Birdshot Chorioretinitis and Fundus Autofluorescence: Novel Insights Into Disease Pathogenesis

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Birdshot chorioretinitis (BSCR), an ocular autoimmune disease, is one of the most well-characterized posterior uveitis syndromes, with its strong HLA-A29 haplotype association and characteristic disease features (i.e., optic disc edema, periphlebitis, cystoid macular edema, and chorioretinal lesions leading to degenerative changes of the retina, choroid, and retinal pigment epithelium). The spectrum of ocular illness mandates judicious monitoring with diagnostic imaging and an understanding of immunotherapeutic options to avert loss of visual acuity and visual field.

In this edition of Investigative Ophthalmology & Visual Science, Böni et al.² describe their large multicenter, prospective experience that systematically defines patterns of fundus autofluorescence (FAF) in BSCR patients. Metrics of visual function including visual acuity, contrast sensitivity, color vision, and visual fields, which can all be impacted in patients with chronic, smoldering active inflammation in BSCR, are correlated with FAF patterns. The authors’ findings of any FAF abnormalities in nearly 80% of eyes and macular FAF abnormalities in 50% of all eyes are particularly notable given that peripheral chorioretinal lesions are most commonly described in the literature. These findings are a reminder that macular pathologies including cystoid macular edema and perifoveal disease require close assessment with both diagnostic imaging and functional testing in BSCR. Moreover, the association between confluent hypoautofluorescence and reduced visual acuity parallels other white dot syndromes where reduced perifoveal FAF signal is associated with visual acuity impairment,³ suggesting FAF as a valuable diagnostic tool for long-term structure/function correlation. Other key findings in this work include the association of retinal vasculitis with peripapillary confluent hypoautofluorescence and macular edema with global hypoautofluorescence. These correlates emphasize the need to evaluate whether FAF signal changes herald active inflammation or whether these abnormalities represent sequelae of disease progression, considerations relevant to the use of FAF imaging for monitoring of patient responses to therapy.

References