Early Presentation of a Known Diagnosis

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Introduction:

A 49-year-old female presented to The Retina Institute for decreased vision in her left eye. The patient reported bumping her head 2 nights earlier and noticed a "kaleidoscope" in her vision the following morning. By the next day, a large, stationary, "donut-shaped" spot was in her central vision. The patient had a history of type 2 diabetes, hypertension, hyperlipidemia, and depression. Review of systems was positive for a recent flu-like illness one week ago.

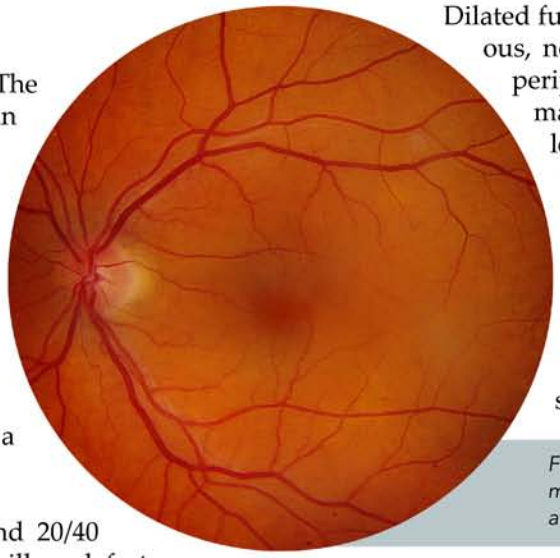

Figure 1: Fundus photos notable for mild mottling and peripapillary atrophy.

Visual acuity measured 20/20 OD and 20/40 OS. There was no relative afferent pupillary defect. Intraocular pressure was normal. Anterior segment exam was quiet and notable for pseudophakia OU.

Dilated fundus exam revealed clear vitreous, normal vasculature, and normal periphery OU. The optic nerve and macula were normal OD but the left eye exhibited mild mottling of the macula OS (Figure 1). Fluorescein angiogram (FA) was normal in the right eye but revealed mild leakage from telangiectatic vessels at the temporal disc OS (Figure 2). OCT was normal OD but showed subretinal fluid (SRF) with intraretinal fluid in the fovea and papillomacular bundle (Figure 3). Humphrey visual fields exhibited a central scotoma and nasal depression OS. (Figure 4).

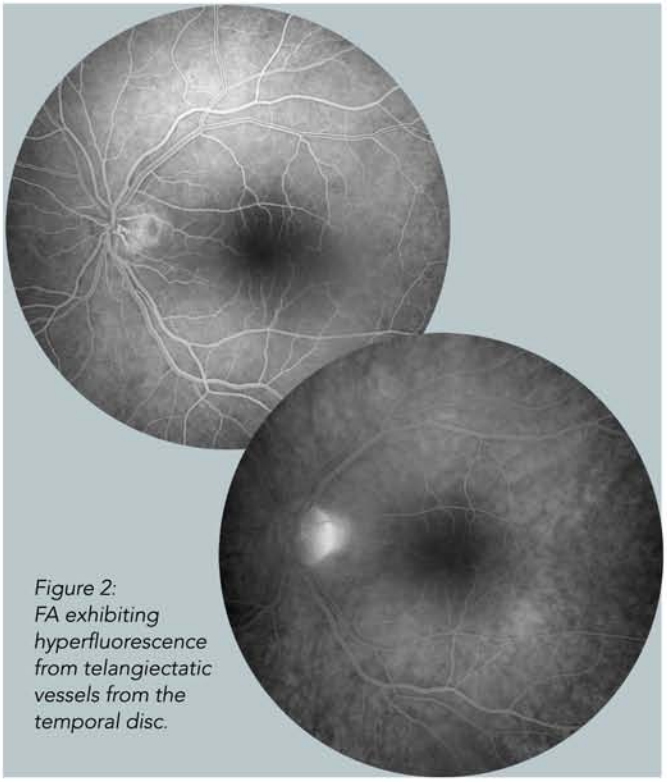

Figure 2: FA exhibiting hyperfluorescence from telangiectatic vessels from the temporal disc.

