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Grand Rounds

A 41-year-old man with bilateral, painless loss of vision

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History

A 41-year-old South Asian man presented at an outside clinic affiliated with MedStar Georgetown University Hospital with painless, progressive vision loss in both eyes. His symptoms started with constant blurred vision and floaters in his left eye 4 weeks earlier, followed 2 weeks later by blurred vision in his right eye. Past medical, surgical, and ophthalmic history were unremarkable. He was not taking medications and had no known drug allergies. Review of systems was negative for skin rashes, joint pains, fevers, auditory, and neurological symptoms. Family history was significant for cataracts. There was no known history of ocular inflammation. He reported drinking alcohol socially and smoking two cigarettes daily.

Examination

Uncorrected visual acuity was 20/40 in the right eye and 20/200 in the left eye, without improvement on pinhole testing. Intraocular pressure by applanation tonometry was 16 mm Hg in the right and 17 mm Hg in the left eye. There was no afferent pupillary defect. Anterior segment examination showed 2+ cell and 1+ flare in both eyes. Fundus examination showed 1+ vitreous cells in both eyes, with choroidal folds more prominent in the left eye, subretinal fluid in both eyes, and an inferior exudative detachment in the left eye (Figure 1).

Ancillary Testing

Blood pressure in clinic was 110/80. Retinal optical coherence tomography (OCT) revealed choroidal folds and subretinal fluid in both eyes, greater in the left eye (Figure 2). Fundus autofluorescence showed multiple areas of hypoautofluorescence, with pockets of serous detachments in the macula of left eye (Figure 3). Fluorescein angiography identified pinpoint areas of leakage, greater in the left eye (Figure 4). B-scan ultrasound showed a thickened choroid and an exudative retinal detachment in the left eye (Figure 5). CBC with differential, ANA panel, RPR, FTA-ABS and Quantiferon gold were normal. A chest X-ray revealed a benign calcific nodule but was otherwise normal.

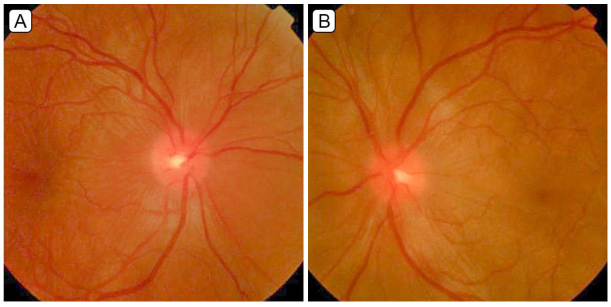

Figure 1. Fundus photographs of the right eye (A), showing mild optic nerve edema, retinal striae, and localized subretinal fluid in the macula, and the left eye (B), showing mild optic nerve edema and serous retinal detachments in the macula and around the optic nerve.

Treatment

The patient was diagnosed with probable Vogt-Koyanagi-Harada (VKH) disease based on the constellation of signs and symptoms. The presence of panuveitis made hypertensive retinopathy and central serous chorioretinopathy less likely. Similarly, the B-scan findings and lack of ocular trauma or eye surgery ruled out posterior scleritis and sympathetic ophthalmia (Figure 5). The patient was started on 60 mg of oral prednisone and prednisolone acetate 1% drops to each eye every 6 hours. His vision rapidly improved after initiating treatment. Six weeks later his visual acuity was 20/20-2 in both

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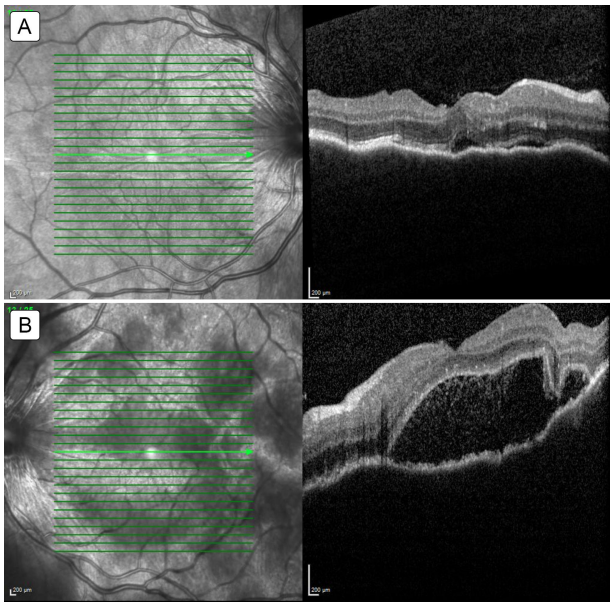

