ORIGINAL ARTICLE

Vogt-Koyanagi-Harada Disease Presenting as Unilateral Neuroretinitis

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ABSTRACT

A 42-year-old female presented with unilateral visual loss associated with systemic symptoms of fever and headache. Although initial ophthalmic examination revealed a unilateral neuroretinitis, investigation for infectious and non-infectious causes of neuroretinitis were negative. At our examination, retinal imaging (suggestive of bilateral involvement) along with the results of lumbar puncture (pleocytosis) and clinical findings was consistent with a diagnosis of Vogt-Koyanagi-Harada disease. The patient was treated with intravenous steroids with prompt resolution of her symptoms. Vogt-Koyanagi-Harada disease may present atypically and should be considered in the differential diagnosis of neuroretinitis.

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Introduction

Neuroretinitis (NR) is an inflammatory disorder characterised by optic disc oedema and subsequent formation of a macular star figure. It can be divided in idiopathic recurrent and infective, cat scratch disease (CSD) being the most common cause of the latter group. We report a case of Vogt-Koyanagi-Harada disease (VKHD) with bilateral markedly asymmetric involvement of retina, optic nerve, and choroid, causing unilateral NR.

Case report

A 42-year-old housewife was admitted to the local hospital because of painless visual loss in the left eye (OS), begun suddenly the week before, associated with headache and fever occurred 10 days before. She denied medications or drug or alcohol abuse. Contrast brain magnetic resonance imaging (MRI) scan of the head and orbit was negative for optic nerve enhancement, space-occupying masses, or signs of multiple sclerosis. The first ophthalmologic consultation revealed a best-corrected visual acuity (BCVA) of 20/20 in OD and 10/20 in OS, a quiet anterior segment in both eyes (OU), a normal intraocular pressure (IOP) in OU, and on retinography

(Figure 1), a normal retina with a mild peripapillary nerve fibre layer swelling and disc hyperaemia in OD, and in OS an optic nerve oedema with haemorrhages associated with a partial macular star. She was diagnosed with NR. Laboratory investigation was negative for *Bartonella henselae* as well as for other causes of neuroretinitis: syphilis, tuberculosis, toxoplasmosis, brucellosis, leptospirosis, herpes, and Lyme disease. Investigation for sarcoidosis (high-resolution computed tomography [CT] scan of the chest, angiotensin-converting enzyme [ACE] and lysozyme levels) was also unremarkable. She was diagnosed with idiopathic neuroretinitis and sent to our hospital for a second opinion. The patient denied any trauma, ocular surgery, or recent vaccination. She complained of painless visual loss in OS associated with fever and headache, whereas in OD she was asymptomatic. She denied kitten in the house but her neighbour had two cats. At our visit, BCVA was still 20/20 in OD but dropped to finger count in OS with a left relative afferent pupillary defect (RAPD). Slit-lamp examination of the anterior segment was normal in OU as well as IOP. Although not apparent (or not present) on the first retinography, on detailed fundus examination we noticed retinal swelling mainly at the posterior pole in OS, which may have accounted for the worsening in

Figure 1. Colour retinography of the fundus when the patient presented the first time to the local hospital shows in OD a normal retinal with a mild peripapillary nerve fibre layer swelling and disc hyperaemia, and in OS an optic nerve oedema with haemorrhages associated with a partial macular star.

visual function. There were no signs of retinal vascular involvement (e.g., periphlebitis), whereas some cells could be seen in the central vitreous in OU. The patient refused to perform visual field testing. Optical coherence tomography (OCT) of the macula ([Figure 2](#)) showed mild chorioretinal folds in OD and severe chorioretinal folds associated with multiple serous retinal detachments in OS, consistent with the clinical findings on fundus examination. The choroid was thickened as well. Ultrasonography confirmed bilateral choroidal thickening (OS > OD) and no sign of posterior scleritis. Fluorescein angiography (FA) showed pooling of fluorescein in the areas of serous retinal detachments in OS with mild disc leakage in OD. On the contrary, indocyanine green (ICG) angiography ([Figure 3](#)) disclosed bilateral

involvement of the choroid with focal areas of delay in perfusion. Repeated antibody test for *B. henselae* was negative. Lumbar puncture was performed: opening pressure was within normal limits, whereas analysis was significant for pleocytosis (32/mL). Protein and glucose were also normals. Repeated contrast brain MRI scan was negative for masses, cerebral sinuses involvement, and meningeal enhancement but showed a mass at the posterior pole of the left eye as compared with the right eye ([Figure 4](#)), consistent with retinal detachment. The patient was treated with intravenous steroids for 5 days and then switched to oral steroids, with prompt resolution of her symptoms and a rapid recovery of the VA. After a month, the retinal involvement greatly improved as well as OCT ([Figure 5](#)) and BCVA increased to 20/20 also in

Figure 2. OCT shows mild chorioretinal folds in the right eye, whereas in OS there are chorioretinal folds associated with multiple exudative detachments of the neural retina associated with retinal septae, very suggestive of VKHD.

