

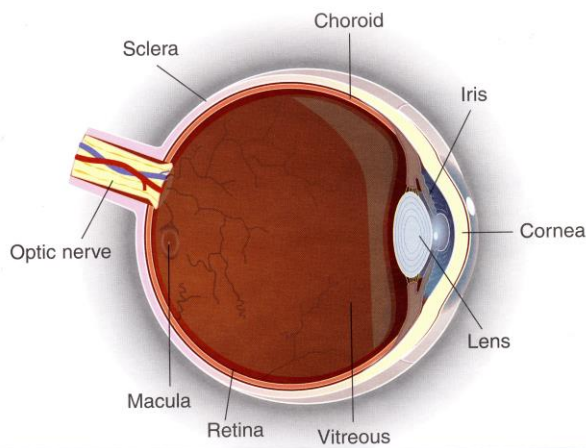
# What You Should Know About Cancer Associated Retinopathy, Melanoma Associated Retinopathy, and other Ocular Paraneoplastic Syndromes

*By David J. Browning, MD, PhD*

Genetic diseases of the retina, such as retinitis pigmentosa, cause most cases of night blindness. Much more rarely, a patient will develop night blindness with no previous history and no family history. When this occurs later in life and is associated with shimmering lights, ophthalmologists begin to suspect that the patient may have an underlying cancer that produces an antibody against the retina causing the problem, a situation called cancer associated retinopathy (CAR).

## Review of Normal Ocular Anatomy

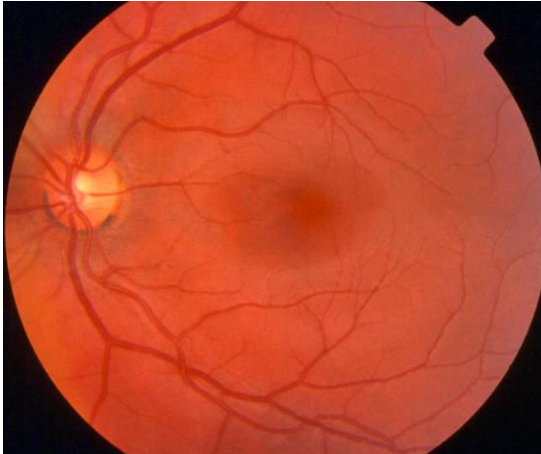
**Figure 1. Eye Anatomy**



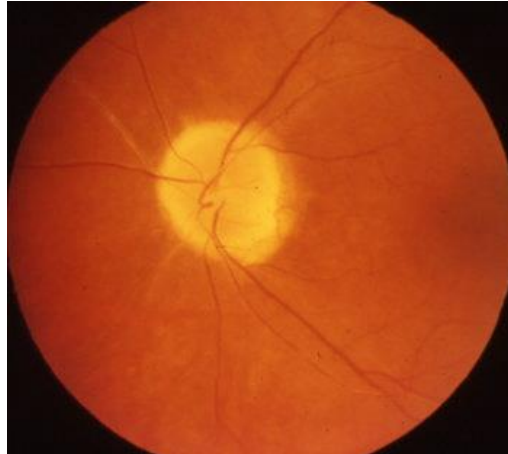
The retina is the neural lining of the back of the eye as shown in figure 1. It is a layered tissue with cells of different types – photoreceptors, bipolar cells, ganglion cells, and others. All cells have molecules on their surfaces that can be sensed and recognized by the roving cells of the immune system. These “identifier molecules”, called antigens, usually protect cells from immune attack because they are properly identified as being part of the body’s normal makeup. Sometimes the immune system wrongly identifies the cells as “not self” and an immune

response or a degenerative response is triggered producing an unwanted normal cell dysfunction and death. Figure 2 is a photograph of a normal, healthy retina. The retina of a patient with CAR may look normal or close to normal with only thinned blood vessels, but under the microscope photoreceptor or bipolar cell death can be seen. Figure 3 is an example of a retina with cancer associated retinopathy.

**Figure 2. Normal retina**



**Figure 3. CAR**



### **What Is A Paraneoplastic Syndrome?**

A neoplasm is another name for a cancer. A paraneoplastic syndrome is a medical condition in which a molecule made by a cancer causes disease in the body remote from the cancer. In the case of cancer associated retinopathy (CAR), an antibody against retinal cell antigens is aberrantly produced by the cancer cells and causes retinal cellular drainage and death. Many such antibodies have been discovered, but the most widely recognized is an antibody called anti-recoverin antibody. When doctors suspect CAR, blood is drawn to look for the presence of anti-recoverin and other antibodies. There are many cases in which the known antibodies causing CAR are not found. We think that these cases have antibodies against retinal antigens that have not yet been identified.

Ocular paraneoplastic syndromes include cancer associated retinopathy, melanoma associated retinopathy, a rarer condition of optic neuropathy, and a myasthenia gravis – like condition in which the eyelids progressively droop.

### **Are Certain Cancers The Predominant Causes Of Cancer Associated Retinopathy?**

Small cell carcinoma of the lung is the most commonly associated cancer with CAR, accounting for approximately two thirds of cases. Many other cancers have been associated with CAR, however, including breast cancer, lymphoma, thymoma, and testicular carcinoma.

Melanoma associated retinopathy is a different but related condition to cancer associated retinopathy. In melanoma associated retinopathy, the night blindness may reach a stable point. Unfortunately, in CAR, the disease frequently progresses to blindness.

## **Is There Any Treatment For CAR?**

The first task is to find the underlying cancer, if possible, and treat it. Occasionally, despite a thorough search, an underlying primary cancer cannot be found. In such cases, a search later on may find the cancer. To treat the loss of sight, doctors may try steroids, injections of immunoglobulin, or plasmapheresis, in which the blood is washed free of large proteins. Unfortunately, in many cases these attempts have minor effects.

### **Final Comments**

Cancer associated retinopathy and other paraneoplastic syndromes can severely handicap vision. Our present therapies are unsatisfactory in general. Further research is necessary to offer patients a better prognosis. The most important take home message is that a diligent and sometimes repetitive search for the underlying cancer is necessary, and treatment is addressed to that root cause of the problem.

After reading this brochure, if you have any questions, please call our office, at 704-295-3180. If you are interested in learning more on your own, we have developed a website dedicated to educating people about retinal disease. Our site is called *The Retina Reference* and it can be accessed at [www.retinareference.com](http://www.retinareference.com). This site includes numerous other pamphlets, which discuss retinal disease and treatment. There is also a forum available on the site, where patients can read about the experiences other patients have had and share their own experiences if they choose to do so. An additional resource we recommend is the website for the National Library of Medicine, on which there is a diverse collection of medical publications. We have included a link to this site on our website, but it can also be directly accessed via [www.pubmed.com](http://www.pubmed.com).

updated 7-30-18