Figure 2. Retinal optical coherence tomography (OCT) showing choroidal folds and subretinal fluid in the right eye (A) and choroidal folds, loculated pockets of subretinal fluid, and hyper-reflective dots in the subretinal space in the left eye (B).

eyes, with resolution of anterior and posterior inflammation and serous detachments. Ten weeks after treatment, OCT of both eyes showed resolution of subretinal fluid (Figure 6). He was tapered off oral prednisone over 6 months. The patient did not experience any systemic symptoms during the course of disease and remained free of recurrent inflammation on his last follow-up examination, 11 months after his initial presentation.

Differential Diagnosis

The differential diagnosis for bilateral exudative retinal detachments includes hypertensive crisis, VKH, sympathetic ophthalmia, central serous chorioretinopathy, uveal effusion syndrome, posterior scleritis, primary intraocular lymphoma, uveal lymphoid infiltration, bilateral diffuse uveal melanocytic proliferation, tuberculosis-associated uveitis, and sarcoidosis.[1,2]

Diagnosis and Discussion

VKH is an autoimmune disorder that affects tissues pigmented with melanin. This disease can affect the visual, auditory, neurologic, and integumentary systems. It characteristically manifests as a bilateral granulomatous uveitis with accompanying loss of vision and other extraocular manifestations, such as pleocytosis, vitiligo, and tinnitus.[3]

VKH more often affects people with darker skin pigmentation, including Asian, Hispanic, Native American, and Middle Eastern ethnic groups.[1,3] The incidence of VKH is variable, accounting for approximately 4% of uveitis cases in the United States and 8% in Japan.[3] Most studies report a slight female preponderance.[1] Affected individuals typically range in age from 20 to 50 years.[4]

To diagnose VKH, patients must not have a history of ocular trauma or eye surgery. The disease is classified as (1) complete, (2) incomplete, or (3) probable. Complete disease is associated with bilateral ocular, neurologic, auditory, and integumentary symptoms. Patients with incomplete VKH have bilateral ocular involvement and neurologic, auditory, or skin findings. Patients with probable VKH present with only bilateral ocular symptoms.[1]

VKH is usually diagnosed based on clinical appearance. Patients almost always present with bilateral exudative retinal detachments, although there may be asymmetry between the eyes. Fluorescein angiography shows multiple punctate hyperfluoresecent foci, with pooling in areas of serous retinal detachment.[3] Focal areas of retinal pigment epithelium loss may give rise to window defects, and optic disc leakage is often present. Indocyanine green angiography demonstrates delayed choroidal perfusion, with multiple hypercyanescent foci of leakage in areas of exudative retinal detachment.[3] Retinal OCT findings include exudative detachments, which often form loculated pockets of subretinal fluid, choroidal excavations, the presence of fibrin in the subretinal space and a thickened choroid. B-scan ultrasound demonstrates choroidal thickening with or without an exudative retinal detachment. Cerebrospinal fluid may show lymphocytic pleocytosis.

