44 year old Italian-American male presents with a three day history of profound vision loss in one eye.

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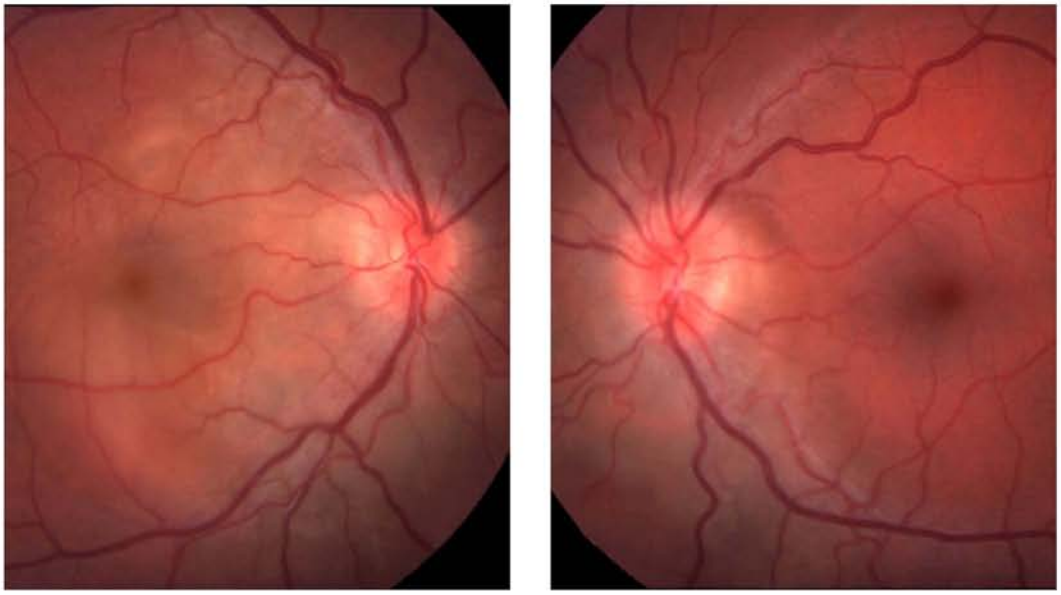
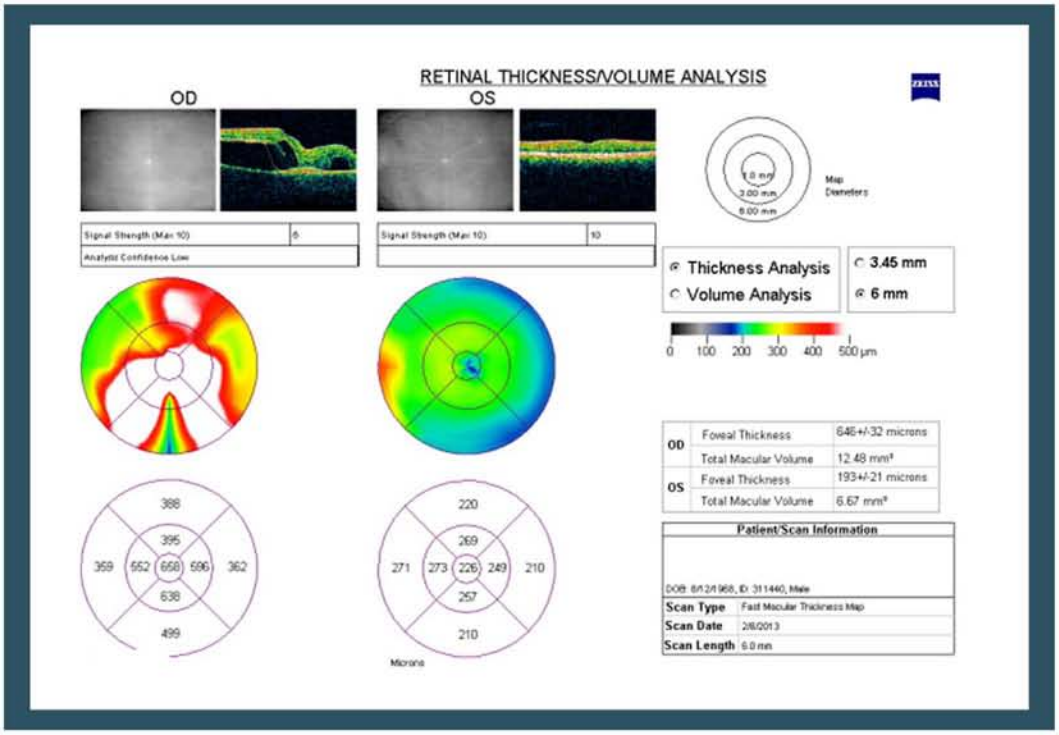
This healthy male presented with painless vision loss in the right eye. His review of systems was unremarkable, and he was on no medications. Visual acuity was count fingers in the right eye and 20/25 in the left. The anterior segments and vitreous were normal. The discs were subtlely edematous. A large, irregular serous detachment involved much of the nasal macula in the right eye, with a small serous detachment noted nasal to the disc in the left eye.



