Case report

Presumed atypical peripapillary Vogt-Koyanagi-Harada disease

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ABSTRACT

Purpose: To describe a case of bilateral presumed atypical Harada disease with sequential, not simultaneous, involvement of the peripapillary retina (subretinal fluid) in a healthy patient with no systemic complaints.

Observation: A 35-year-old healthy white man presented with sudden paracentral visual loss in the left eye. His medical history was unremarkable. However, he reported a similar episode 20 months earlier in the right eye that was associated with macular serous retinal detachment. The right eye showed evidence of reactive peripapillary atrophy and pigmentary alteration in the macula. Optical coherence tomography scans of the posterior left eye segment revealed diffuse thickened choroid, papillomacular subretinal exudate and discontinuity of the ellipsoid layer with suggestion of vitreous cellularity. Autofluorescence imaging of the left eye showed peripapillary hyperautofluorescence. A fluorescein angiogram revealed progressive staining and pooling of the peripapillary retina with corresponding retinal vasculitis. Indocyanine green angiography revealed multiple hypocyanescent lesions with an area of hypercyanescence temporal to the disc. Rheumatologic evaluation and laboratory tests were all negative. Chest tomography was normal. Considering the apparent absence of infectious diseases, the patient was started on 60 mg/day prednisone. After 8 days, visual acuity improved to 20/250, improving to 20/20 vision six months after a slow steroid wean.

Conclusion: We believe our case represented a variant of the Vogt-Koyanagi-Harada syndrome in an atypical situation, because the patient fulfilled the presumed criteria. Furthermore, the findings of clinical and complementary examinations led to this nosological entity to the exclusion of others.

Importance: The point of this case is to alert ophthalmologists to the existence of this atypical presentation of the disease so that it should be included among the differential diagnoses of pathologies that present with these findings.

1. Introduction

Serous retinal detachment represents a diagnostic challenge because of its etiological diversity. Inflammatory and infectious diseases should be considered in order to direct appropriate clinical management.1

We describe a case of bilateral presumed atypical Harada disease with sequential, not simultaneous, involvement of the peripapillary retina with subretinal fluid, in a healthy patient with no systemic complaints.

1.1. Case report

A 35-year-old healthy white man presented with sudden paracentral visual loss in the left eye. His medical history was unremarkable. He had reported a similar episode 20 months earlier in the right eye that was associated with macular serous retinal detachment (Fig. 1-a and 1-b). At that time, the exam of the left eye showed no abnormalities (Fig. 1-c), and the results of systemic and laboratory investigations, including a study of the cerebrospinal fluid, were normal. He was diagnosed elsewhere with an optic disc pit maculopathy in the right eye. Laser treatment associated with a pneumatic retinopexy was performed and this was believed to have resulted in complete regression of the retinal detachment (Fig. 1-d). By the time we first examined the patient, visual acuity was 20/25 in both eyes. Extrinsic ocular motility, pupillary reflexes and anterior segment biomicroscopy were normal in both eyes. Right eye retinography revealed evidence of reactive peripapillary atrophy and pigmentary alteration in the macula (Fig. 2-a). In the left eye, retinography revealed peripapillary serous retinal detachment...
Auto-fluorescence imaging in right eye revealed peripapillary hypo-auto-fluorescence and in the left eye there was peripapillary hyper-auto-fluorescence. Fluorescein angiogram (FA) in the left eye revealed progressive staining and pooling of the peripapillary retina with corresponding retinal vasculitis (Fig. 2-c). Indocyanine green angiography (ICGA) revealed multiple hypocyanescent lesions with an area of hypercyanescence temporal to the disc (Fig. 2-c). Spectral-domain optical coherence tomography (OCT-SD) scans through the posterior left eye segment revealed a diffuse thickened choroid, papillomacular subretinal exudate and discontinuity of the ellipsoid layer with suggestion of vitreous cellularity (Fig. 2-d). There was no evidence of pits in either eye. Laboratory tests were required at that time.