Diagnosis:

The differential diagnosis for this patient's disc edema, intraretinal fluid, and subretinal fluid includes

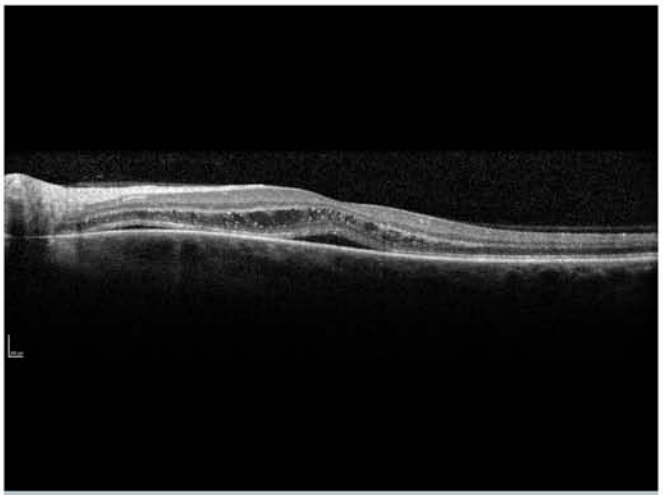

Figure 3: OCT with intraretinal fluid and SRF in the fovea and PM bundle.

exudative age-related macular degeneration, central serous retinopathy, diabetic macular edema, cystoid macular edema, neuroretinitis, and optic nerve pit. There was no drusen, retinal vasculature leakage, diabetic changes, inflammation, or visible optic nerve pit. The patient did endorse a recent fever and was an enthused owner of multiple cats so a laboratory work-up was sent for Lyme and Bartonella titers. Lyme antibodies were negative but Bartonella antibodies were positive for Henselae IgM/IgG and Quintana IgG. The patient was diagnosed with early neuroretinitis (aka cat-scratch disease) before formation of a clinically detectable macular star.

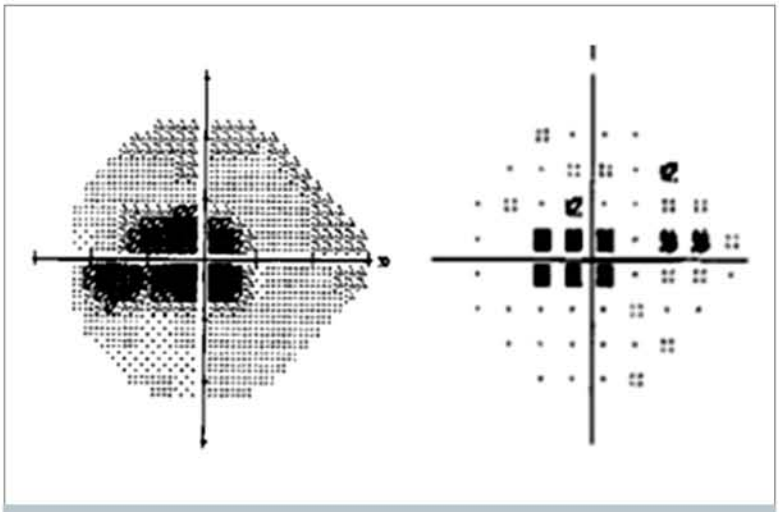

Figure 4: HVF (with pattern deviation) OS demonstrating a paracentral scotoma and nasal depression OS.

Bartonella spp. are intracellular bacteria that cause prolonged intraerythrocytic bacteremia in their hosts and are typically transmitted by insects such as cat fleas, phlebotomine sandflies, human body lice or via cat scratches, bites, or saliva. To date, more than 30 Bartonella spp. have been isolated from humans but Bartonella henselae, B. quintana and B. bacilliformis are responsible for the majority of infections. Bartonella is confirmed serologically and an increase in immunoglobulin IgM titer with a significant increase in IgG antibodies titer is considered an acute Bartonella infection. Approximately two-thirds of patients have positive testing.