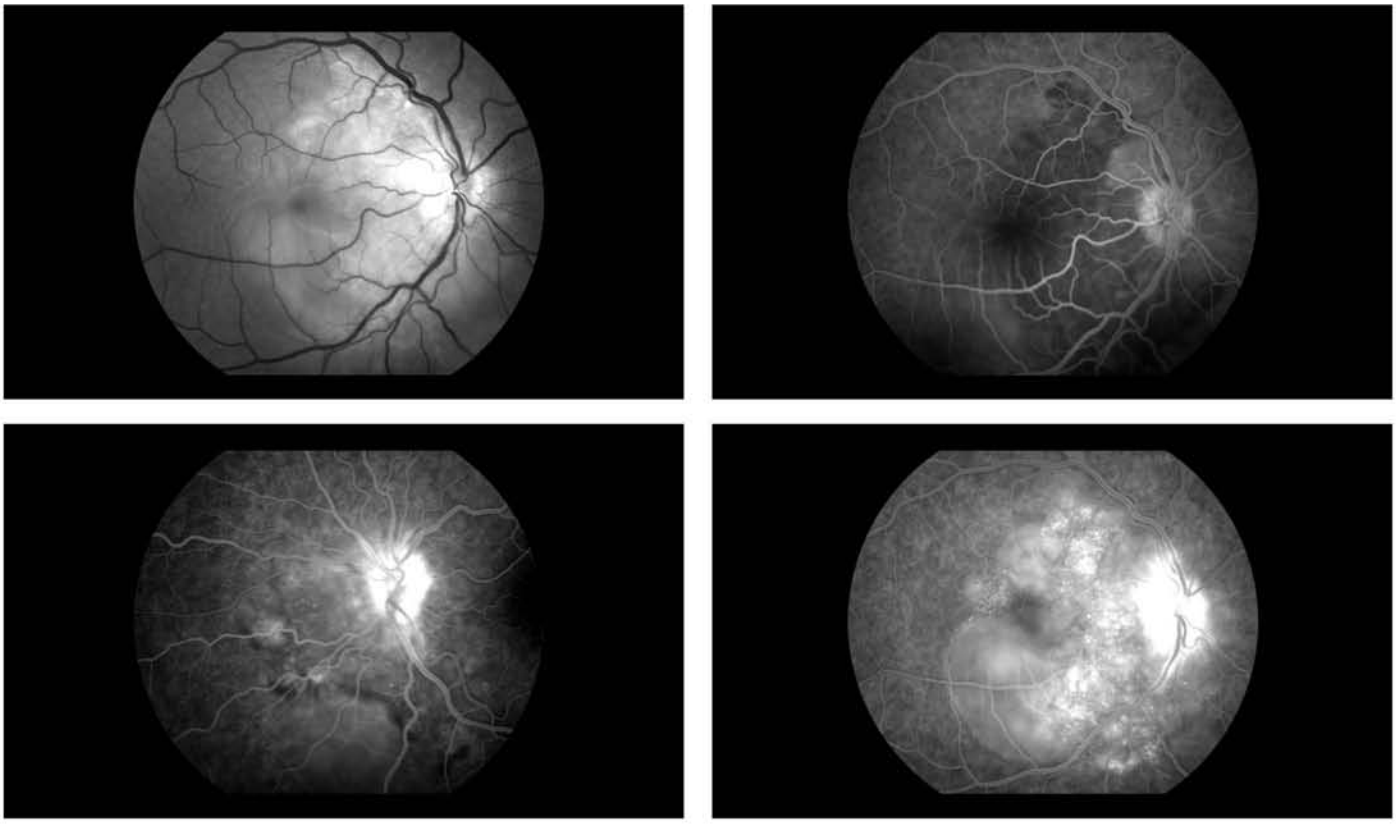


A time domain OCT revealed a large bi-lobed neurosensory detachment in the right eye.