Figure 3. Top row: FA shows severe disc leakage OS associated with pooling of the dye in the areas of exudative retinal detachments. OD is relatively silent, showing a mild disc leakage with hyperfluorescence. Bottom row: ICG angiography shows multiple choroidal filling defects concentrated at the posterior pole in OU.

Figure 4. Contrast brain MRI scan of the patient (T2 weighted). Note thickening of the posterior pole of the left eye, consistent with serous retinal detachment. Right eye is normal.

Figure 5. One month follow-up. Top row: Retinography shows absence of optic disc oedema. A mottling of the retinal pigmented epithelium (RPE) is evident in OS, with a mild temporal disc pallor. Bottom row: OCT is normal in OD whereas in OS there is dystrophy of the RPE associated with a normal macular profile.

the affected eye. At sixth month of follow-up, the patient is stable with no recurrence and a BCVA OU 20/20. Nine months after our examination, poliosis

was evident in OD (**Figure 6**). The clinical presentation with neurological involvement along with retinal imaging and the results of lumbar puncture was

Figure 6. Poliosis in OD, present at 9 months after onset.

consistent with a diagnosis of complete Vogt-Koyanagi-Harada disease (VKHD). As far as we know, this is the first report of VKHD presenting as unilateral neuroretinitis.

Discussion

Vogt-Koyanagi-Harada disease is a severe, bilateral, granulomatous panuveitis with serous retinal detachments, disc oedema, and vitritis associated with headache, nausea, meningismus, alopecia, vitiligo, and hearing loss.¹ It presents clinically in four different phases: prodromal, acute uveitic, convalescent, and chronic recurrent. However, not all patients present with the complete constellation of these extraocular findings and prodromal phase may be absent; hence, diagnosis may be difficult or in doubt in some cases, until signs and symptoms finally develop and fulfil the diagnostic criteria of this syndrome. Ancillary tests, especially multimodal imaging, can reliably provide supportive evidence for the diagnosis of early cases and atypical presentations,² as was the present case. Moreover, the occurrence of serous retinal detachments in the acute phase has a positive predictive value of 100 of being VKHD.³ New imaging techniques are continuously changing our view on many optic nerve and retinal disorders, and NR does not escape this revolution. In fact, since the first description by Theodore Leber in 1916,⁴ our understanding of NR has continued to evolve and a number of articles have reported additional cases of NR due to various infective agents. Nomenclature has also evolved from the term “stellate retinopathy” to optic disc oedema with a macular star (ODEMS) proposed by Brazis and Lee.⁵ During this period, retinal imaging has improved as well, and OCT along with FA is commonly used in ophthalmic departments to improve follow-up of patients and possibly to collect additional information on the pathogenesis of many retinal disorders. Indocyanine green angiography is more sensitive than FA in detecting choroidal vasculature abnormalities and has been used to clarify the pathophysiology of NR in some cases. Kitamei et al.,⁶ using FA and ICG angiography coupled with OCT, demonstrated focal leakage of dye from a single arteriole on the disc surface in a case of idiopathic NR. In fact, correlating results from multimodal retinal imaging may further our understanding of this disorder.

The case presented here is unique because eye involvement was apparently unilateral as suggested by retinography and by patient's complaint, but multimodal imaging was paramount in detecting a bilateral, albeit highly asymmetric, involvement of both eyes, especially of the choroid. It remains unexplained why optic disc involvement was so different between both eyes, causing an evident optic disc oedema in OS and only a mild disc hyperaemia in OD. The macular incomplete star developed in the papillomacular bundle because of the retinal and peripapillary exudation from the optic nerve. Neuroretinitis so far has been described in a variety of infectious and non-infectious disease. Particularly important is cat scratch disease (CSD) caused by *Bartonella henselae*, which accounts for two thirds of cases in one series,⁷ and which was excluded here by repeated laboratory tests. A new classification of NR has been proposed, basing on causative agent and recurrence over time: idiopathic NR, cat scratch NR, and recurrent-idiopathic NR.⁸ In conclusion, the authors suggest performing ICG angiography in every case of idiopathic, chronic, or recurrent NR, since it may provide additional informations and sometimes the main clue to the diagnosis, especially in those cases of NR labelled so far as idiopathic.

To the knowledge of the authors, this is the first case of unilateral neuroretinitis caused by Vogt-Koyanagi-Harada disease, and also the first case of Vogt-Koyanagi-Harada disease presenting as neuroretinitis.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

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