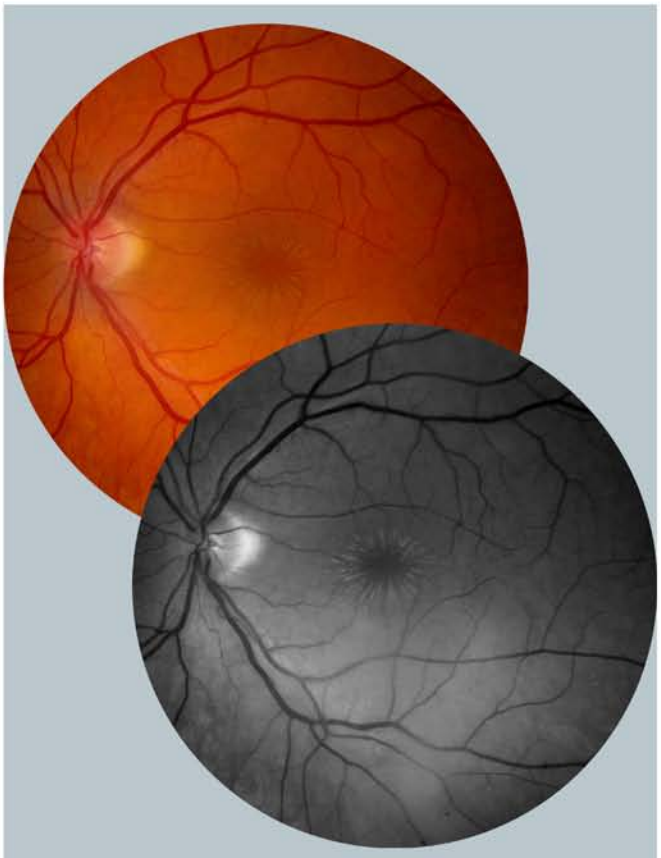

Figure 5: Fundus and red-free photograph with macular star one week after presentation.

Systemic and Ocular Manifestations:

Bartonella infections are usually a self-limiting disease in immunocompetent patients. Systemic clinical manifestations of the intraerythrocytic stage include lymphadenopathy, fever, malaise, nausea, myalgia, arthralgia, and hepatosplenomegaly. Bartonella spp. can subsequently colonize secondary foci leading to endocarditis, hepatitis, encephalopathy, meningitis, transverse myelitis, hemolytic anemia, glomerulonephritis, and osteomyelitis. Immunocompromised patients can also exhibit bacillary angiomatosis which can resemble Kaposi's sarcoma histologically.

Ocular manifestations include anterior and/or posterior uveitis, neuroretinitis, retinochoroiditis or Parinaud's oculoglandular conjunctivitis (characterized by ipsilateral preauricular and submandibular lymph node swelling). It usually presents unilaterally but bilateral

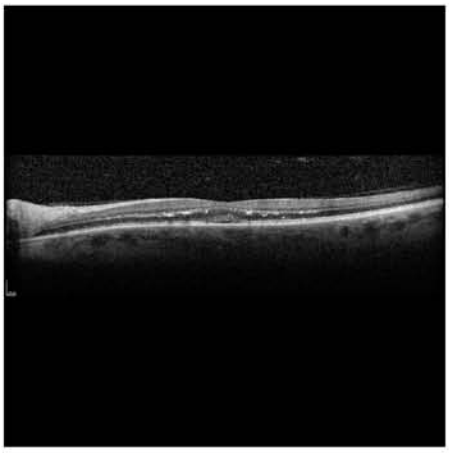

Figure 6: OCT with decreased SRF and hyperreflective intraretinal deposits in the outer plexiform layer one week after presentation.

cases have been reported. The primary process of inflammation in the posterior segment of the eye involves the disc vasculature with subsequent exudation of fluid into the retinal outer plexiform layer around the peripapillary retina/macula. Neuroretinitis occurs in 1-2% of patients with Bartonella infection and is characterized by optic disc swelling, retinal hemorrhages, intraretinal and/or subretinal fluid, and partial or complete macular star (retinal deposits located in the outer plexiform layer). This process typically takes 2-4 weeks to resolve. More serious complications include retinal vascular occlusion, neovascular glaucoma, macular hole, choroiditis, retinal granulomas, and papillary vasoproliferative changes.