A fluorescein angiogram revealed areas of delayed choroidal perfusion, multifocal pinpoint RPE leaks, areas of placoid hyperfluorescence, pooling of subretinal fluid and optic nerve staining bilaterally.

A cell blood count, ESR, RPE/FTA, HIV antibodies, serum lysozyme, and PPD were unremarkable.

On the basis of the clinical exam and negative serologic studies, a diagnosis



of incomplete Vogt-Koyanagi-Harada (VKH) disease was made.

VKH disease is a multi-systemic disorder characterized by granulomatous Panuveitis and exudative retinal detachments that is often associated with neurologic and cutaneous manifestations.

Asians, Middle Eastern and Hispanics are more susceptible. VKH is rare in people of European or African descent.

An immunologic predisposition is suspected with HLA Haplotype DR4 having the strongest association. There is no apparent sexual predisposition, and most patients present in the second to fifth decade. Children are rarely diagnosed.

The pathophysiology is thought to be T helper cell mediated autoimmune attack of melanocytes in the uvea, skin, inner ear, and central nervous system.

Apparently 50% of patients experience a *prodromal phase* which may include fever, headache, nausea, vertigo, orbital pain, or tinnitus. In the prodromal phase 80% of patients have cerebral spinal fluid abnormalities which include lymphocytic pleocytosis or elevated protein.

The *acute uveitic phase* is often bilateral with posterior uveitis, disc edema, and serous retinal detachment. Later anterior uveitis with iridocyclitis, iris nodules, and mutation-fat precipitates are common. Many patients experience otolaryngologic manifestations including hearing loss, tinnitus, and vertigo.

In the *chronic phase* retinal pigment epithelial migration and Dalen-Fuchs nodules appears with vitiligo, poliosis, an alopecia. A minority of patients experience a *recurrent phase* with repeated retinal detachment, glaucoma, choroidal neovascularization, and subretinal fibrosis.

Early and aggressive treatment with corticosteroids and/or immunosuppressive drugs is essential. Treatment should be continued for a minimum of 6 to 12 months before considering withdrawal. Auditory system disorders respond well and are generally completely reversed in two to three months.

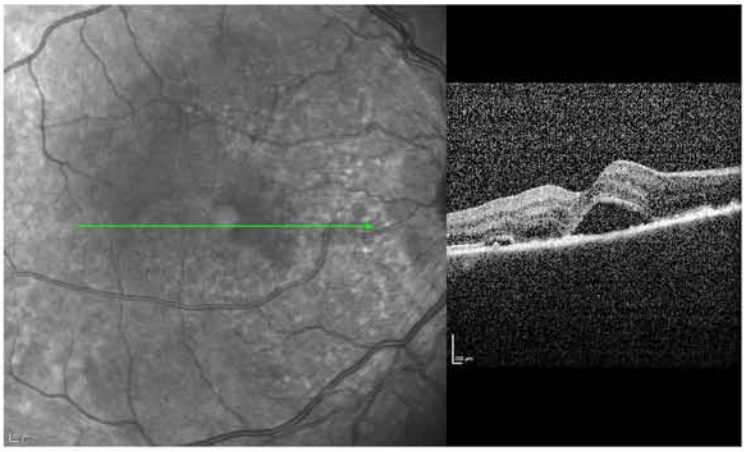
The visual prognosis is variable. The most important prognostic factors include good visual acuity one month after starting treatment, early high-dose steroids, and age of onset.