There are four stages of VKH: (1) prodromal, (2) acute uveitic, (3), convalescent, and (4) chronic recurrent. During the first stage, patients present with viral symptoms, such as headaches, nausea, dizziness, and fever. During the acute uveitic stage, patients have decreased vision due to bilateral posterior uveitis and serous retinal detachments. During the convalescent stage, skin findings such as vitiligo, alopecia, and poliosis may be seen. Signs of hyper- or hypopigmentation in the eye may be present, including a “sunset-glow” fundus, depigmented chorioretinal lesions, called “Dalen Fuchs nodules,” and perilimbal vitiligo, known as Sugiura’s sign.[1,2,4] The chronic recurrent stage is characterized by episodes of panuveitis and development of iris nodules, posterior synechiae, glaucoma, cataracts, subretinal fibrosis, and chorioretinal atrophy.[1,4]

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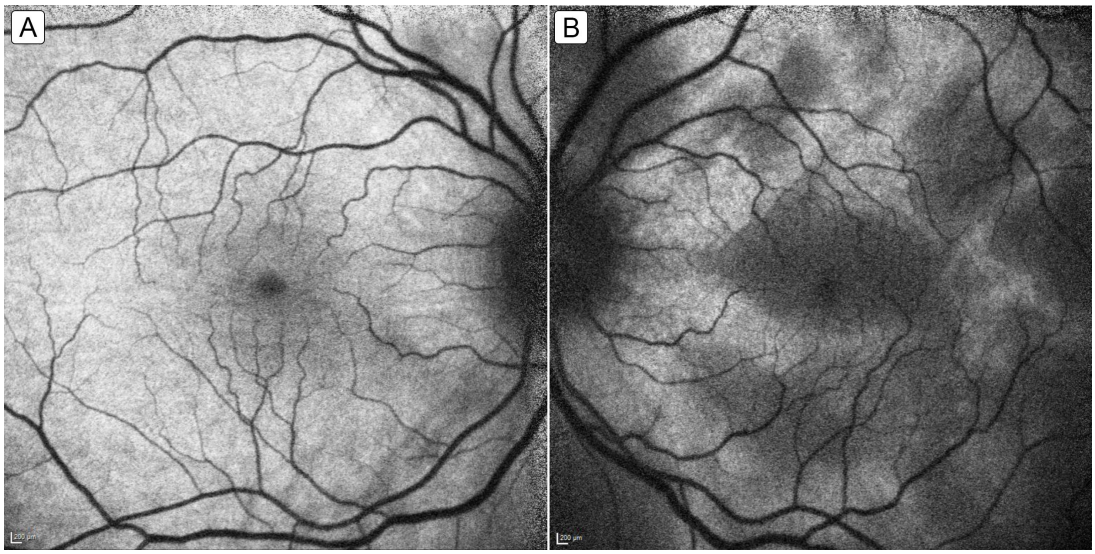

Figure 3. Fundus autofluorescence. The right eye (A) shows mild hypo-autofluorescence around the fovea and optic nerve; the left eye (B), patches of hypo-autofluorescence corresponding to pockets of serous detachments in the macula.

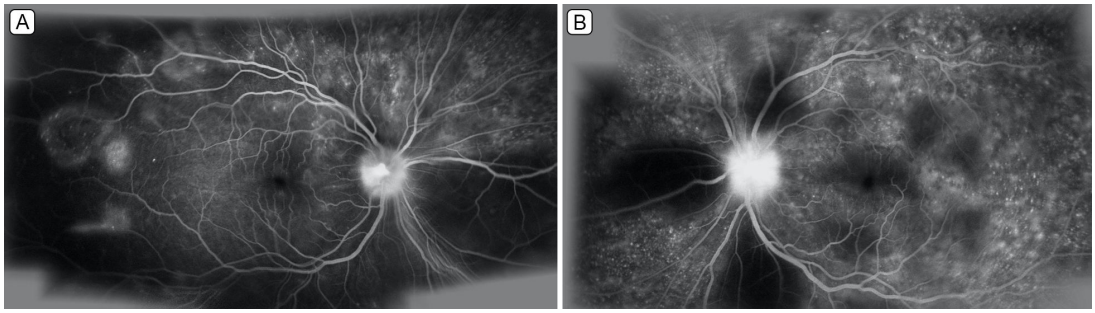

Figure 4. Late-phase fluorescein angiography showing multiple pinpoint foci of leakage in the macula and retinal periphery with optic nerve leakage, greater in the left eye (B) than in the right eye (A). There are areas of hypo-fluorescence due to blockage from serous detachments.