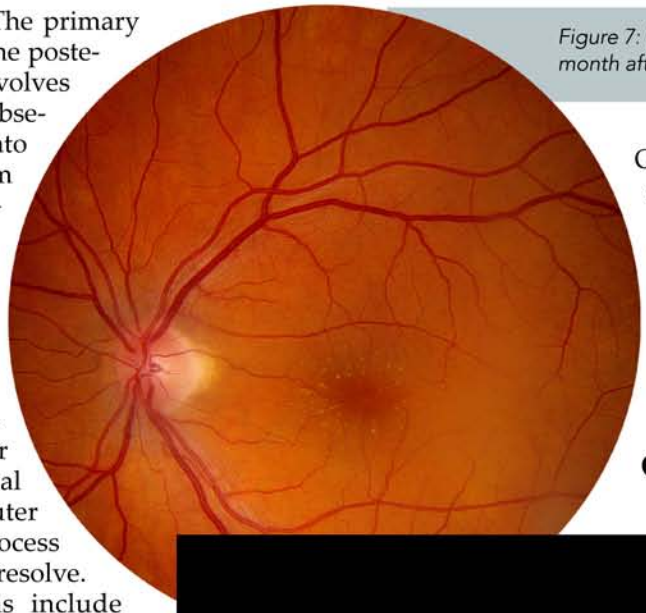

Figure 7: Near resolution of macular star one month after presentation.

One month after presentation, the subjective central scotoma decreased and acutiy was 20/30. The macular star was nearly completely resolved with decreased intraretinal hyperreflective deposits on the OCT, (Figure 7 and 8).

Treatment:

Indication for treatment is based on severity of ocular involvement and/or systemic manifestations. Prompt antibiotic treatment may improve visual outcome and hasten visual recovery. Recommended antibiotics for bartonellosis include doxycycline, azithromycin, trimethoprim/sulfamethoxazole, ciprofloxacin, gentamicin, and rifampicin, which have reported to have good efficacy. Several studies have shown that patients treated with concomitant corticosteroids for ocular manifestations had a good response as well.

Follow-up:

One week after initial presentation, this patient's central scotoma persisted and vision fell to 20/60 OS. A macula star formed and serologic results confirmed Bartonella infection, (Figure 5). OCT showed decreased SRF and the formation hyperreflective deposits in the outer plexiform layer, (Figure 6). The patient was subsequently started on Doxycycline 100 mg BID for one week and sent for an infectious disease consultation.

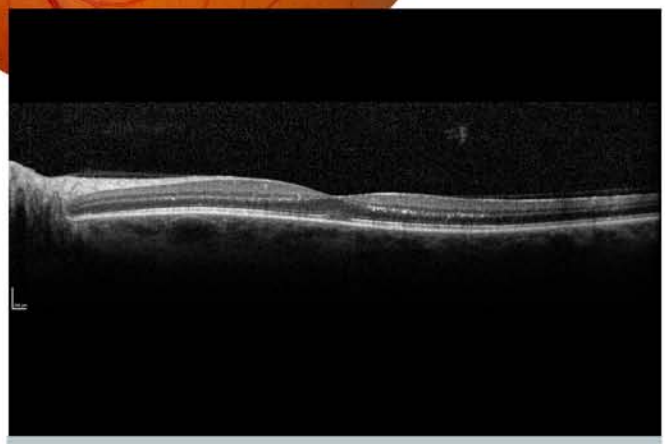

Figure 8: Resolution of SRF, few persistent hyperreflective deposits in OPL one month after presentation.

Conclusion:

Neuroretinitis from Bartonella infection can manifest with disc edema, retinal hemorrhages, macular star, intraretinal and/or subretinal fluid. Serologic testing can confirm the diagnosis but careful ocular exam/imaging as well as a detailed review of systems can determine if antibiotic therapy +/- steroids may be indicated. While most cases are self-limiting, treatment may hasten recovery, improve outcomes, and prevent serious ocular and systemic complications.

References:

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