What You Should Know About Vogt – Koyanagi – Harada Disease

By David J. Browning, MD, PhD

Vogt - Koyanagi - Harada Disease (VKH) is a disease of several body systems, including the eyes, brain, and skin. It causes blurred vision from inflammation of the choroid and leakage of fluid under the retina. It can cause headaches and pain with neck motion due to inflammation of the meninges, which coat the brain and spinal cord. Skin depigmentation (vitiligo), can occur, as well as hair loss (alopecia), and eyelash depigmentation (poliosis).

What Causes VKH?

VKH is an autoimmune disease. The body's immune cells attack cells in the body that contain melanin pigment. Usually immune cells distinguish cells belonging to one's self from invading cells (like viruses and bacteria) that need to be eradicated. In autoimmune diseases like VKH, recognition goes awry, and unwanted attacks on self cells occur. These episodes can occur out of the blue, or may be triggered by injuries to skin or by viral infections.

Eye Manifestations

In the eye, VKH causes inflammation of the front of the eye (iritis) and of the back of the eye (choroiditis). Taken together, both types of inflammation comprise the general diagnosis of uveitis, a general term for ocular inflammation. Uveitis secondarily produces fluid leakage under the retina (serous retinal detachment), scar tissue formation, lens clouding (cataract), occasionally abnormal blood vessel growth, and high pressure inside the eye (glaucoma). Figure 1 shows a normal retina. Figure 2 shows the retina of a patient with VKH. Blisters of subretinal fluid are present.

Figure 1. Normal Retina

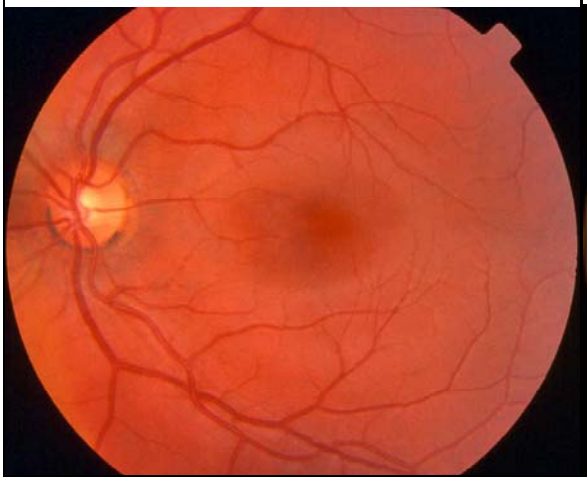
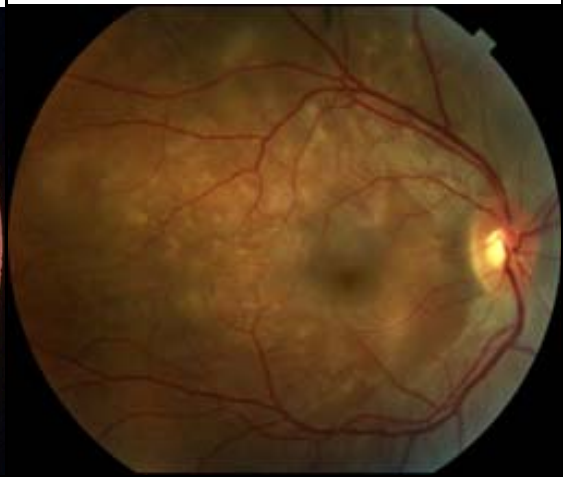


Figure 2. Retina with VKH



Who Gets VKH?

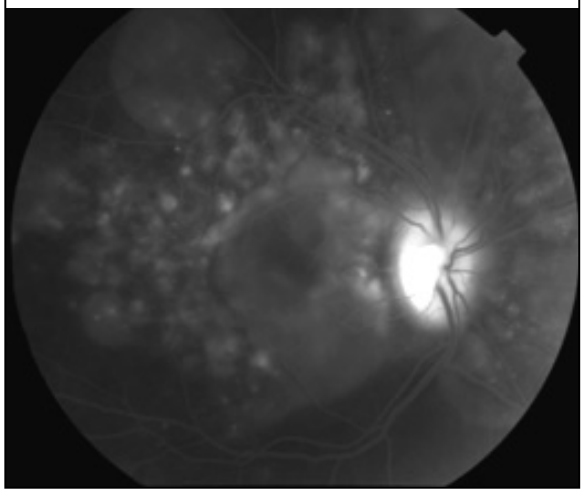
Certain genetic characteristics predispose a patient to VKH. People with genes that code for the HLA-DR4 and –DR53 antigens develop VKH more frequently than patients not possessing these genes. These genes tend to be more common in patients with Asian or American Indian ancestry.

Women seem to develop VKH more than men. The ratio of women to men is roughly 3:1. Although the onset of disease may be in childhood, usually the first attack occurs between the ages of 20 and 30. Both eyes are usually affected, although often somewhat asymmetrically, and disease recurrences are common.

Diagnostic Testing

Although most cases of VKH are fairly straightforward to diagnose, sometimes confusing pictures are seen, and clinical testing may be helpful. Blood tests to rule out syphilis, sarcoid, Lyme disease, and hematologic diseases may be helpful. Sometimes a spinal tap is done to check for lymphocytes in the spinal fluid. Pictures of the retina may be taken to show swelling and fluid collections (OCT) or to show areas of leakage (fluorescein angiography). Figure 3 shows areas of leakage under the retina in a patient with VKH. Ultrasound testing may be helpful to show swelling of the choroid.

Figure 3. Fluorescein Angiogram



Treatment

Almost all cases of VKH are first treated with high dose steroids by mouth or intravenous injection. Gradually the dosage is tapered when the disease is controlled. Tapering must be gradual to prevent recurrence, and regular monitoring is necessary to detect side effects such as decreased bone density or intraocular pressure elevation. In certain cases, VKH does not respond to steroids or the side effects of steroids become intolerable. In such cases, immunomodulating drugs including azathioprine, cyclosporine, or chlorambucil may be used. These drugs require regular laboratory testing to make sure that bone marrow does not become dangerously suppressed or kidney function damaged.

Final Comments

Although a serious and sight threatening disease, VKH can be successfully controlled with powerful drugs. Careful follow-up and involvement of internists,

neurologists, and other doctors in the care of patients can improve the chances of a good outcome.

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