

An acute on chronic presentation of Vogt-Koyanagi-Harada (VKH) disease

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Abstract

VKH disease is an idiopathic chronic granulomatous inflammatory disorder. It has ophthalmic, neurological and cutaneous manifestations. The clinical picture is variable and dependent on the stage of presentation. We report on a patient who presented with a mixed picture of early and late onset symptoms with clinical findings of acute on chronic inflammation.

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Keywords

Vogt-Koyanangi-Harada disease; granulomatous inflammation; ophthalmology; neurology.

Case report

A 31-year-old female of Bangladeshi origin presented with a month's history of worsening headaches, photophobia, neck stiffness and reduced vision.

Visual acuity was 6/36 in either eye with no pinhole improvement. Examination revealed an active pan uveitis. Fundoscopy revealed marked optic disc hyperaemia, marked peripheral hypopigmentation of the choroid and associated hyperpigmented patches of retinal pigment epithelium (RPE) clumping. There was also bilateral chronic cystoid macular oedema (Fig. 1(a) and (b)). These findings were confirmed on fundus fluorescein angiography (FFA) (Fig. 2(a) and (b)).

Syphilis serology was negative as was screening of serum ACE for sarcoid. Routine haematology and chemistry were normal. Cerebrospinal fluid (CSF) analysis showed normal protein and glucose with no oligoclonal bands. There was no growth on CSF culture and Ziehl Neelsen (ZN) and gram staining were negative. Computed tomography (CT) brain scan was also normal.

Diagnosis

A diagnosis of VKH was made on the basis of her history of meningismus and fundus findings. She was started on high dose oral prednisolone 80 mg per day and reviewed weekly. She was also started on a proton pump inhibitor and bisphosphonate in view of the prolonged course of steroids that was envisaged. The meningismus settled down rapidly over a number of days. Over the course of 4 months vision improved to 6/12. She was then very slowly taken off the steroids and has remained stable ophthalmically.

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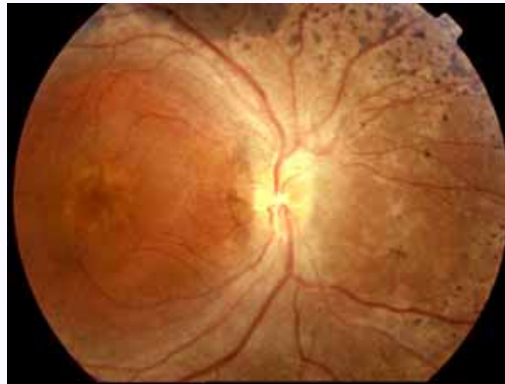
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(a)



(b)



Fig. 1. Fundoscopy revealing marked optic disc hyperaemia, marked peripheral hypopigmentation of the choroid and associated hyperpigmented patches of retinal pigment epithelium (RPE) clumping and bilateral chronic cystoid macular oedema.

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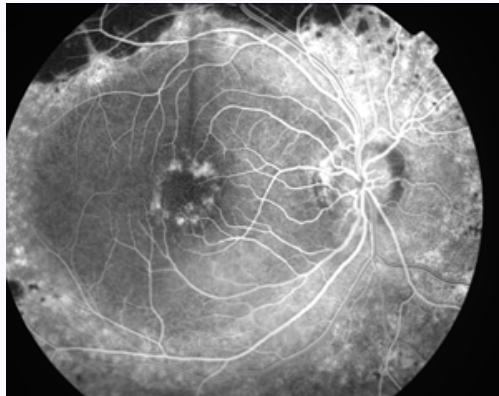
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(a)



(b)

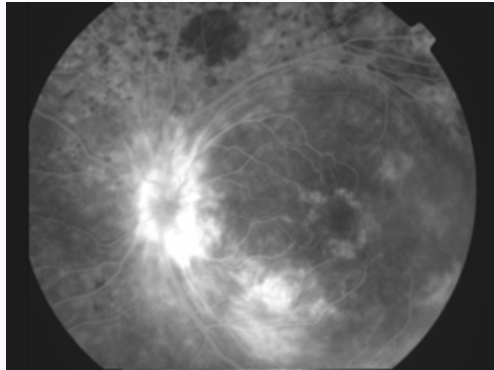


Fig. 2. Fundus fluorescein angiography (FFA) confirming the same features shown in Fig. 1.

Clinical evidence and unusual features

The aetiology of VKH disease is generally believed to have an immunologic basis^[1]. Autoimmune T cell activity against an unknown melanocytic antigen, a tyrosinase or tyrosinase-related protein, is most commonly suggested^[2,3]. Proposed triggers include cutaneous injury or viral infection^[4]. The diagnosis of VKH is dependent on the stated criteria which include the exclusion of several other entities^[5]. The similarity to sympathetic ophthalmia both in the laboratory findings and the clinical picture mean that a history of trauma or surgery must be excluded^[6]. Other conditions which also share a similar clinical picture and therefore must be excluded are posterior scleritis, syphilitic and tuberculous uveitis, Lyme disease, sarcoid, intraocular lymphoma, central serous chorioretinopathy and uveal effusion syndrome^[7,8]. A full work-up including serology, angiography and B scan ultrasonography is usually mandatory.

Ocular manifestations of the disease vary depending on the stage of presentation. Four phases are commonly described. An initial prodromal phase with meningismus or auditory system involvement occurs. Diffuse choroiditis marks the anterior uveitic phase and can manifest as exudative retinal detachment and papillitis. The chronic phase has many manifestations which are not specific for the disease. The two possible exceptions to this are the 'sunset glow fundus' due to choroidal depigmentation and the 'Sigiura sign' which is perilimbal vitiligo. Less specific signs can be used to make the diagnosis if depigmentation is absent and these include retinal pigment epithelium (RPE) clumping or migration, chronic or recurrent anterior uveitis and nummular chorioretinal scars. Evidence of involvement of other systems of the body will unequivocally establish the diagnosis of VKH^[8-10].

Inflammation of the meninges and auditory system manifesting as meningitis, dysacusis, or tinnitus are diagnostic findings.

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Our patient exhibits an acute presentation of definite VKH as adhered to by the revised diagnostic criteria. This is, however, on a background of late presentation VKH. She responded well to steroids and has thus far remained relatively free of complications of treatment except for a slight tendency towards a cushingoid appearance. We feel her case is unusual in that she manifested both acute and late diagnostic criteria at presentation.

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