AUTOIMMUNE RETINOPATHY

Cause of Progressive Vision Loss

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Autoimmune retinopathy is an opthalmic disease causing progressive vision loss that may occur as a primary autoimmune condition or in association with various cancers

Immune-Mediated Disorders

Autoimmune and cancer-associated retinopathies are ophthalmic disorders in which autoantibodies damage the retina and its components, causing progressive vision loss.

The retina in the eye consists of seven layers of tissue through which the signals that result in vision are processed. Autoimmune retinopathies can occur rarely as distinct autoimmune disorders or more commonly as paraneoplastic syndromes. Paraneoplastic syndromes are remote effects (occurring away from the primary affected organ) that occur in about 15 percent of all cancers, usually those involving neurologic, dermatologic and endocrine systems.

Paraneoplastic syndromes aren't related to the primary cancer but are the result of an autoimmune process in which autoantibodies are formed against antigen proteins in tumor tissue. These antibodies then cross-react with or react with similar antigenic proteins in the body. In those disorders affecting the retina, cancer-associated retinopathy or CAR antibodies are found. CAR antibodies are primarily directed against recoverin, a 23-kd retinal protein that is expressed by some tumor cells. Cancer-associated retinopathies are the most common type of paraneoplastic syndrome seen.

Symptoms

Symptoms in both cancer-associated retinopathies and autoimmune retinopathies are similar. Patients typically develop a rapid, painless vision loss associated with photopsias. Photospias are light flashes that may occur together with floaters or separately. Photopsias are typically perceived in subdued lighting or darkness and range from subtle light twinkles to bright flashes that suggest neon signs or camera flashes. Symptoms in retinopathies usually occur in both eyes although one eye may be affected before the other, and symptoms may progress over weeks to months.

Signs and symptoms can vary depending on the retinal elements that are affected. Either the rods or cones or both elements may be affected. Individuals with cone involvement have symptoms of photosensitivity (light sensitivity), prolonged glare after light exposure (hemeralopia), reduced visual acuity and loss of vision. Patients with rod involvement have difficulty seeing in dim lighting (nyctalopia), prolonged dark adaptation and

peripheral field vision loss. In both cases, flashing lights, transient dimming of vision, and flickering, smoky or swirling vision are common symptoms. On examination, patients usually show reduced visual acuity and loss of color vision.

Who is Affected?

Men and women are affected equally. When the retinopathy is associated with cancer, the most common disorders are small-cell lung cancer, gynecological and breast cancers, melanoma, lymphoma, pancreatic, prostate, bladder, laryngeal, or colon cancers. Early in the course of the ophthalmic disease, findings on retinal examination may be normal, which makes diagnosis difficult. For a proper diagnosis, an electroretinographic examination is necessary and the results are confirmed by the presence of circulating CAR antibodies.

Although vision loss can be unrelated to cancer and occur as a primary autoimmune retinopathy, in most cases of cancer-associated retinopathy, vision loss can occur before the malignancy becomes apparent. In retinopathy associated with melanoma (melanoma associated retinopathy or MAR), eye symptoms do not usually occur until the melanoma is diagnosed and has begun to metastasize or spread.

Treatment

Corticosteroids are used to decrease antibody titers in autoimmune and cancer-associated retinopathies and help stabilize vision. However, they usually cannot reverse vision loss. Treatment for cancer in cancer-associated conditions doesn't usually alter the visual complications.

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