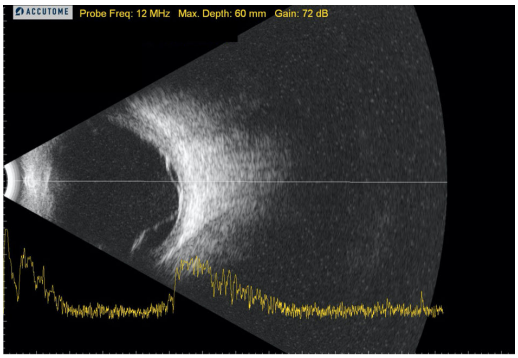

Figure 5. B-scan ultrasound of the left eye showing a thickened choroid and exudative retinal detachments; there was no evidence of the classic T sign seen in posterior scleritis.

Standard treatment for VKH involves initiating oral prednisone (1–1.5 mg/kg/day) or intravenous methylprednisolone (1 g daily for 3 days) followed by a switch to oral prednisone, with a gradual taper over 3–6 months.[3] Relapses may occur if the patient is taken off corticosteroids too early. In cases where inflammation is inadequately controlled or recurrent, immunosuppressive therapy is usually initiated.

Some researchers have advocated using antimetabolites, cyclosporine, and biological agents as first-line and maintenance therapy for VKH, suggesting they may lead to better long-term visual outcomes.[1,3,4] Periocular steroid injections and intravitreal fluocinolone implants can be used as adjunctive therapy.[3,4]

With adequate treatment with corticosteroids, the prognosis for VKH is generally good. By following the treat-

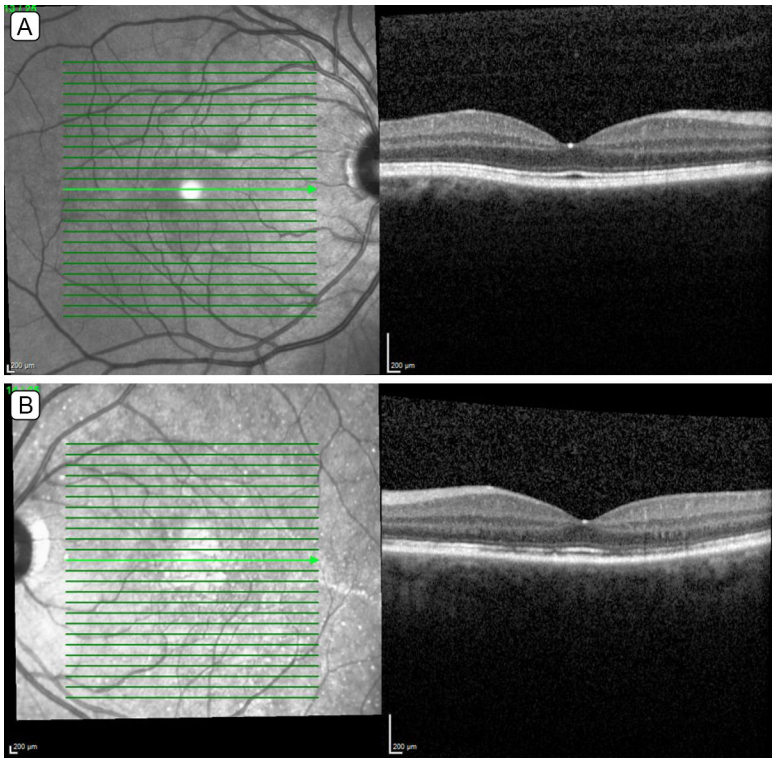

Figure 6. Retinal OCT 10 weeks after treatment demonstrates resolution of subretinal fluid in both the right (A) and left (B) eyes; the left eye shows patchy loss of the ellipsoid zone.

ment regimen with the necessary follow-ups, the majority of patients achieve a visual acuity of 20/40 or better and avoid complications, such as cataract, glaucoma, choroidal neovascularization, and subretinal fibrosis.[1]

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