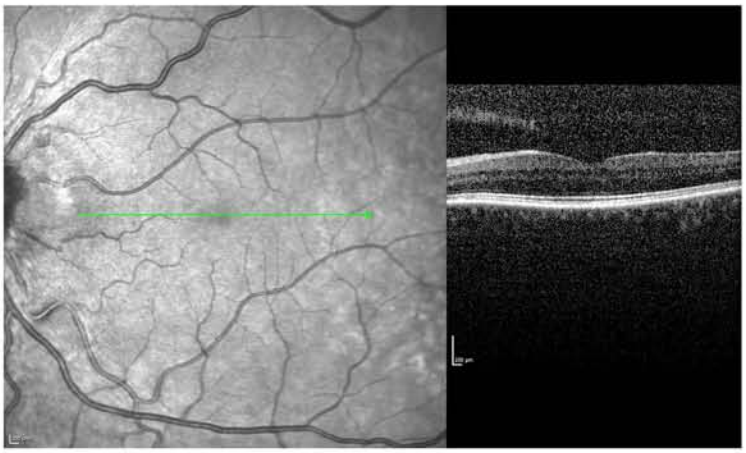
The differential diagnosis includes syphilis, TB, toxoplasmosis, fungal infections, AIDS, sympathetic ophthalmia, and severe hypertension.

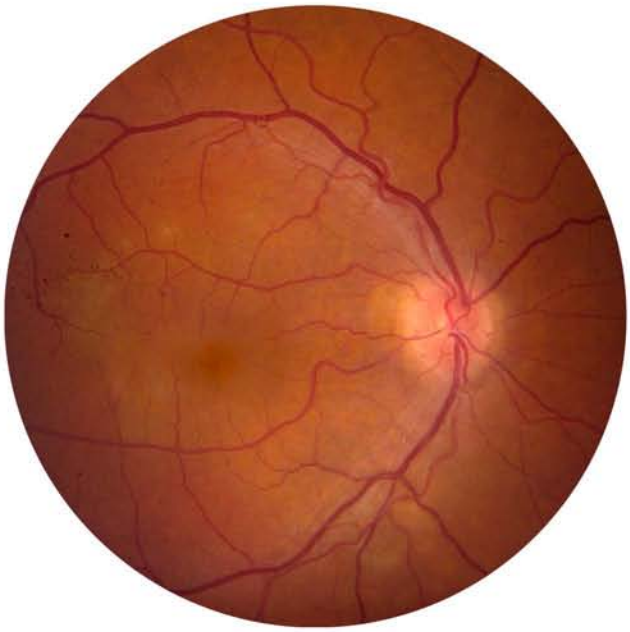
Recent advances in OCT technology employ a longer wavelength of light source permitting a more

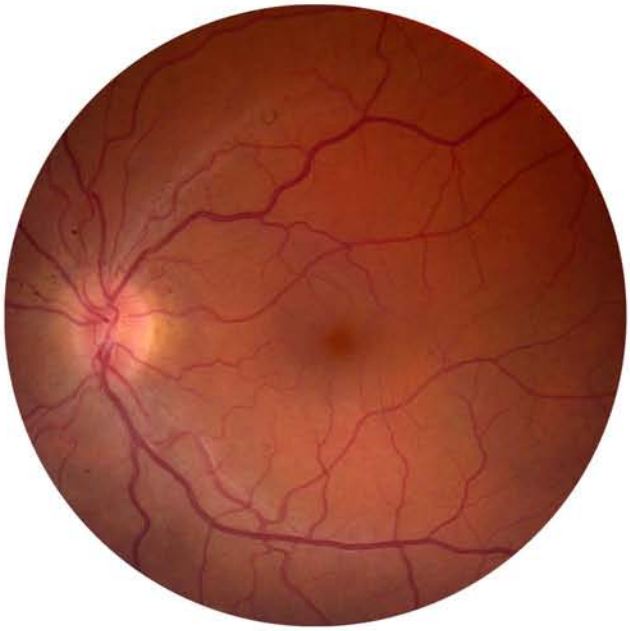
precise visualization of the choroid. This precision may be useful in assessing VKH disease severity, enabling titration of treatment.

At four months our patient on steroids was doing well. The right eye had improved from count fingers to 20/40 with a small subfoveal serous detachment. The left eye remained 20/25. The disc edema had resolved.









The Retina Institute participates in numerous national clinical trials. Visit the Studies section on our website at tri-stl.com for information regarding these trials and patient enrollment.