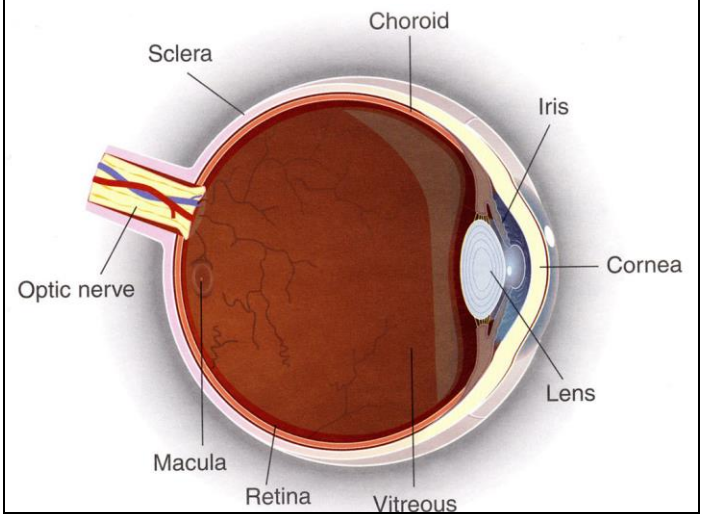
What You Should Know About Birdshot Chorioretinopathy

By David J. Browning, MD, PhD

Birdshot chorioretinopathy, also called vitiliginous chorioretinopathy, is an autoimmune disease of the eye in which depigmentation and inflammation occur in the retina and choroid, the neural and vascular linings, respectively, at the back of the eye. Figure 1 shows these anatomic structures. Women are affected more often than men with most patients developing the disease between the ages of 35 and 65. If left untreated, visual acuity slowly declines and inability to see at night or under low light conditions develops.

Figure 1. Anatomy of the Eye



What Causes Birdshot Chorioretinopathy?

The exact cause has yet to be discovered, but we know that host immune factors play an important role because 90% of affected patients possess the HLA-A29 antigen, one of the cell markers that allows immune surveillance cells to distinguish self from non-self. One hypothesis suggests that a triggering infection produces an immune reaction that becomes misdirected, attacking the choroid and outer retina.

Diagnosis

Most patients with birdshot chorioretinopathy develop painless blurring and floaters in both eyes. The ophthalmologist notes depigmented spots radiating from the optic disk, especially in the nasal fundus. A normal retina is shown in figure 2. Figure 3 shows the fundus of a patient with birdshot chorioretinopathy. The blood vessels frequently become leaky as determined by a series of photographs taken after intravenous injection of fluorescein dye (fluorescein angiography), and cysts of fluid can collect in the macula, the center of the retina where reading is done. Optical coherence tomography (OCT) can be used to measure this fluid build-up and monitor response to treatment. Sometimes electroretinography is used to monitor disease course. In this test, the voltage generated by the retina in response to light flashes is measured. Decreasing voltage over time implies worsening of disease.

Figure 2. Normal Retina

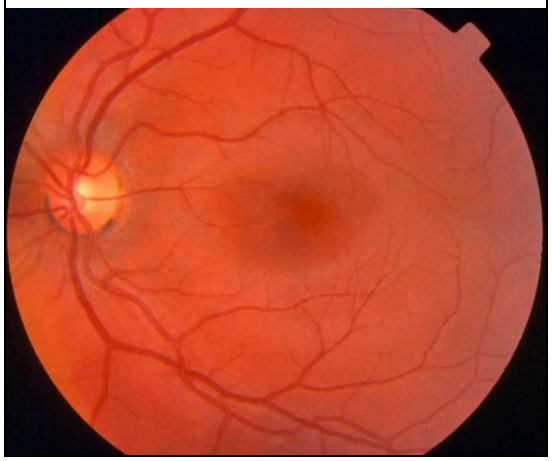
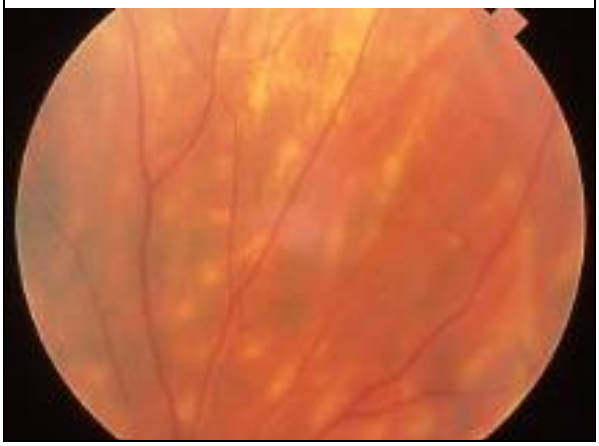


Figure 3. Birdshot



Treatment

The earliest treatments that occasionally worked were trials of corticosteroids, and even now these may be used sometimes. More consistently effective have been nonsteroidal immunomodulating drugs such as cyclosporine and mycophenolate mofetil. Both drugs require periodic blood testing to assess the possibility for renal and hematologic toxicity.

Final Comments

Once birdshot chorioretinopathy develops, treatment usually continues for years or even permanently to prevent loss of vision. With conscientious follow-up and care, the progress of disease in most patients can be halted, and vision maintained.

If you have questions, please contact Dr. Browning by email at contact@retinareference.com. If you would like to read more about uveitis, and excellent resource is the National Library of Medicine website, on the PubMed page. This site provides a wide variety of medical publications and can be accessed via any search engine or directly at the following link: www.pubmed.com.

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