During this period, the patient presented with worsening vision in the left eye, and an increased area of serous detachment reaching the fovea could be seen on retinography (Fig. 2-e), red-free, auto-fluorescence and infra-red. The OCT-SD showed an increase in the sub-retinal fluid, affecting the foveal zone, in addition to the increase of the amorphous substance and the vitreous cellularity (Fig. 2-f).

Laboratory tests, including hemogram, erythrocyte sedimentation rate, C-reactive protein, rheumatoid factor and antinuclear antibodies were normal. We excluded the following infectious diseases: HIV, syphilis, toxoplasmosis, cytomegalovirus, herpes simplex, herpes zoster and Bartonella henselae. Chest tomography was normal. Considering the apparent absence of infectious diseases, the patient was started on 60 mg/day prednisone. After 8 days, visual acuity improved to 20/250, improving to 20/20 vision six months after slow steroid wean. This improvement could be demonstrated on retinography (Fig. 2-g), OCT-SD (Fig. 2-h) and infra-red.

2. Discussion

These fundoscopic findings in a previously-healthy young white man revealing rapidly progressive low visual acuity suggested a heterogeneous group of pathologies that could be responsible.

Infectious causes, including syphilis and tuberculosis, were excluded due to negative serologies. Chest tomography showed no abnormalities, making the possibility of sarcoidosis highly unlikely.

Ocular ultrasonography and neuroimaging showed no signs suggestive of scleritis, tumor or lymphoma that were not consistent with the FA and ICGA.
Some diseases belonging to the group of white dots syndrome should be considered. Acute zonal occult outer retinopathy (AZOOR), acute posterior multifocal placoid pigment epitheliopathy (APMPPPE) and serpiginous choroiditis may be accompanied by vitritis and low visual acuity, depending on the extent of involvement of the external retina and fovea. The peripapillary RPE atrophy observed in the patient's right eye can be found in more advanced cases of AZOOR and serpiginous choroiditis. Chao et al. described a case of multiple evanescent white dots syndrome (MEWDS) with similar characteristics to those found in our case, including thickening of the choriocapillaris and serous detachment observed on optical coherence tomography. However, despite the presence of early hypofluorescent lesions in FA, also found in APMPPE, multifocal choroiditis and panuveitis, there were no typical angiographic or fundoscopic patterns of any spectrum of these pathologies, as seen in this case.

Although the diagnosis of Vogt-Koyanagi-Harada syndrome (VKHS) requires bilateral ocular involvement, there have been reports in the literature of unilateral and bilateral non-simultaneous involvement. Typical findings of VKHS that are found in FA (such as pin points) were not detected in our patient. Furthermore, a circumscribed type of peripapillary serous retinal detachment that slowly advanced to the macula in both eyes is an atypical presentation of Harada disease as well. Our patient initially presented with peripapillary serous retinal detachment in the left eye, eighteen months after a similar event in the right eye that had been misdiagnosed as a papilla pit. We believe both eyes suffered from the same syndromic condition, but in distinct periods, because the clinical presentation and findings from the complementary exams were similar. This observation reinforces the probable diagnosis of VKHS, with bilateral ocular involvement occurring non-simultaneously.

The diagnostic criteria for VKHS were published in 2014 (Table 1). In this case report, the patient met the criteria for the probable form, because trauma and previous ocular surgeries were excluded, as were other ocular and systemic pathologies.

### 3. Conclusion

We believe our case represented a variant of the VKHS in an atypical situation, because the patient fulfilled the presumed criteria. Furthermore, the findings of clinical and complementary examinations led to this nosological entity to the exclusion of the others. It is important to promptly diagnose atypical VKHS, so that early therapy can be directed in order to improve visual prognosis.

Patient consent: Written consent to publish this case has not been obtained.

This report does not contain any personal identifying information.

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#### Conflicts of interest

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