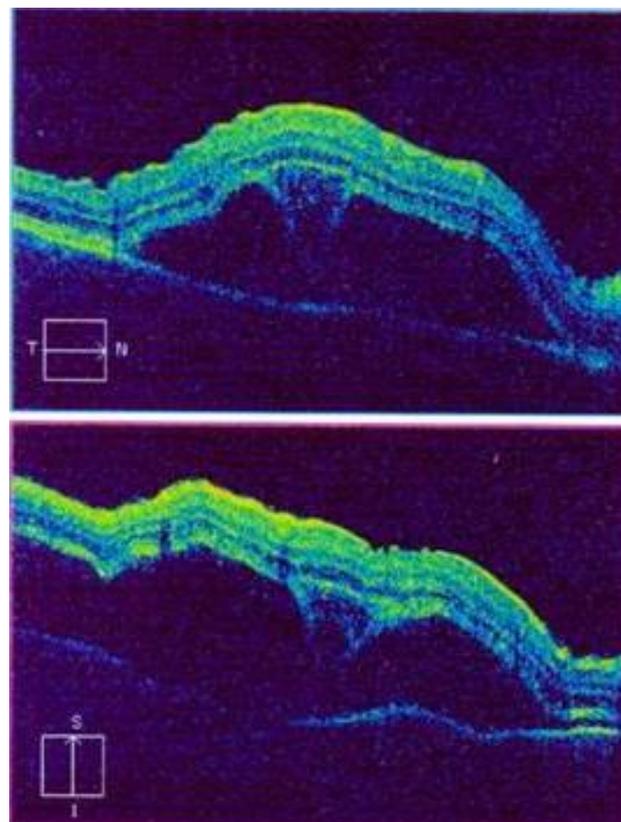


Figure 4: Optical coherence tomography of the right eye, with retinal detachment and subretinal fluid.

Conclusion

VKH disease is a rare pathology, in Mexico it has a prevalence of 2.4%, it is more common in Asians, with a higher prevalence in India up to 21.08% [8]. It is usually difficult to diagnose since there are about 30 diseases that present uveitis. To reach the diagnosis of VKH is complicated, although there are criteria, these are rather exclusionary. Probably, the majority of Vogt-Koyanagi-Harada disease cases published is "incomplete". This fact could make us think about the inadequacy of the diagnostic criteria, or about the inadequate characterization of the disease. Possibly, the diagnostic criteria for VKH disease should be subjected to a new review, to specify the most frequent clinical data in recent years, since they were published almost a couple of decades ago. Due to its presentation and frequency, there is relatively little clinical information, which translates into a diagnosis and delayed treatment, allowing sequelae to occur generally [8,9]. The case presented here is about a pathology that is extremely rare in our environment, being the first case that has been reported in this region of Mexico, however, had an apparently timely diagnosis, although it had a torpid evolution in terms of affection visual, despite timely

treatment of first choice, escalating to azathioprine. The use of monoclonal antibodies was not possible due to lack of recourse. We consider the report of this clinical case to be of utmost importance, in order to raise awareness of one more form of clinical presentation of VKH disease, such as the sudden onset of total amaurosis in the early stages of the disease, and not in a gradual manner as regularly are reported.

Conflict of Interests

None

Funding

None

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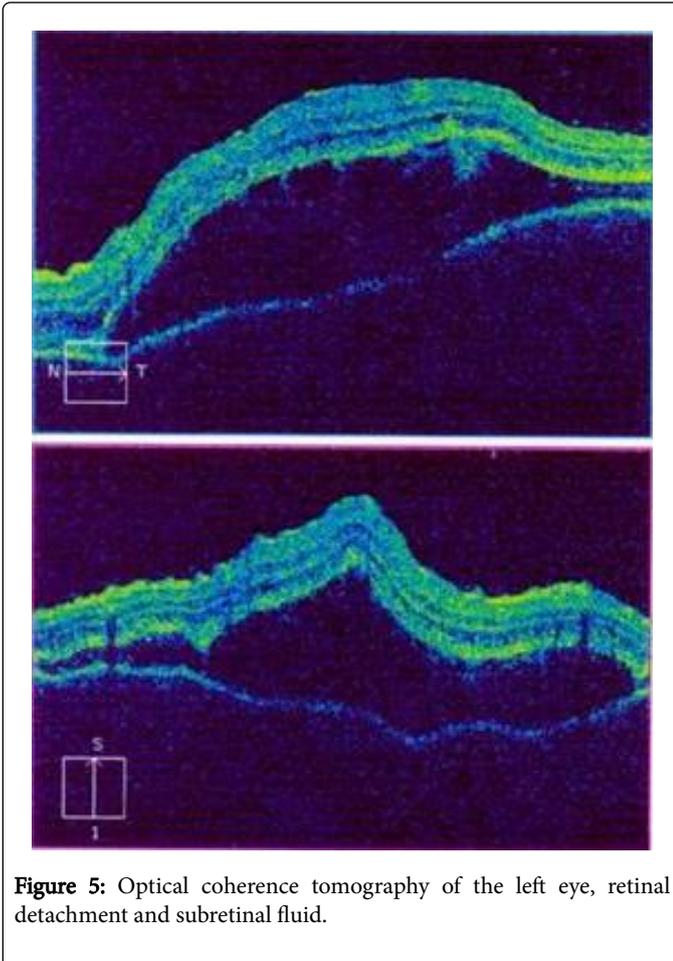


Figure 5: Optical coherence tomography of the left eye, retinal detachment and subretinal fluid.