
AMERICAN UVEITIS SOCIETY

Birdshot Retinochoroidopathy

A Patient Education Monograph prepared for the American Uveitis Society **June 2004**

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NOTE: The opinions expressed in this monograph are those of the author(s) and not necessarily those of the membership of the American Uveitis Society, its leadership, or the Editorial Board of UveitisSociety.org. All medical decisions should be made in consultation with one's personal physician.

INTRODUCTION

Birdshot retinochoroidopathy (also called birdshot chorioretinopathy and commonly shortened to simply "birdshot") is an inflammatory condition affecting both eyes. It is most often seen in Caucasians of northern or middle European descent. There is some evidence that birdshot may be more common in women, but not all studies support this. Birdshot most commonly begins when patients are in their early 50s, but the condition may start earlier or later in life.

HISTORY

Birdshot retinochoroidopathy was first well described in the early 1980's although it is clear that individuals who fit the description were reported earlier.

COURSE OF THE DISEASE

Birdshot retinochoroidopathy most often has a gradual onset with a subtle increase in symptoms. However, it can also present with the relatively rapid onset of floaters and decreased vision. Birdshot retinochoroidopathy is usually a chronic progressive disease, but which follows a differing course amongst patients. One of the greatest challenges faced by physicians and patients with this disease is how to prevent long term, low grade inflammation, which can lead to permanent loss of vision.

DIAGNOSIS AND TESTING

There is no test specific enough to establish without a doubt the diagnosis of birdshot retinochoroidopathy. Rather, the diagnosis is based on clinical findings in combination with blood tests. The name "birdshot" comes from the light, cream colored, round to oval lesions in the back part of the eye. There can also be inflammation of the retinal veins and swelling of the macula (the most sensitive part of the retina) as well as damage to the retina. In addition to the clinical findings, about 90% of Caucasian patients with birdshot retinochoroidopathy carry a genetic marker termed human leukocyte antigen (HLA)-A29. However, the vast majority of people who are HLA-A29 positive will never develop birdshot retinochoroidopathy. This means that HLA-A29 in and of itself is not the only reason one develops the condition and that other factors must

play a role as well.

Other forms of inflammation within the eye can mimic birdshot retinochoroidopathy and additional testing may be required to rule out such causes as sarcoidosis, syphilis, or intraocular lymphoma, among others.

TREATMENT

It is not always clear when to begin treatment. Furthermore, not every specialist feels that every patient needs to be treated. Some patients with minimal symptoms and good vision may not require treatment. While there appears to be a general trend toward more aggressive therapy, there are widely differing views on the subject. As with all such conditions, the decision of when and how to treat is something all patients should discuss frankly with their treating physician.

When treatment is decided upon, typically oral corticosteroids are the first agent used, but most individuals who require long term treatment are likely to be switched to an immunosuppressive agent such as cyclosporine, mycophenolate, methotrexate, azathioprine or others. Choice of which medication or combination of medications is best for this disease is guided primarily by a patient's other medical conditions. Studies are currently ongoing to understand which tests and clinical findings are best to follow for guiding decisions about response to therapy. Local injections of corticosteroids around or inside the eye may be used, especially for macular swelling. The role of new treatments, such as the corticosteroid implant is not yet established.

CAUSE OF THE CONDITION

The cause of birdshot retinochoroidopathy is not known. However, the tight correlation of the disease with the HLA-A29 marker suggests an autoimmune process.

PROGNOSIS

The prognosis for retention of good vision is guarded in patients with birdshot retinochoroidopathy. While some individuals will still have good vision after a prolonged period of time, many individuals have persistent complaints, such as difficulty with night and color vision. Others may have complaints such as "vibrating" vision, despite seeing well on the eye chart. In some cases there is significant visual loss due to recurrent complications such as macular swelling. It is very important that individuals with birdshot retinochoroidopathy maintain a close relationship with their treating physicians to ensure that complications such as macular edema can be aggressively treated when appropriate.

RESEARCH AND FUTURE OUTLOOK

Current research is primarily geared toward a better understanding of ways to monitor disease activity, the best ways to treat the disease long term, and research to look at the basic underlying causes of the disease.

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