AMERICAN UVEITIS SOCIETY

Vogt-Koyanagi-Harada Disease

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by Russell W. Read, MD
Assistant Professor of Ophthalmology and Pathology
University of Alabama at Birmingham
Birmingham, AL, USA

Introduction

Vogt-Koyanagi-Harada disease, or VKH, is an inflammatory condition that affects both eyes, as well as the inner ear, skin, hair, and coverings of the brain. Certain groups of individuals appear to be predisposed to the development of VKH, such as Asians (including East and Southeast Asians and Asian Indians), Middle Easterners, Native Americans, and Hispanics. A common ancestry among these groups is believed to exist, with groups now living in the New World having arrived after crossing a land bridge across the Bering Strait. Individuals of Caucasian and African heritage may also develop VKH, though much less commonly.

History

VKH has been known for over 1000 years. The name of the condition comes from three physicians who described the different phases of the condition, though at that time it was not realized that the features described were part of the same condition. Later reports brought this condition together under the name of Vogt-Koyanagi-Harada disease.

Course of Disease

At the onset of VKH, patients frequently note symptoms such as severe headache, neck stiffness, ringing in the ears (tinnitus), fever, and possibly scalp tenderness. Sometimes these symptoms are so severe that patients may be admitted to the hospital for meningitis (inflammation of the membranes surrounding the brain). Within a few days to weeks, patients note the onset of blurry vision and sometimes eye pain. Eye examination at this point classically reveals leakage of fluid under the retina, and inflammation of the optic nerve head (the nerve at the back of the eye which relays visual signals to the brain). The middle layer of the eye (the uvea, and specifically the choroid) is swollen.

Diagnosis and Testing

Unfortunately, there are no blood or X-ray tests that can tell an eye doctor without a doubt that a patient has VKH. Rather, VKH is diagnosed based on the history of the disease in a particular

patient in combination with the findings present on examination and testing. The eye doctor will obtain a complete history and carefully examine the eyes for acuity or vision, for eye pressure, and for the presence of inflammation in the eye, using special instruments which magnify the eye under bright light. Special testing such as a fluorescein angiogram or ultrasound may be performed. Since no tests exist to diagnose VKH absolutely, the eye doctor will likely perform blood tests and a chest X-ray to ensure that other diseases which may look similar to VKH are not present. Some of these other eye diseases which can mimic VKH include sympathetic ophthalmia, sarcoidosis, intraocular lymphoma, uveal effusion syndrome, and multifocal central serous chorioretinopathy.

Treatment
The main form of treatment of VKH is corticosteroids such as prednisone. High doses are typically required to bring the inflammation into control. Some patients may require admission to the hospital to receive corticosteroids through the vein. It is believed by some doctors that early, aggressive treatment may result in fewer complications and less likelihood of recurrent disease in the future. No well-designed clinical research trials have proven this, however. VKH can be a difficult disease to treat, and may require the use of immunosuppressive therapy to achieve the desired control.

Cause of Condition
While the cause of VKH is not yet completely understood, scientists believe that the condition is caused by an attack mounted by the patient's own immune system against areas of the body that contain melanin, a substance that gives hair, skin, and the eyes their coloration. Interestingly, the lining of the brain and the inner ear contain this substance as well, thus tying together the various sites that are involved in VKH, and lending weight to this theory.

Prognosis
When diagnosed early and treated appropriately, patients with VKH have a good chance of retaining useful vision. Some studies suggest that VKH may be more aggressive in children and African Americans.

Research and Future Outlook
Current research efforts for VKH are along two fronts: 1) Finding the exact target of the abnormal immune attack, and 2) Determining the best treatment protocols to control the disease quickly and preserve sight with the added benefit of preventing recurrences.