Infectious Nontuberculous Serpiginous Choroiditis

Serpiginous choroiditis (SC) is a rare, usually bilateral, progressive, recurrent, idiopathic inflammatory disease that primarily affects the inner choroid and retinal pigment epithelium. It was first described in 1904 by Junius but was defined as we now recognize it in the 1970s. Multiple names have been used to define this entity, but the term proposed by Gass in 1987 (serpiginous choroiditis) is the most widely accepted. As other cases were reported with atypical characteristics compared with the classic criteria, the nomenclature was broadened to include subtypes of SC, such as macular SC, ampiginous choroiditis, relentless placoid chorioretinitis, and macular helicoid chorioretinal degeneration. However, some authors consider serpiginous-like choroiditis (SLC) both a subtype of SC and a distinct clinical entity. Unlike SC, SLC is characterized by multifocal lesions in the posterior pole and periphery, usually sparing the juxtapapillary area, but with significant vitritis in all the cases. It has been associated with tuberculosis, although other causes, such as toxoplasmosis and sarcoidosis, mimic the entity. We describe herein the first 2 reported cases to date of SC secondary to Francisella tularensis and Bartonella henselae.

Report Of Cases. Case 1. A 51-year-old white man was seen at the emergency department with decreased visual acuity and the presence of floaters in his left eye. On examination, his best-corrected visual acuity was 20/20 OD and 20/125 OS. The intraocular pressure and anterior segment examination findings were unremarkable. A fundus examination of the left eye revealed multifocal yellow subretinal infiltrates, progressing centrifugally in an irregular serpentine fashion and sparing the peripapillary area (Figure 1A). Results of the fundus examination in the right eye were unremarkable. Fluorescein angiography (FA) showed early blockage of fluorescein and progressive diffuse staining and late leakage at the rim (Figure 1B and C).

Serum serological test results were negative for Lyme disease, syphilis, Brucella, toxoplasmosis, viral hepatitis, B henselae, Rickettsia, and human immunodeficiency virus. Mantoux test findings were negative, and chest radiographs were unremarkable. Test results for antinuclear antibodies, rheumatoid factor, antinuclear cytoplasmic antibodies, and antiphospholipid antibodies were also negative. Given that the patient hunted as a hobby, serological testing was performed for F. tularensis, with positive titers of 1:160, which increased to 1:640 after 10 days. Treatment with ciprofloxacin hydrochloride (500 mg twice daily) was initiated for 20 days. One month after treatment, the patient’s visual acuity had improved to 20/30, and the results of the fundus examination showed atrophy of the lesions (D), and fluorescein angiography showed no activity (E and F).

Case 2. A yellow subretinal infiltrate is seen in the right eye in the macular area (A), with hyperautofluorescence surrounded by a hypoautofluorescent halo on fundus examination (B) and leakage at the rim in the last frames of fluorescein angiography (C). D. Three months after antituberculosis treatment, a lesion was seen in the left eye. After treatment of the left eye with moxifloxacin hydrochloride, progression of the lesion ceased (E), and fluorescein angiography showed no activity (F).
examination and FA demonstrated no active lesions (Figure 1D-F).

After 12 months, the disease showed no progression. Lesions observed on fundus examination and FA remained inactive, and serological test results for *F. tularensis* were negative for organisms.

**Case 2.** A 36-year-old white man reported blurry vision and flashing lights in the right eye for 2 days. His best-corrected visual acuity was light perception OD and 20/20 OS. The intraocular pressure and anterior segment examination findings were unremarkable. A fundus examination of the affected eye disclosed a macular yellow subretinal infiltrate, sparing the peripapillary area (Figure 2A). Fluorescein angiography showed early blockage of fluorescein and progressive staining, with leakage at the rim in the late phases (Figure 2C). The results of laboratory studies and serological testing were negative except for the Mantoux test findings (18 mm) and IgG titers of 1:128 for *B. henselae*. Anti-tuberculosis treatment and a corticosteroid regimen for 9 months were started. Three months after the beginning of treatment, a new lesion was observed in the left eye (Figure 2D), so moxifloxacin hydrochloride (400 mg once daily) was initiated for 1 month against *B. henselae*, improving the inflammatory status and stopping the progression of the macular lesion (Figure 2E). Six months later, lesions observed on fundus examination and FA remained inactive (Figure 2F), and serological test results for *B. henselae* were negative for organisms.

**Comment.** Serpiginous choroiditis is a rare and sight-threatening disorder classically characterized by peripapillary involvement, with centrifugal progression in a serpiginous morphologic form (snake shape) and a lack of systemic disease. However, the observation of patients with atypical characteristics led to a modification of the strict concept of SC, expanding it to cover new subtypes. Serpiginous choroiditis is generally associated with idiopathic causes, as SC is related to tuberculosis; however, among all the reported cases of SC, only 7% have had positive Mantoux test results. From our viewpoint, all the types of SC and the variant termed *serpiginouslike choroiditis* represent a spectrum of one disease regardless of the responsible factor. The choroid can react to any inflammatory or infectious insult in the classic form or atypical form of SC, no matter what the cause.

Case 1 herein could be defined as SLC secondary to *F. tularensis*, although it deviates from defined features that characterize SLC; it shows a multifocal pattern but without juxtapapillary involvement, so it could be defined as atypical SLC, which reinforces the theory of one unique disease with varied spectra of presentation. Serpiginouslike choroiditis may be a variant form of SC in which tuberculosis could have a more relevant role than other origins, probably not yet identified. However, serological testing for other causes is advisable regardless of the subtype of SC. Case 2 is an example of SLC due to *B. henselae*, which responded to antibiotic therapy, as did case 1.

Serpiginouslike choroiditis cases associated with systemic disease, both infectious and autoimmune, have been published, with modifications of the diagnostic criteria for classic SC. Herein, we report 2 new cases of infectious SLC. Both cases demonstrate the need for ruling out an infectious origin in all the patients with SC regardless of subtype.

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