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Birdshot Chorioretinopathy

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Birdshot chorioretinopathy, commonly referred to simply as “birdshot”, is a rare form of posterior uveitis which mainly affects the retina and choroid. The disease occurs in women more often than men, typically Caucasian from northern Europe, and most often between the ages of 30 and 70. “Birdshot” can be a severe and blinding disease if unrecognized or undertreated. The etiology of the disease is unknown; however, autoimmunity is considered to be the underlying cause. There is a class of genes known as human leukocyte antigens (HLA), and specifically one called HLA-A29, that is present in 80–98% of patients with birdshot disease. Infection, by virus or bacteria, in susceptible individuals is thought to act as a trigger, with the disease then being self-propagated by an autoimmune mechanism.

Birdshot chorioretinopathy is a chronic eye condition, with symptoms that develop and progressively worsen over several months to years. The symptoms include, floaters and light flashes, blurry or hazy vision (sometimes described as looking through murky water), decreased color and/or night vision, and visual field defects. Patients usually do not complain of pain, light sensitivity, or redness. Interestingly, central vision is preserved until the very late stages of the disease. Birdshot is confined to the eyes, and there are no systemic diseases typically associated with it.

Clinical examinations are the most important tool in the diagnosis of birdshot chorioretinopathy. A full history, review of systems, and complete eye examination should be performed to help differentiate birdshot from other infectious or noninfectious causes of uveitis. Eye examination often shows little to no inflammation in the “front” of the eye; however, dilated exam can reveal a dense collection of inflammatory cells and debris in the vitreous, pallor of the optic nerve, attenuation of retinal vessels, retinal vasculitis, macular edema, and the presence of characteristic birdshot lesions, creamy yellow-white spots involving the choroid. Cataract can be found as a result of long-standing inflammation or

chronic use of corticosteroids. Rarely, new blood vessels are seen between the choroid and the retina which can cause severe vision loss even after inflammation is treated.

Color vision testing can show mild to severe deficit. Fundus autofluorescence may reveal abnormal dark areas corresponding to birdshot lesions. Fluorescein angiography may show dark or bright areas of birdshot lesions or evidence of macular edema, retinal vasculitis, optic nerve inflammation, or choroidal neovascularization. Indocyanine green angiography, which is used to evaluate the choroid, can show dark lesions that may not be visible on dilated retinal exams or fluorescein angiography. Visual field testing with a blue-on-yellow protocol (shortwave automated perimetry, or SWAP), which is more sensitive than normal white-on-white visual field testing, will show visual field defects. Optical coherence tomography (OCT) can reveal macular edema or epiretinal membranes. Lastly, standard electroretinogram can be a useful tool in the diagnosis of birdshot and in assessing response to therapy, particularly the 30-Hertz photopic protocol, which often shows delayed implicit times and decreased signal amplitudes that may improve with treatment.

A broad range of laboratory tests can be done to rule out other inflammatory and infectious causes of posterior uveitis. However, the HLA-A29 gene is highly sensitive and specific for birdshot chorioretinopathy. All other laboratory test results are negative or within normal range in birdshot chorioretinopathy.

Birdshot disease is a treatable and potentially curable condition. Unfortunately, damage to the retina, choroid, and other parts of the eye is irreversible, making timely recognition and initiation of therapy essential for achieving a good outcome and preserving all visual functions. Corticosteroids, which are commonly used early in the treatment of most non-infectious uveitis, are not effective in birdshot and may even worsen the long-term prognosis. Our research has found that patients who have received oral corticosteroids have shown a higher rate of recurrence of disease, even after achieving remission on immunomodulatory therapy; however, injection of steroid into the eye (intravitreal injection) was not shown to have this risk. Additionally, generally speaking, topical eye drops are not effective in birdshot chorioretinopathy.

Immunomodulatory therapy is the standard of care for treating birdshot chorioretinopathy. Our years of experience treating patients with birdshot have shown that the most effective initial treatment strategy involves combination therapy with mycophenolate mofetil (CellCept®), an antimetabolite, and modified cyclosporine A, a T cell lymphocyte inhibitor. In cases of intolerance or poor efficacy, mycophenolate is sometimes replaced with another antimetabolite such as azathioprine, or the patient is transitioned to intravenous therapy with a TNF-α inhibitor such as infliximab (Remicade®).

Other biologic response modifier agents may also be tried after infliximab. Stubborn and poorly controlled disease on these medications may require use of an alkylating agent such as cyclophosphamide (Cytoxan®). New corticosteroid implants such as dexamethasone implant (Ozurdex®) and triamcinolone acetonide implant (ILuvien®) A corticosteroid implant surgically placed within the eye, fluocinolone (Retisert®), can be used for patients who are not able to achieve remission on or intolerant to immunodulatory therapy.

In regard to birdshot chorioretinopathy complications, cataract and glaucoma can occur as a result of active inflammation or long-term treatment with corticosteroids. Cataracts can be removed, ideally when inflammation has been treated and the eye is quiet for an extended period of time. Glaucoma is initially treated with eye drops, followed by laser therapy, and eventually glaucoma surgery if other treatments fail. Retina Problems with the central retina, especially macular edema and epiretinal membranes, are frequently seen. Macular edema may be resolved when inflammation is treated, however if it persists, it can be treated with topical NSAIDs or oral acetazolamide. Epiretinal membranes can be surgically removed only if they cause progressive vision loss or vision distortion. Finally, choroidal neovascularization can be successfully treated with anti-VEGF therapy, such as bevacizumab (Avastin®), or with corticosteroid injections.

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